

International Pediatric Orthopaedic Pocketbook (IPOP)

A Global HELP
Publication

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2009

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International Pediatric Orthopaedic Pocketbook (IPOP)

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Author's Comment

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1 Andry's Tree

Pediatric orthopedics is a subspecialty of medicine that deals with the prevention and treatment of musculoskeletal disorders in children. In 1741, Nicholas Andry, professor of medicine at the University of Paris, published his treatise describing different methods of preventing and correcting deformities in children [1]. He combined two Greek words, *orthos*, or straight, and *paidios*, child, into one word, "orthopedics," which became the name of the speciality concerned with the preservation and restoration of the musculoskeletal system. Pediatric orthopedics is central to this specialty because of Andry's original focus on childhood problems, because of the large proportion of orthopedic problems that originate during the early period of growth, and finally, because pediatric orthopedics offers a dynamic and inherently interesting subspecialty.

A knowledge of normal and abnormal growth and development is vital to an understanding of pediatric orthopedics [2]. This knowledge increases our comprehension of the musculoskeletal system, improves our understanding of the causes of disease, and makes us better able to manage the varied orthopedic problems of childhood.

Dividing the period of growth into seven stages provides a convenient framework to review both normal and abnormal growth and development [3]. During the first stage, reproductive cells or gametes are formed.



2 Femoral torsion. Femoral torsion is often familial. Many common musculoskeletal problems have a genetic basis.

Category	Period
Gamete	Prior to fertilization
Early Embryo	0–2 weeks
Embryo	2–8 weeks
Fetus	8 weeks to birth
Infant	Birth to 2 years
Child	2 years to puberty
Adolescent	Transition to maturity

3 Growth phases. The period of growth can be divided into seven phases.

2 Growth / Embryo

Normal Growth

Gamete

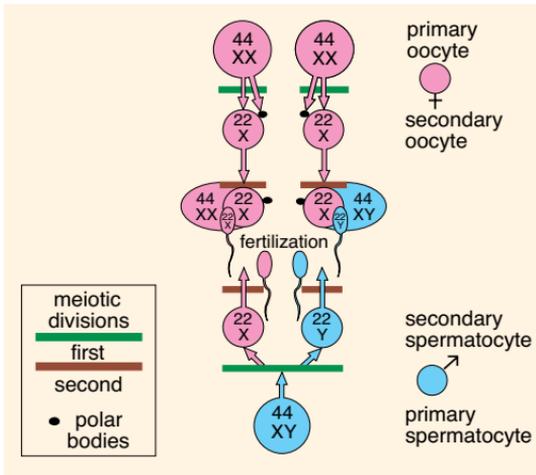
Gamete is a collective term for ovum and sperm. During gametogenesis, meiotic division halves the chromosome number. Genetic material, which may include defective genes, is shuffled, and mature ova and sperm are formed [1].

Early Embryo

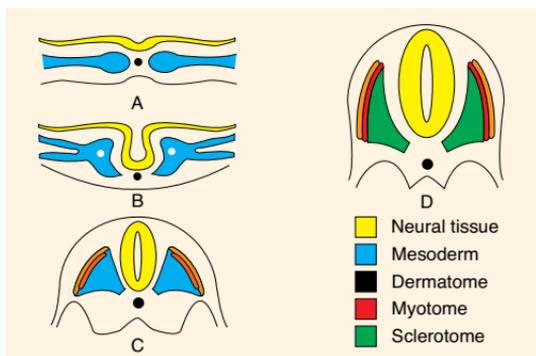
This early embryonic phase encompasses the 2-week period from fertilization to the implantation of the embryo.

First week During the first week following fertilization, the zygote repeatedly divides as it moves through the fallopian tubes to the uterus. The zygote becomes a morula, then a blastocyst. The blastocyst implants itself on the posterior uterine wall.

Second week During this week, the amniotic cavity and trilaminar embryonic disc are formed [2]. The early embryo is usually aborted if a lethal or serious genetic defect is present. During these first two weeks, the early embryo is less susceptible to teratogens than during the following embryonic period.



1 Gametogenesis. The ovum and sperm are formed by two meiotic divisions that halve the chromosome number and shuffle genetic material. Fertilization combines the traits of both parents to create a unique individual.



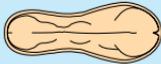
2 Trilaminar Disc. The neural tube closes. The mesoderm differentiates into dermatome, myotome, and sclerotome.

Embryo

The organ systems of the body develop during the embryonic period. Differentiation to more specialized tissue occurs through complex mechanisms such as induction. Induction is the process by which cells act on other cells to produce entirely new cells or tissue.

Third week This is the first week of organogenesis. During this week, the trilaminar embryonic disc develops, somites begin to form, and the neural plate closes to form a neural tube.

Fourth week During this week, the limb buds become recognizable [1]. Somites differentiate into three segments. The dermatome becomes skin, the myotome becomes muscle, and the sclerotome becomes cartilage and bone. The apical ectodermal ridge develops in the distal end of each limb bud. The ridge has an inductive influence on limb mesenchyme, which promotes growth and development of the limb. Serious defects in limb development may originate at this time.

	AGE wks.	SIZE mm.	Shape	Form	Bones	Muscles	Nerves
Embryo				Trilaminar notochord			Neural plate
				Limb buds	Sclerotomes	Somites	Neural tube
				Hand plate	Mesenchyme condenses	Premuscle	
		12		Digits	Chondrification	Fusion myotomes	
		17		Limbs rotate	Early ossification	Differentiation	
		23		Fingers separate		Definite muscles	Cord equals vertebral length
Fetus	12	56		Sex determined	Ossification spreading		
	16	112		Face human	Joint cavities	Spontaneous activity	
	20-40	160-350		Body more proportional			Myelin sheath forms; cord ends L3

1 Prenatal development. This chart summarizes musculoskeletal development during embryonic and fetal life.

4 Growth / Embryo

Fifth week The hand plate forms and mesenchymal condensations occur in the limbs.

Sixth week The rays of the digits become evident and chondrification of mesenchymal condensations occurs.

Seventh week The notches appear between the digit rays. Failure of the separation of rays results in syndactylism. During this week, the upper and lower limbs rotate in opposite directions [1]. The lower limb rotates medially to bring the great toes to the midline, whereas the upper limb rotates about 90° laterally to position the thumb on the lateral side of the limb.

Eighth week The fingers separate completely, the embryo assumes a human appearance, and the basic organ systems are completed.

Fetus

The fetal period is characterized by rapid growth and changes in body proportions.

Ninth to twelfth weeks The first bone, the clavicle, ossifies by a process of intramembranous deposition of calcium. The upper limbs become proportionate compared to the rest of the body, but the lower limbs remain short.

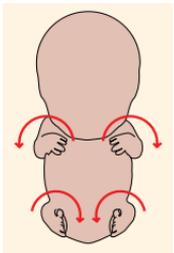
Thirteenth to twentieth weeks Growth continues to be rapid. The lower limbs become proportionate and most bones ossify. The fetal period is characterized by rapid growth and changes in body proportions.

Twentieth to fortieth weeks Growth continues and body proportions become more infant-like.

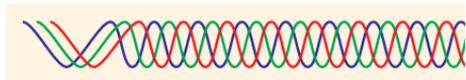
Connective Tissue

During early fetal life, the basic structure of connective tissue is formed largely of two families of macromolecules—collagens and proteoglycans.

Collagen Collagen is a family of proteins containing a triple helix of peptide chains [2]. Although at least ten different types of collagen are known, five types are most common [3].



1 Limb rotation. During the seventh week, the upper limb rotates laterally. The lower limb rotates medially to bring the great toes to the midline.



2 Collagen helix. A triple helix of peptide chains form the basic collagen structure.

Location	Type	Comment
Interstitial	I	Ubiquitous, skin, tendons
	II	Cartilage and nucleus pulposus
	III	Like type I, but absent in bone
Segmentation	IV	Lens and kidney
	V	Minor component of bone

3 Collagen types. Five basic collagen types in human connective tissue.

The biosynthesis of collagen starts in the endoplasmic reticulum, where the basic molecule is assembled. In the extracellular space, procollagen is formed. It is arranged into fibrils and reinforced by cross-linkages to become collagen. Collagen is the major component of connective tissue.

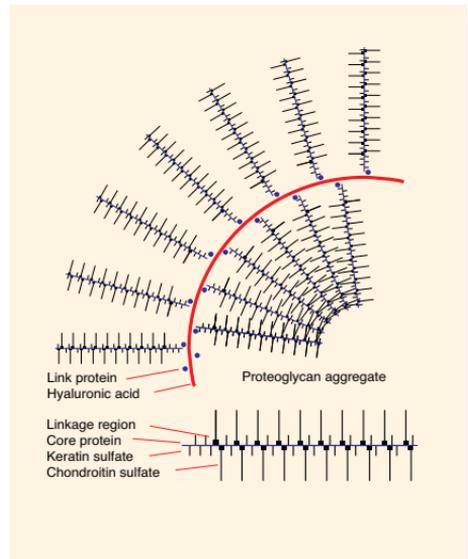
Disorders of collagen are common. They may be minor, producing only increased joint laxity [1], or severe, causing considerable disability. The major collagen disorders are classified according to the site of the defect in the pathway of collagen biosynthesis.

Proteoglycans (mucopolysaccharides) Proteoglycans are macromolecules that form the intracellular matrix of hyaline cartilage and the other connective tissues. Polypeptides or proteins attach to glycosaminoglycan to become proteoglycans [2]. Proteoglycans attach to a hyaluronic acid by a link protein to become an aggregate with a molecular weight in excess of one million. Proteoglycans are highly hydrophilic, and in water, they combine with many times its weight of water to create an elastic matrix that is ideal for joint lining. Hyaline cartilage is composed of about equal amounts of proteoglycans and collagen, and it combines with about three times their weight of water. Defects in the formation of these complex molecules produce a variety of diseases.

Mucopolysaccharide (MPS) storage diseases result from a deficiency of specific lysosomal enzymes necessary for the degradation of glycosaminoglycans. These diseases are caused by intracellular accumulation of partially degraded molecules that result in cell dysfunction or death.



1 Clinical manifestations of collagen types. Variations of collagen types are common in pediatric orthopedics. This child has developmental hip dysplasia with extreme joint laxity.



2 Proteoglycan aggregate. These massive molecules combine with water to form a resilient matrix such as that of hyaline cartilage.

6 Growth / Bone Formation

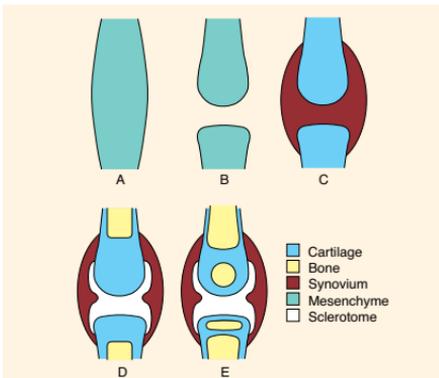
Synovial Joints

Synovial joints develop first as a cleft in the mesenchyme, which then chondrifies and cavitates [1]. Cavitation is completed by about the fourteenth week, with the inner mesenchyme becoming synovium and the outer mesenchyme becoming the joint capsule. Normal joint development requires motion, and motion requires a functioning neuromuscular system. Thus, defective joints are often seen in infants with neuromuscular disorders such as myelodysplasia or amyoplasia.

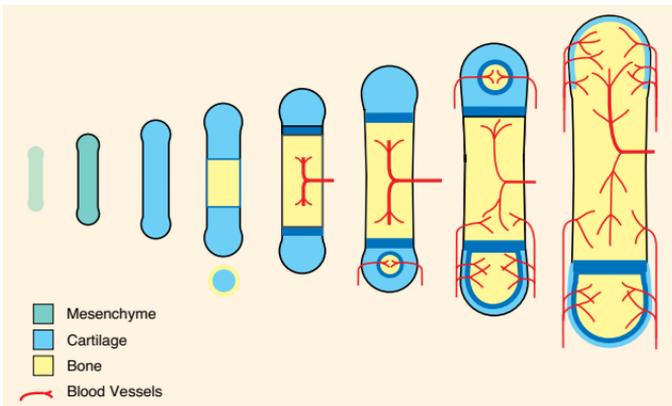
Bone Formation

Bones form in stages. First, mesenchymal cells condense to become models for future bones. The second stage, chondrification, is a time of rapid interstitial growth. Finally, cartilage is converted to bone by intramembranous and endochondral ossification.

Endochondral ossification takes place in most bones [2]. During the fetal period, primary ossification centers develop in long bones within the diaphysis. Ossification first occurs under the perichondrium. Within the cartilage, hypertrophied cells degenerate. Next, vascular ingrowth occurs, and then the core of the cartilage model is ossified to form the primary ossification center. Endochondral ossification proceeds at the cartilage–bone interphase. Later, secondary ossification centers develop at the ends of the bones, and the cartilage interposed between the primary and secondary ossification centers becomes the growth plate.



1 Synovial joint formation. The synovial joints form first as condensations of mesenchyme. Cavitation, chondrification, synovial differentiation, and finally ossification complete the basic structure.



2 Endochondral ossification. A typical long bone is preformed in mesenchyme. Chondrification precedes ossification.

Primary ossification centers for long bones usually develop before birth [1], whereas primary ossification centers for smaller bones, such as the patella and most carpal and tarsal bones, develop during infancy. Secondary ossification centers develop during infancy and early childhood. They fuse with the primary centers during late childhood, adolescence, and early adult life. Because osseous maturation continues throughout childhood and adolescence in a reasonably orderly fashion, the extent of ossification, as radiographically documented, has become the standard for assessing maturation.

Woven bone is formed during the fetal period. This bone has less structure, a relatively higher collagen content, and more flexibility than lamellar bone. This flexibility becomes essential during the transverse of the birth canal. Woven bone is gradually replaced by lamellar bone during infancy, and little remains in childhood.

Cortical thickness also increases throughout childhood. For example, the diameter of the diaphysis of the femur increases faster than the diameter of the medullary canal. This produces an increasing diaphyseal thickness with advancing age. This increasing thickness, lamellar structure, and proportion of calcium give mature bone great tensile strength but little flexibility. These changes are important factors in producing the varying patterns of skeletal injury seen during infancy, childhood, and adult life.

Growth Plate

The growth plate of long bones develops between the primary and secondary ossification centers. The function of the growth plate is to produce longitudinal growth [1 next page]. This is accomplished by a complex process of proliferation and maturation of chondrocytes, matrix production, and mineralization, followed by endochondral ossification. Growth plates with more limited growth potential develop at other sites. These include the periphery of round bones, such as the tarsal bones or vertebral bodies, and the sites of muscle attachments, such as the margins of the ilium. Such sites are referred to as *apophyses*.

The typical long bone epiphysis is divided into zones that reflect morphological, metabolic, and functional differences.

The reserve zone (RZ) is adjacent to the secondary ossification centers and is a zone of relative inactivity. The RZ does not participate in the longitudinal growth of the bone, but it does provide some matrix production and storage functions.



1 Radiograph of bones of a newborn infant. This radiograph shows primary ossification of the skeleton. Much of the skeleton is cartilage at this age.

8 Growth / Growth Plate

The proliferative zone (PZ) is the zone of cartilage cell replication and growth. A high metabolic rate and abundant blood supply, oxygen, glycogen, ATP, and collagen make this rapid growth possible.

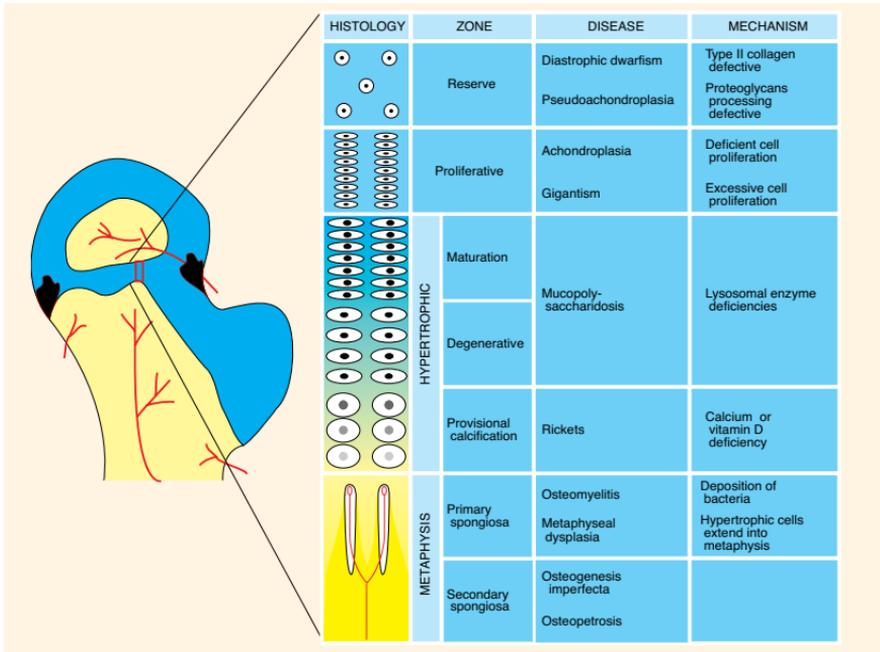
The hypertrophic zone (HZ) consists of three subzones: maturation, degeneration, and provisional calcification segments. In the HZ, the cartilage cells increase in size and the matrix is prepared for calcification. This is associated with a decline in blood supply, oxygenation, and glycogen stores and with a disintegration of aggregated mucopolysaccharides and chondrocytes. In the subzone of provisional calcification, a unique collagen X is synthesized that accepts calcium deposition.

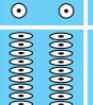
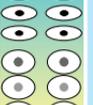
The metaphysis is the site of vascularization, bone formation, and remodeling. The calcified matrix is removed, and fiber bone is formed and replaced by lamellar bone.

The periphery includes the growth plate and metaphysis, which are the primary sites for infections, neoplasms, fractures, and metabolic and endocrine disorders. Problems in the growth plate constitute a significant portion of diseases of the musculoskeletal system in childhood.

Bone Growth

The rate of growth may be retarded by many factors, such as injury, disease, and medical procedures. Brief periods of growth retardation may produce growth arrest lines. These lines may be visible on radiographs [1 next page].



HISTOLOGY	ZONE	DISEASE	MECHANISM	
	Reserve	Diastrophic dwarfism	Type II collagen defective	
		Pseudoachondroplasia	Proteoglycans processing defective	
	Proliferative	Achondroplasia	Deficient cell proliferation	
		Gigantism	Excessive cell proliferation	
	HYPERTROPHIC	Mucopolysaccharidosis	Lysosomal enzyme deficiencies	
				Provisional calcification
		METAPHYSIS	Primary spongiosa	Osteomyelitis
	Metaphyseal dysplasia		Hypertrophic cells extend into metaphysis	
	Secondary spongiosa		Osteogenesis imperfecta	
		Osteopetrosis		

1 Growth plate. This section from the proximal femoral epiphysis is enlarged to show the histology and disordered growth that occurs at various levels of the growth plate.

Nervous System Development

During the third week of fetal life, the neural plate develops as a thickening of the dorsal portion of the ectoderm [2]. The neural plate then infolds to form the neural groove in the center, with neural folds on each side. During the fourth week, the neural groove closes to become the neural tube, and the neural crest separates and becomes interposed between the neural tube and surface ectoderm.

The neural crest becomes the dorsal root ganglia and the dorsal or sensory roots. The ventral or motor roots arise from the basal plates on the ventrolateral aspect of the neural tube. The combination produces the peripheral nerves.

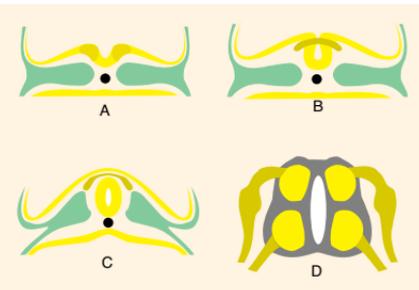
Peripheral nerves grow into the forming limb buds of equivalent somites, penetrating the mesenchyme, and are distributed to the developing muscles. Cutaneous sensation is also provided in a segmental fashion.

Myelination of the spinal cord forms during the late fetal period and continues into early infancy.

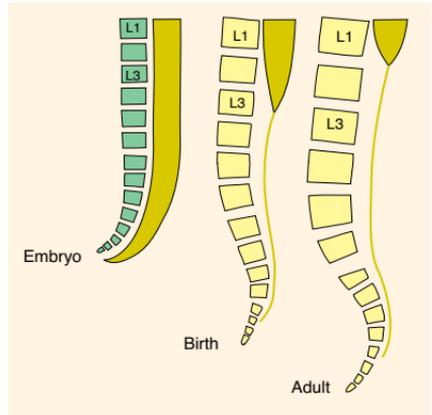
Initially, the neural and bony elements of corresponding somites lie opposite each other. Thus, the caudal end of the spinal cord fills the spinal canal, and the spinal nerves pass through the corresponding intervertebral foramina. By the 24th fetal week, the cord ends at S1; at birth, at L3; and in the adult, at L1 [3]. This differential growth rate results in the formation of the caudal equina: the accumulation of the nerves traversing the subarachnoid space to the intervertebral foramina. The end of the cord is attached to the periosteum opposite the first coccygeal vertebra by the filum terminale. The filum is the residual of the embryonic spinal cord.



1 Growth lines. Note the growth arrest lines (arrows) in this child with developmental hip dysplasia. Presumably the anesthetic and closed reduction caused the arrest. Bone growth since the arrest is shown by the width of the new metaphyseal bone.



2 Development of the nervous system. The nervous system is formed from the neural plate. (A) Infolding. (B) Neural crest. (C) Tube closure. (D) Dorsal and ventral root formation.



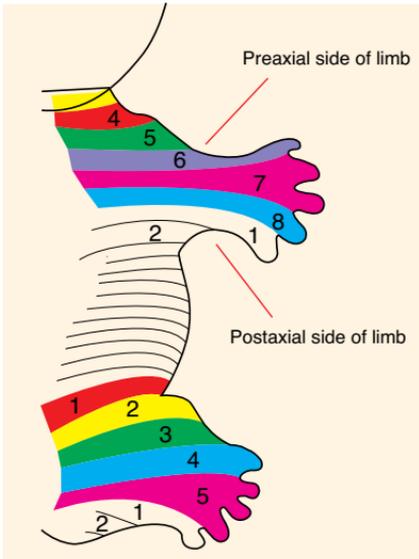
3 Spinal cord vertebral column relationship. During the fetal period, the spinal cord fills the vertebral canal. With growth, the cord ends at a progressively higher level.

10 Growth / Muscle Development

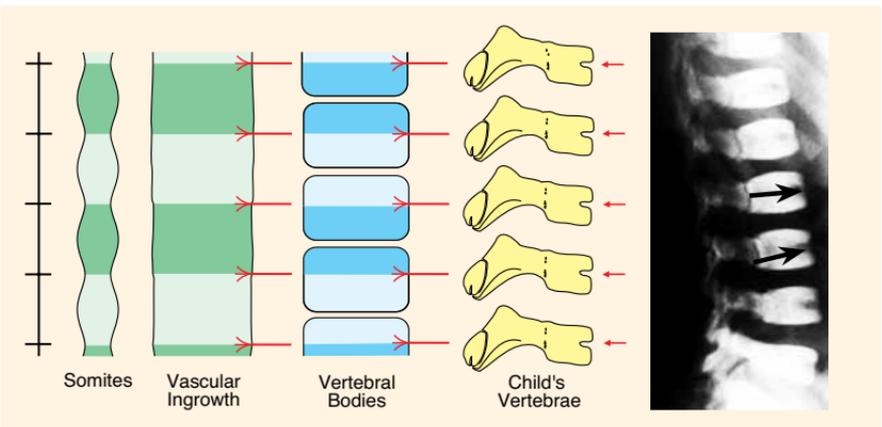
Somites produce a dermatomal pattern of sensory distribution. This simple pattern [1] becomes complicated by the rotation of the limb.

Muscle Development

Mesoderm of the somites' myotome segments produce myoblasts, which in turn produce the skeletal muscle of the trunk. Somatic mesoderm produces the limb buds' mesenchyme, which then forms limb muscles. Limb muscles develop from mesenchyme of the limb buds, which originate from somatic mesoderm. Individual muscles are present by the eighth fetal week. Muscle fibers increase in number before and after birth. Between 2 months of age and maturity, muscle fibers increase about 15-fold in the male and 10-fold in the female. Increase in the size of fibers occurs most rapidly after birth, increasing the muscle component of body weight from about one-fourth at birth to nearly half in the adult.



1 Dermatomes. Somites produce dermatomes that are simple and well delineated. The simple pattern is altered by subsequent limb rotation.



2 Vertebral intersegmental development. The vertebral bodies form as intersegmental structures. As blood vessels grow between somites, their final position is midvertebral. The site of blood vessel entry and somite fusion is sometimes seen radiographically as an anterior notch in the vertebral body of the child (arrows).

Vertebral Column Development

The axial system develops during the embryonic period. During the fourth week, mesenchymal cells from the sclerotome grow around the notochord to become the vertebral body and around the neural tube to form the vertebral arches [1]. Cells from adjacent sclerotomes join to form the precursor of the vertebral body, an intersegmental structure. Between these bodies, the notochord develops into the intervertebral disc. Cells surround the neural tube to become the vertebral arches.

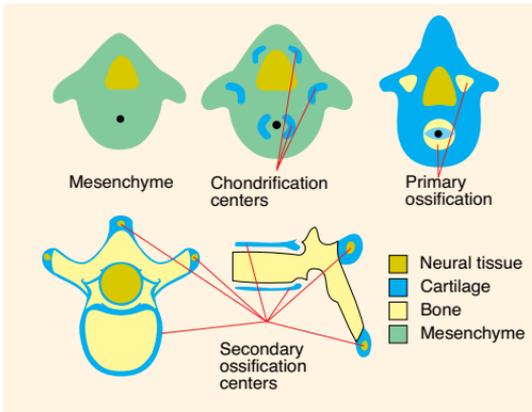
During the sixth fetal week, chondrification centers appear at three sites on each side of the mesenchymal vertebrae. The centrum is formed by the coalition of the two most anterior centers. Chondrification is complete before the ossification centers appear [2]. The centrum, together with an ossification center of each arch, make a total of three primary ossification centers for each vertebra.

During early childhood, the centers of each vertebral arch fuse and are joined to the vertebral body by a cartilaginous *neurocentral junction*. This junction allows growth to accommodate the enlarging spinal cord. Fusion of the neurocentral junction usually occurs between the third and sixth years. Anterior notching of the vertebrae is sometimes seen in the infant's or child's vertebrae and shows the site of somite fusion [2 opposite page].

Secondary ossification centers develop at the ends of the transverse and spinous processes and around the vertebral end plates at puberty. These fuse by age 25 years. Congenital defects are common in the axial system. Variations in the lumbar spine occur in about one-third of individuals. Spina bifida occulta is common. Hemivertebrae result from a failure of formation or segmentation. Such lesions are frequently associated with genitourinary abnormalities and less frequently with cardiac, anal, and limb defects and with tracheoesophageal fistula.



1 Sclerotome growth. Cells from the sclerotome grow around the notochord and neural tube.



2 Vertebral development. Vertebrae develop first as mesenchyme, then cartilage, and finally bone. Secondary ossification centers develop during childhood and fuse during adolescence or early adult life. From Moore (1988).

12 Growth / Infancy

Infancy

Infancy extends from birth to 2 years of age. It encompasses the period of most rapid growth and development after birth.

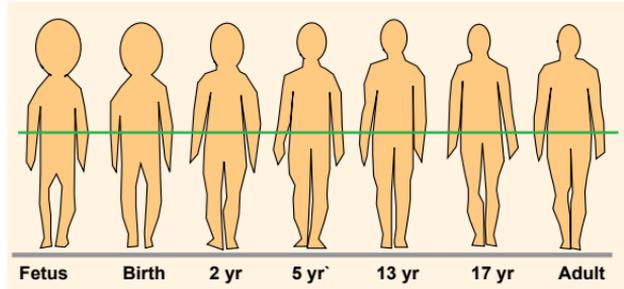
Body proportions Growth of various body parts are different from one another. Upper limb growth occurs earlier than lower limb growth, and the foot grows earlier than the rest of the lower limb. In childhood, the trunk grows most rapidly; in adolescence, the lower limbs grow the fastest. Throughout growth, body proportions gradually assume adult form [1].

Growth is greatest in early infancy, declines during childhood, and briefly increases again during the adolescent growth spurt. A child is about half his or her adult height at 2 years of age and about three-fourths by 9 years of age [2].

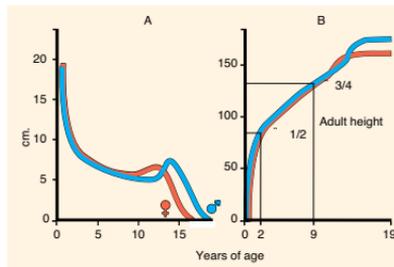
Growth rates from various epiphyses varies. In the upper limb, growth is most rapid at the shoulder and wrist in contrast to the lower limb where most growth occurs just above and below the knee [1 opposite page].



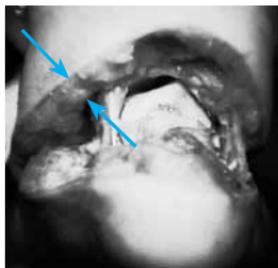
3 Growth variations. These individuals show the wide variations in growth. Courtesy of Dr Judy Hall.



1 Changes in body proportions with growth. At maturity, the position of the center of gravity (green line) is the level of the sacrum. From Palmer (1944).



2 Growth Rate. A. Growth rates for girls (red) and boys (blue) by age. The greatest rate of growth occurs during infancy. B. Growth rate as a fraction of adult height. About half of an individual's adult height is reached by age 2 years and three-fourths by age 9 years.

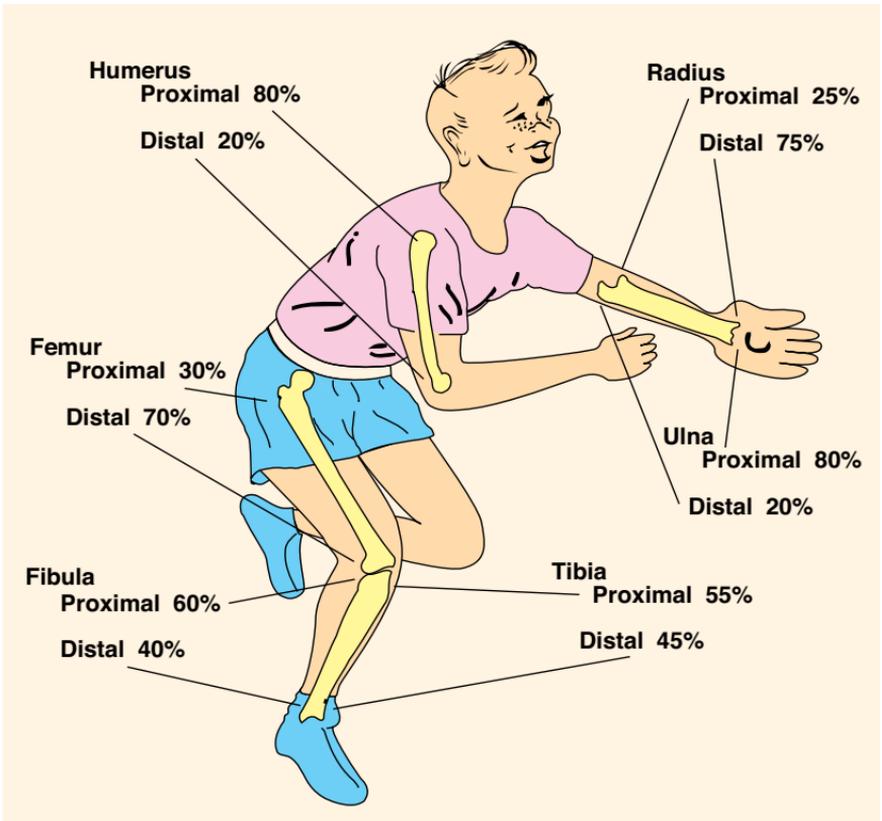


4 Subcutaneous fat in infancy. Note the thickness in the subcutaneous fat (arrows) in this infant undergoing clubfoot correction.

The growth rate of tissues varies with age. Subcutaneous fat, which provides nutritional reserve and protection from cold and injury, develops during the first year. The fat also obscures the longitudinal arch of the foot, giving the infant a flatfooted appearance [4 opposite page]. The percentage of muscle increases with age, but the percentage of neural tissue declines with advancing age.

Growth control factors are systemic and local.

- *Systemic factors* play a key role. Endocrine, nutritional, and metabolic disorders significantly alter growth [3 opposite page].
- *Local factors* may retard or accelerate growth [1 next page]. Procedures known to accelerate growth have been used in an attempt to lengthen the short limb due to poliomyelitis. Unfortunately, the gain in length is not predictable and is not enough to be clinically useful.



1 Epiphyseal contribution of long bone growth. From Blount (1955).

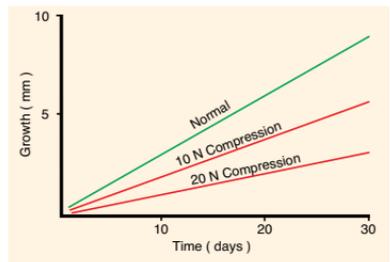
14 Growth / Infancy

Compression of the physis retards growth in proportion to the load applied [2]. This has been studied in rats. Forelimb amputations result in upright walking. This bipedal walking causes significant anterior wedging of the lower lumbar vertebrae, presumably due to the greater loads applied to the anterior portion of the vertebral bodies.

Growth control factors are inherent in each growth plate. When juvenile limbs are transplanted onto adult rats, they continue to grow.

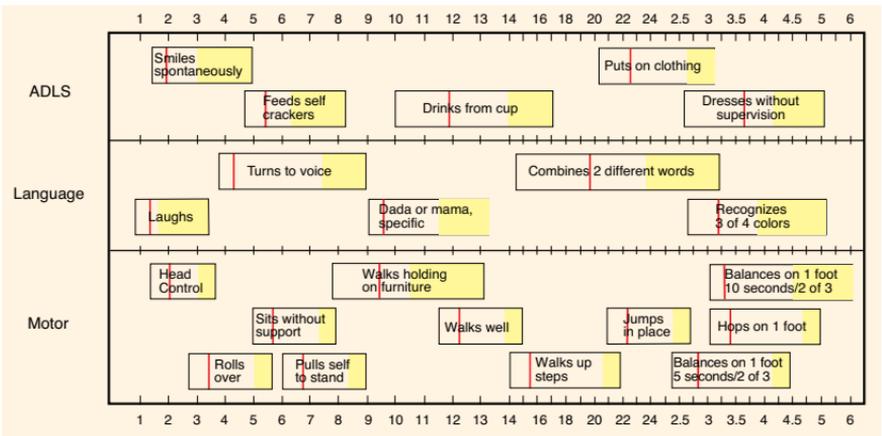
Gross motor development The standard for assessing motor development is the age of acquisition of gross motor skills. Such skills are easily measured and useful in assessing development [3]. Infants usually show head control by about 3 months, sit by 6 months, stand with support by 12 months, and walk unsupported by 15 months. These general guidelines are useful when screening.

Local Factors	Effects Growth
Physcal compression	Retards
Denervation	Retards
Physcal ischemic injury	Retards
Sympathectomy	Accelerates
AV fistula	Accelerates
Periosteal division	Accelerates
Periosteal stripping	Accelerates
Diaphyseal fracture	Accelerates
Foreign body reaction	Accelerates
Chronic osteomyelitis	Accelerates



1 Local factors affecting growth.

2 Physcal compression effect on growth. Growth rate is reduced by compression (N = Newtons). From Bonnell (1983).



3 Denver developmental screening test. From Frankenberg (1967).

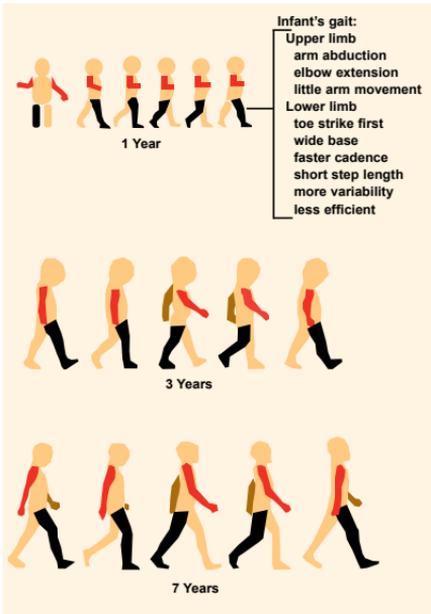
Childhood

Childhood extends from the middle of the second year until adolescence. During this time, growth and development continue but at a slower rate than in infancy. Because childhood lasts so long, the majority of growth and development occurs during this period.

Gait during infancy is less stable and efficient than that of the child or adult [1]. Early gait is characterized by a wide-base irregular cadence, instability, and poor energy efficiency. The instability of gait in the infant is due to a high center of gravity, low muscle to body weight ratio, and immaturity of the nervous system and posture control mechanisms.

Developmental variations occur during infancy and childhood [2]. These variations are commonly mistaken for deformities. They include flatfeet, in-toeing, out-toeing, bowlegs, and knock-knees. These conditions resolve with time and seldom require any treatment. These conditions are covered in more detail in Chapters 4 and 5.

Prediction of adult height is valuable in managing certain deformities, particularly anisomelia (limb length inequality). A variety of methods for predicting adult height are available. A simple method involves establishing the percentile of height by plotting the child's height on the growth chart by bone age rather than by chronologic age. This percentile is projected out to skeletal maturity to provide an estimate of adult height [1 next page].



1 Development of normal gait. Adult gait pattern is achieved by about 7 years of age in the normal child. From Sutherland (1980) children.



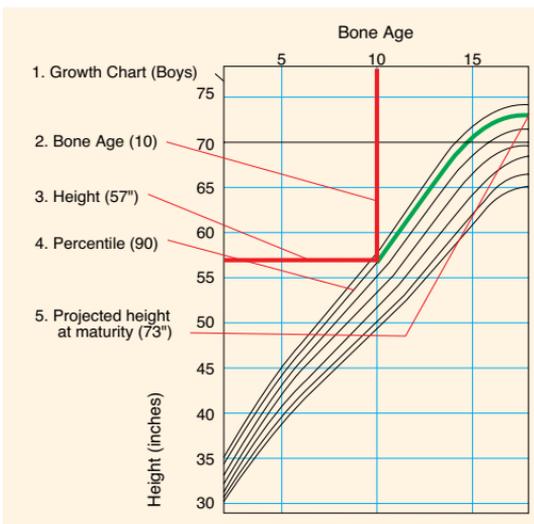
2 Developmental variation in normal children. Common variations include knock-knees (left), flatfeet (top right), and femoral torsion (lower right).

16 Growth / Adolescence

Adolescence

Adolescence extends from the beginning of puberty until skeletal maturity. Certain diseases, such as scoliosis and slipped capital femoral epiphysis, develop during this time.

During adolescence, psychosocial factors receive a higher priority than in childhood. Physical appearance becomes increasingly important. Preexisting deformities or disabilities that may have caused little concern during childhood suddenly produce great distress. A boy with a small calf associated with a clubfoot deformity will request exercises to build up the limb size. A girl will become aware of old operative scars on her knee that before were ignored. A girl with an abductor lurch, present since infancy, may become concerned about it for the first time at age 13 years.



1 Prediction of adult height.

Predict adult height by plotting the child's bone age (vertical red line) against the current height (horizontal red line) to determine the percentile value (green). Follow the percentile (green line) to skeletal maturation to estimate final adult height.



2 Obesity and orthopedic problems. Two common serious orthopedic problems, slipping of the capital femoral epiphysis (black arrow) and tibia vara (yellow arrow), are commonly associated with obesity.

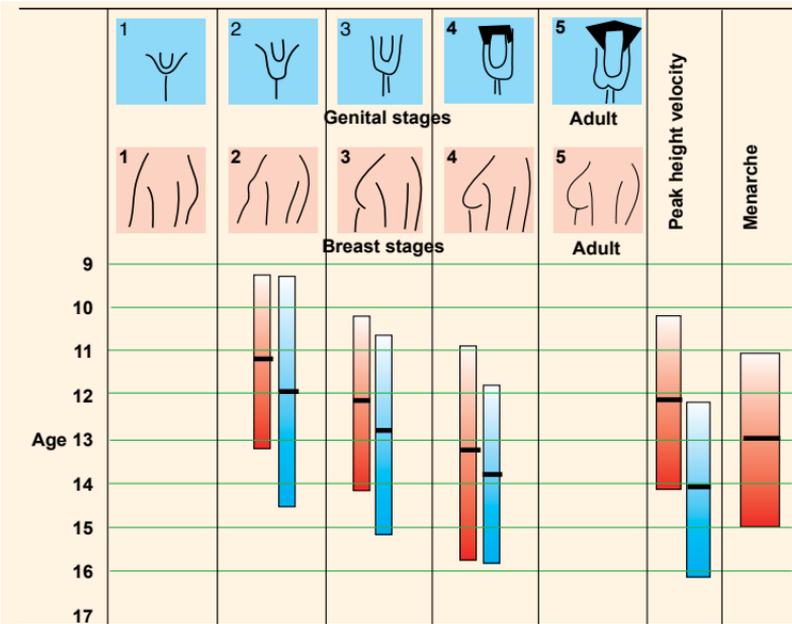


3 Leg length inequality. Bone age determination is helpful in planning correction by epiphysiodesis.

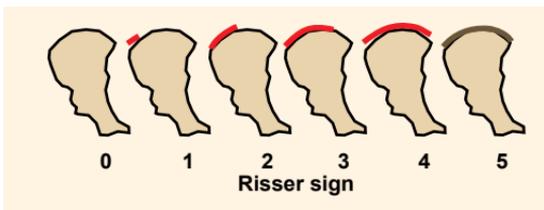
Obesity Obesity in children is becoming more common. The added weight is a factor in the development of several orthopedic problems. These include slipped capital femoral epiphysis and tibia vara [2 opposite page].

Determining maturation level Knowing the amount of growth remaining is important to the timing of physcal fusion and thus in correcting leg length inequality [3 opposite page] and in managing patients with scoliosis.

- **Hand-wrist radiographs** Use the Greulich-Pyle atlas to estimate the bone age.
- **Tanner stages** The level of maturation is based on the physical examination. Because this assessment requires an assessment of breast and genital development [1] in a sensitive age group, its use is limited.
- **Risser sign** is based on the extent of ossification of the iliac crest as assessed on the AP radiograph [2]. This sign has been commonly used in assessing maturity when managing scoliosis.
- **Other signs** such as the velocity of height gain and the status of the tri-radiate cartilage (acetabulum) are becoming useful maturational indices.



1 Tanner maturation index. Using physical signs, the level of maturation is assessed for males (blue) and females (red). The columns show the 3–97% levels. Mean values are shown by the black bars.



2 Risser sign. The extent of ossification of the iliac apophysis is commonly used to assess the skeletal maturation of patients with scoliosis. Risser 0 = no iliac apophysis; Risser 5 = fusion of the apophysis with the ilium.

Abnormal Growth

Disorders affecting the musculoskeletal system are relative common [1]. These and other conditions that cause limitation of activity in children have tripled during the past four decades because children with disabilities are more likely to survive today than in the past.

Congenital Defects

Multifactorial inheritance is the most common cause of congenital defects [2]. Of newborn infants, 3% show major defects and an additional 3% are discovered later during infancy. About 20% of perinatal deaths are attributable to congenital problems. Single minor defects are present in many newborns. Because infants with multiple minor defects have a higher incidence of major malformations, the finding of minor defects should prompt a careful search for more serious problems. Musculoskeletal problems account for about one-third of congenital defects. Hip dysplasia and clubfeet make up half of the primary musculoskeletal defects.

Although inherited disorders may manifest themselves during infancy, the majority of musculoskeletal problems of infancy are due to environmental factors, such as malnutrition, infection, and trauma.

Chromosomal Abnormalities

Chromosomes have been mapped to show the location of defective genes that create disorders often seen in orthopedic clinics [1 opposite page]. The linkage of genes causing diseases with genes controlling distinguishable characteristics makes possible the identification of individuals at risk for certain diseases. For example, on chromosome 9, the gene carrying nail-patella syndrome is linked to the gene of ABO blood type. Offspring with the same ABO blood type as an affected parent will carry the syndrome.

Many chromosomal abnormalities are due to changes in number, structure, or content of chromosomes. Numerical changes in chromosomes are due to a failure of separation or nondisjunction during cell division. Nondisjunction results in monosomy or trisomy gametes. Monosomy of sex chromosomes produces the XO pattern of Turner's syndrome.

Disease	Prevalence estimated per 1000
Cerebral palsy	2.5
Trisomy 21	1.1
Developmental hip dysplasia	1.0
Clubfoot	1.0
Sickle cell disease	0.46
Muscular dystrophy	0.06

1 Prevalence of orthopedic disorders.

Cause	Percentage
Chromosomal aberrations	6
Environmental factors	7
Monogenic or single gene	8
Multifactorial inheritance	25
Unknown	4

2 Causes of congenital defects. From Moore (1988).

Trisomy of sex chromosomes causes 47XXX females who may have only mild mental retardation, whereas 47XXY causes Klinefelter's syndrome and 47XYY causes a disorder characterized by aggressive behavior. Trisomy of autosomes (nonsex chromosomes) is common and frequently affects chromosome 21, which causes Down syndrome [2]. Trisomy 13 and 18 cause significant defects but are less common.

Chromosomal structural defects occur spontaneously or secondarily to the effects of teratogens [3]. Teratogens are agents that induce defects and cause a variety of syndromes. Deletions of portions of chromosomes 4, 5, 18, and 21 produce specific syndromes. For example, deletion of the terminal portion of the short end of chromosome 5 causes the "cri du chat" syndrome. Other common changes include translocations, duplications, and inversions.

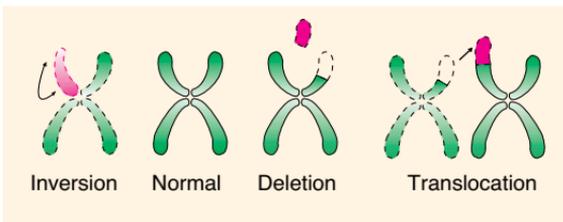
Single gene defects may be inherited or produced by spontaneous mutation. Once established, the defect is inherited according to Mendelian laws. Thus, the individual's genetic makeup is largely determined by a random process during meiosis and fertilization.

Chrom.	Disorder
1	Rh blood group, Gaucher's, CTM diseases
5	MPS VI, cri du chat syndrome
6	Histocompatibility complex
7	MPS VII, Ehlers–Danlos VII, some Marfan's
9	ABO typing, nail–patella syndrome
15	Prader–Willi syndrome
X	Duchenne dystrophy, chondrodysplasia

1 Chromosome disorder location. Localization of musculoskeletal disorders to specific chromosomes.



2 Down syndrome hip instability. Due to the excessive joint laxity, recurrent dislocations (arrow) may occur in these children.



3 Chromosome structural defects. Various structural defects include inversions, deletions, and translocations.

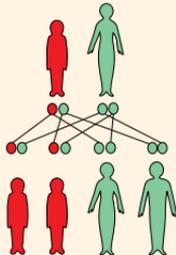
Inherited Disorders

Fertilization restores the diploid number of chromosomes and composites the traits of both parents. Fertilization may produce an abnormal zygote if the ovum or sperm carries defective genes. These conditions are transmitted by several mechanisms.

Dominant inheritance results in a disorder caused by a single abnormal gene [1]. Autosomal dominant conditions usually produce structural abnormalities [2 and 3 opposite page]. Variable expressivity and incomplete penetrance suppress or minimize the expression of dominant inheritance.

Recessive inheritance is expressed only if both gene pairs are affected [2]. Metabolic or enzymatic defects that cause diseases such as the mucopolysaccharidoses are often inherited by autosomal recessive inheritance.

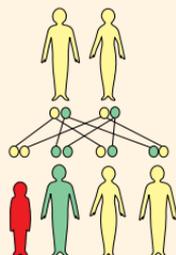
X-linked inheritance involves only the X chromosome [3]. In the male, the genetic inactivity of the Y chromosome allows even the recessive abnormal gene of the X chromosome to be manifested. A classic example of X-linked recessive inheritance is pseudohypertrophic muscular dystrophy. The female is the carrier, but only male offspring are affected. In recessive X-linked inheritance, the female is affected only in the rare situation in which both genes of the genetic pair are abnormal.



A pedigree chart illustrating dominant inheritance. The first generation consists of one affected individual (red) and one unaffected individual (green). They have four children: two affected (red) and two unaffected (green). The second generation consists of four affected individuals (red) and four unaffected individuals (green).

- Achondroplasia
- Brachydactyly
- Cleidocranial dysostosis
- Marfan syndrome
- Multiple epiphyseal dysplasia
- Nail-patella syndrome
- Neurofibromatosis
- Polydactyly

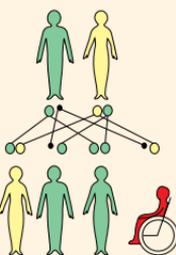
1 Dominant inheritance. The dominant gene (red) causes structural defects in both parent and offspring. Musculoskeletal disorders transmitted by dominant inheritance are listed.



A pedigree chart illustrating recessive inheritance. The first generation consists of two unaffected individuals (yellow). They have four children: two carriers (yellow) and two affected individuals (red). The second generation consists of one affected individual (red) and three unaffected individuals (yellow).

- Congenital insensitivity to pain
- Diastrorphic dwarfism
- Gaucher disease
- Hurler syndrome
- Morquio syndrome
- Scheie's syndrome
- Hypophosphatasia

2 Recessive inheritance. Carriers of the recessive genes (yellow) are expressed (red) only if both gene pairs are abnormal. Musculo-skeletal disorders transmitted by recessive inheritance are listed.

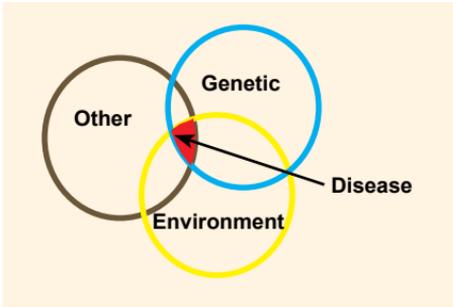


A pedigree chart illustrating X-linked inheritance. The first generation consists of one unaffected individual (yellow) and one carrier (yellow). They have four children: one affected individual (red) and three unaffected individuals (yellow). The second generation consists of one affected individual (red) and three unaffected individuals (yellow).

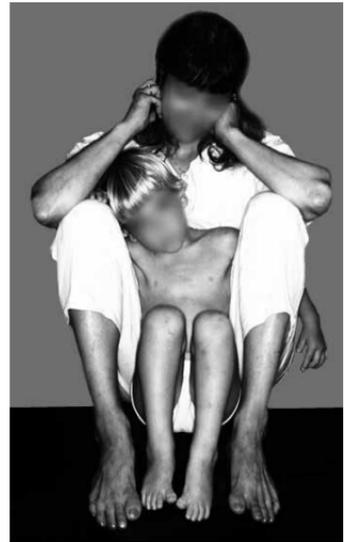
- X-linked dominant**
- Vitamin D refractory rickets
- X-linked recessive**
- Hemophilia
- Pseudohypertrophic muscular dystrophy

3 X-linked inheritance. X-linked defects (yellow) are carried by the female and expressed in the female if the gene is dominant. Most defects are recessive and are expressed only in the male (red).

Polygenic inheritance (or multifactorial inheritance) involves multiple genes and an environmental "trigger" [1]. Such common conditions as hip dysplasia [4] and clubfeet [5] are transmitted by this mechanism.



1 Polygenic inheritance. Many common orthopedic problems are transmitted by this mode. Genetic, environmental, and possibly other factors combine to cause the problems.



2 Familial toe deformities. The mother and child have the same toe abnormalities. Toe and finger deformities are often familial.



3 Toe deformities. These toe deformities are exactly the same in the mother and child.



4 Hip dysplasia. Developmental hip dysplasia is a common condition with a multifactorial etiology.



5 Clubfoot in utero. High-resolution ultrasound shows a clubfoot deformity. Clubfeet are common deformities with a multifactorial etiology.

22 Growth / Abnormal Morphogenesis

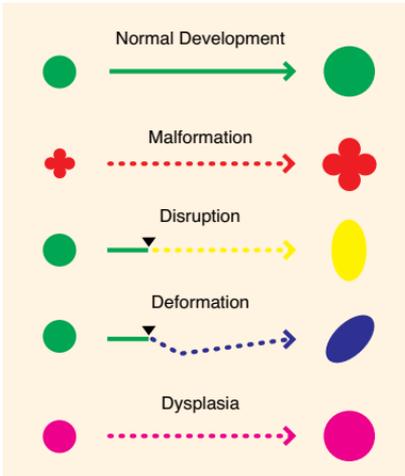
Abnormal Morphogenesis

Abnormal morphogenesis is classified into four categories [1].

Malformations are defects that arise in the period of organogenesis and are of teratogenic or genetic origin. Phocomelia and congenital hypoplasia [3] are examples.

Dysplasias result from altered growth that occurs before and after birth [2].

Disruptions occur later in gestation when teratogenic, traumatic, or other physical assaults to the fetus interfere with growth. Ring constriction due to amniotic banding [2 and 3 opposite page] are examples.



1 Classification of abnormal morphogenesis. These categories provide a practical basis for understanding congenital defects. From Dunne (1986).



2 Achondroplasia. Achondroplasia is one of many osteochondral dysplasias commonly seen in orthopedic clinics.



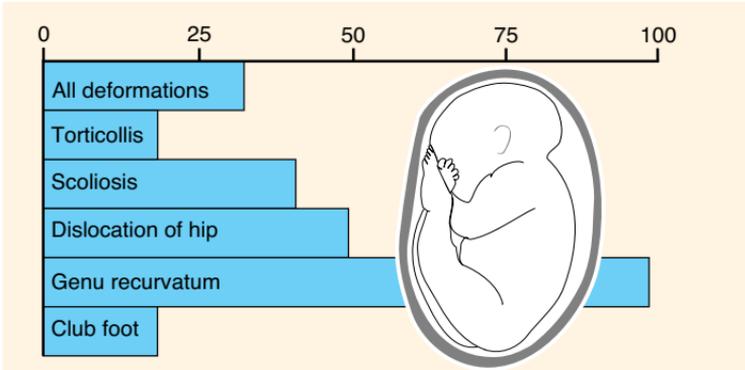
3 Limb hypoplasia. Major limb defects are malformations arising from interruption of limb development.

Deformations occur at the end of gestation and are due to intrauterine crowding [1 and 4]. These deformities are milder and usually resolve spontaneously during early infancy.

Developmental Deformities

Metabolic disorders such as rickets cause osteopenia and a gradual bowing of long bones.

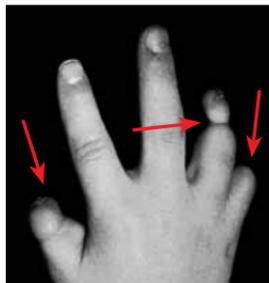
Inflammatory disorders may damage the growth plate or articular cartilage, causing shortening or angular deformity. Less commonly, chronic inflammation that does not affect the growth plate from conditions such as rheumatoid arthritis or chronic osteomyelitis may induce hyperemia and accelerate bone growth, thus causing bone lengthening.



1 Breech position. Common musculoskeletal defects associated with breech position. From Clarren (1977).



2 Congenital constriction bands. Intrauterine adhesion caused this deep circumferential band.



3 Constriction bands causing hand deformity. Amputation of the thumb and little finger and hypoplasia of the ring finger result from bands.



4 Molding deformity. Intrauterine crowding caused this calcaneovalgus foot deformity.

24 Growth / Iatrogenic Deformities

Physical activity may alter bone growth. For example, long-term non-weight-bearing activity, as was once prescribed in treating Perthes disease, resulted in slight shortening of the involved leg. Similarly, professional tennis players who start their careers as children show relative overgrowth of the dominant upper limb.

Neuromuscular deformity may occur from muscle imbalance such as in the child with spasticity from cerebral palsy. Adductor spasm positions the head of the femur on the lateral acetabular rim causing deformity and erosion of the cartilage of the labrum, which in turn causes subluxation and eventual dislocation of the hip [1]. The combination of contractures, immobility, gravity, and time create the so-called windswept deformity common in spastic quadruplegia.

Trauma may cause deformity by malunion or growth plate damage [2]. If the growth plates are not damaged, growth contributes to the correction of residual malunion deformity through the process of *remodeling*.

Idiopathic disorders Sometimes the cause of the developmental deformity is not determined [1 opposite page].

Iatrogenic Deformities

The cradleboard, by positioning the infant's hip in extension, is a known cause of developmental hip dysplasia [2 opposite page]. In some cultures, iatrogenic deformities are created in girls to enhance their beauty. Placing rings around the neck [3 opposite page] of young girls and binding of the feet [4 opposite page] has produced deformity and severe disability.



1 Hip deformity in cerebral palsy. This boy with cerebral palsy (left) developed an adduction deformity (red arrows) and a secondary dislocation (yellow arrow) of the right hip.



2 Growth arrest lines. This post-traumatic physeal bridge (black arrow) caused asymmetrical growth of the distal tibia, as shown by the growth arrest line (yellow arrows).



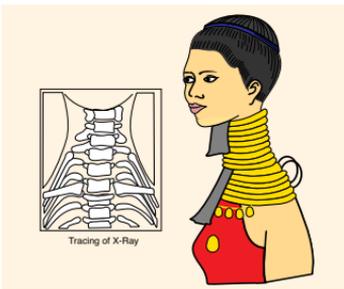
1 Idiopathic growth

acceleration. This girl pictured in the 1940s has massive overgrowth of the left upper extremity producing a grotesque disability. The girl died during the operation to remove the extremity.



2 Cradleboard.

Cradleboards extend the infant's hips, causing an increased incidence of hip dysplasia.



3 Thoracic deformity.

Rings placed around the neck in childhood produce constriction of the upper thorax in the adult woman (Padaung tribe, east Burma). From Roaff (1961).



4 Bound feet. A woman's feet show the effect of foot binding during childhood. The foot becomes triangular in shape (left and middle) and small in size so that it fits the shoe (right). The shoe is less than 6 inches in length.

Chapter 2 – Evaluation

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Evaluation leading to an accurate diagnosis [1] is the first and most important step in optimal management. Every condition requires a diagnosis, but only some require active treatment. The evaluation of the child is often more difficult than that of the adult. The child is a poor historian, and examination of the child can be difficult. Dealing with the family may be challenging. The history given by the parents is often laced with emotion. Reporting is often complicated by varying gender and generational hierarchy. The physician often finds that managing the child's problem is easier than dealing with the family. Establishing rapport during the first visit is essential.

Establishing Rapport

The goal is to reduce the fear in the child and establish confidence with the parents and family.

Dress

Studies have shown that casual dress promotes approachability and more formal dress enhances confidence. Dress in a way that suggests you have good judgment and are more appropriate for the situation. More formal dress may be more appropriate in a major referral center than elsewhere. Avoid making a statement by dress. This usually translates into selecting conservative clothing that promotes an image of good taste.



1 Diagnosis. Evaluation requires integration of clinical, imaging, and laboratory findings.

Initial Introduction

On entering the examination room, acknowledge everyone in the room [1]. Consider the cultural background of the family and conform to gender order for introductions. Shake hands with everyone including the child. Determine the relationship of each person with the patient.

Be professional yet friendly. Establishing a good rapport with *everyone* in the family may be critical to properly managing the child. Later, when difficult management decisions must be made, having rapport with every member of the family is necessary to avoid pressure on the parents to seek additional opinions. Once started, serial consultations usually end with some unnecessary treatment of the child.

Calming the Child

Reducing the child's fear is the next objective. Consider examining the infant or younger child on the parent's lap [2]. Ask the child on whose lap he or she wishes to sit. Children will often select the family member who they believe will offer the greatest safety.

Be friendly with the child. Suggest that this will be a game. Make some positive statements about the child, such as "Mary, you are such a nice child." Ask some child-oriented questions, such as "What is your pet's name?"

Start gently examining the child while taking the history from the family. This first step is to convince the child that the examination will not be painful. This is the time for the screening examination, starting with the area most removed from the problem. Being gentle often results in the child becoming less threatened and more cooperative.

Sometimes, these measures fail and the infant or young child remains aggravated and uncooperative. This is the time to move to strategy two—a firm approach [1 opposite page].

Tips for the Physician

1. Knock on the door before entering to give anyone undressed a chance to cover-up before you go in.
2. Touch the patient either with a handshake or with a pat on the shoulder.
3. Introduce yourself and your colleagues to everyone in the examining room. Attempt to identify the cultural expectations to establish the order of introductions. Shake everyone's hand.
4. Establish the reason for the clinic visit.
5. Sit down in the room, preferably lower than the patient.
6. Show the family the x-ray, especially if it is normal.
7. Avoid technical terms.
8. Avoid leaving the room during consultation unless definitely necessary. Avoid looking at your watch.
9. Do not discuss other patients' treatment.
10. Avoid trying to impress patient with your credentials; the family has already selected you as their physician.
11. Discuss the problem, options, and recommendations.
12. Try to assess the family's reaction to the discussion. Continue discussion until the family's expectations are fulfilled.
13. Offer to provide follow-up if the family appears to need continued reassurance.

1 Suggestions for establishing rapport.



2 Efficient, comfortable examination. Positioned on the parent's lap, the infant or child is most secure and quiet.

History

The child's complaints usually fall into the categories of deformity, altered function, or pain. Assessment of these complaints should take the patient's age into consideration. For example, the toddler usually manifests discitis (an intervertebral disc space infection) by altered function in the form of an unwillingness to walk. The child with discitis may primarily show a systemic illness, whereas the adolescent often complains of back pain.

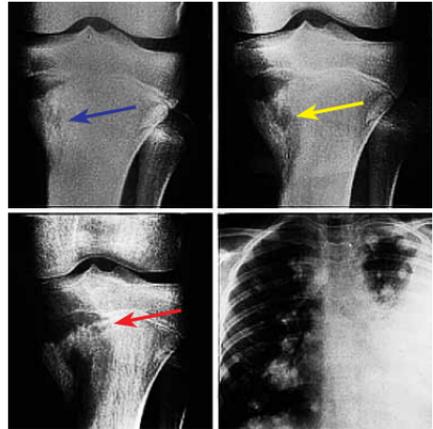
A common pitfall in diagnosis is inappropriately attributing the child's problem to trauma. Although trauma is a common event in the life of a child, serious problems such as malignant tumors or infections may be mistakenly attributed to an injury [2].

Deformity

Positional deformities such as rotational problems, flatfeet, and bowlegs are common concerns but seldom significant [3]. More significant problems, such as congenital or neuromuscular deformities, require careful evaluation. Inquire about the onset, progression, and previous management. Are there old photographs or radiographs that document the course of the deformity? Is there associated pain or disability? Does the deformity cause a cosmetic problem and embarrass the child? Is it noticeable to others? Finally, be cautious about relying solely on the family's estimation of the time of the deformity's onset. Often a deformity originates long before it is first noticed.



1 When coaxing fails. Perform the examination without the cooperation of the child.



2 Confusing trauma history. A 12-year-old boy gives a history of knee trauma and pain. The initial radiograph was considered normal, but a lesion is present (blue arrow). One month later, the lesion has enlarged (yellow arrow). A diagnosis of Osgood–Schlatter disease was made. A radiograph 2 months later showed further expansion of the lesion (red arrow). A radiograph of the chest just prior to death showed multiple pulmonary metastases from osteogenic sarcoma. Attributing this problem to “trauma” was disastrous.



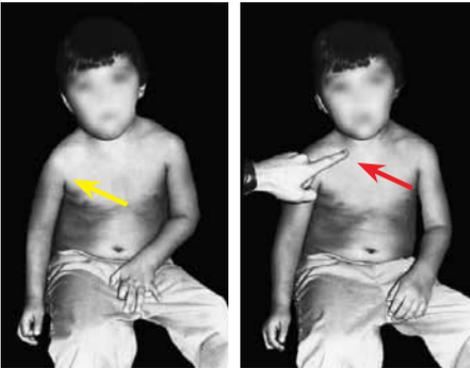
3 Familial flatfeet. Since the father has flatfeet, it is more likely that the child's flatfeet will persist into adult life.

Altered Function

Function can be altered by deformity, weakness, or pain. Pain is a common cause of altered function in the infant and child; the most common example is a limp. A toddler's fracture of the tibia may be manifested by a limp or an unwillingness to walk. The young child with toxic synovitis may simply limp; the older child might complain of pain. The newborn whose clavicle is fractured during delivery shows a loss of arm movement on the affected side. This may be confused with a birth palsy. Altered function due to trauma, inflammation, or infection without neurologic damage is referred to as *pseudoparalysis*.

Pain

The expression of pain is age related. The infant may simply avoid moving the painful part, may fuss and cry, or cry continuously if the pain is severe. The child may show altered function, avoid moving the affected part, or complain of discomfort [1]. The adolescent usually complains of pain.



1 Pseudoparalysis. Use of the arm (yellow arrow) is restricted because of pain. A painful lesion of the right clavicle (red arrow) was due to a leukemic infiltrate.



2 Importance of medical history. This boy had normal function of his right arm (arrow) as an infant. During early childhood, he developed weakness of the arm (arrow), and a diagnosis of cerebral palsy was made. The weakness increased, and finally during adolescence, he was found to have a tumor involving the cervical spinal cord (arrow). He became quadriplegic. The progressive nature of the condition is inconsistent with a diagnosis of cerebral palsy. A medical history of progression would have prompted an earlier diagnosis and may have prevented this disastrous outcome.

The perception and expression of pain differs widely among individuals, particularly as adolescents grow more adult-like in their responses. A young athlete might minimize his discomfort to improve his chances of participating in the next sporting event. Others might exaggerate the problem. Some adolescents minimize pain by pain-relieving positioning. A herniated disc or an osteoid osteoma may cause scoliosis. This scoliosis results from positioning the spine in a pain-relieving posture. This secondary deformity rather than the underlying condition may be the focus of the evaluation. Unless the underlying condition is identified by the physician, a serious diagnostic error can occur.

Past History

The past history is essential, not only for understanding the background and general health of the child but also for gaining insight into the current problem. Important aspects of past history include the following:

Birth history Were the pregnancy and delivery normal?

Development Have the developmental milestones been met at the appropriate age? When did the infant first sit and walk? About one-third of late walkers are pathologic. In children with conditions such as cerebral palsy, walking is always delayed and may be important in establishing whether or not the condition is progressive [2 opposite page].

Mother's intuition The mother's intuition is surprisingly accurate [1]. For example, the mother's sense that something is wrong with her infant is one of the most consistent findings in infants with cerebral palsy. Take the mother's concerns seriously.

Family history Do others have problems similar to those of the patient? If so, what disability is present? A surprisingly large number of orthopedic problems run in families, and knowledge of the disability, or absence of disability, provides information regarding the patient's prognosis.



1 Mother's intuition. The mother with painful degenerative arthritis from developmental hip dysplasia (red arrow) sensed something was abnormal about her infant's hip. Her concern, based on intuition, was discounted by the primary physician, and the asymmetry present on examination was attributed to the child's mild hemiparesis. This resulted in a delay in diagnosis of developmental hip dysplasia until 18 months of age (yellow arrow).

Physical Examination

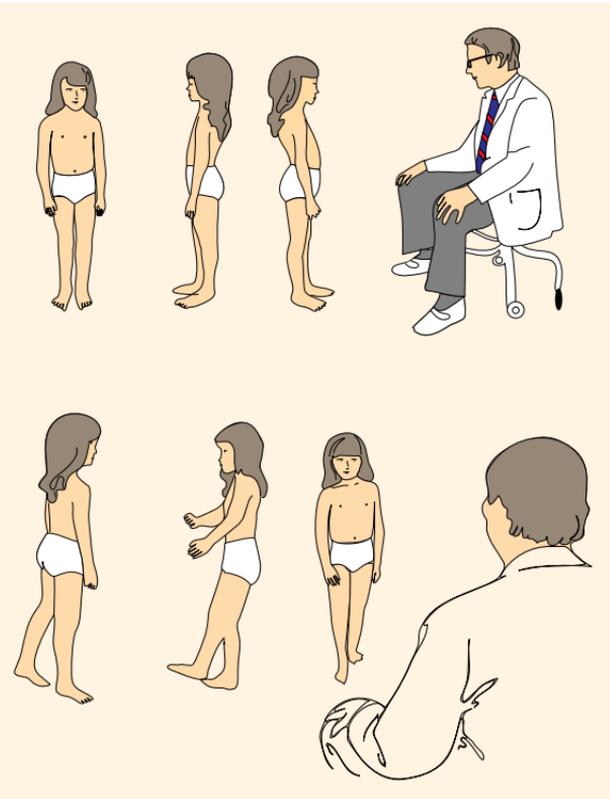
Examination of the musculoskeletal system should include two steps: (1) a screening examination and (2) a complete musculoskeletal evaluation performed to assess a specific complaint. The history and physical examination provide the diagnosis in most cases. It should be thorough and carefully performed. With the proper approach, it is usually possible to perform an adequate examination even without the cooperation of the infant or child.

Approach

Approach the child in a friendly and gentle fashion. Examining the child on the mother's lap is helpful. If the child is still nervous, keep your distance while obtaining the history. Reassure the child that all you plan to do is to watch her walk or move her legs. If the child is still nervous, examine the parent or sibling first. The child may find it reassuring for you to go through the examination with the parent first. If the child will not cooperate in walking, carry her to the opposite side of the room. The child will usually walk or run back to the parents. If the child has pain, always examine the painful site last.

Screening Evaluation

Examine the child in his underclothing. Examine the adolescent in a gown or, even better, in a swimsuit. Perform the screening examination [1] first before focusing on the principal complaint. This screening ensures that you do not miss any other orthopedic problems and will provide a general



1 Inspect from front, side, and back. Observe the child walking normally, then on heels and toes.

overview of the musculoskeletal system necessary to understand the specific problem. It is essential to see the whole child to avoid missing important clues in diagnosis, such as the midline skin dimple that may accompany an underlying spinal deformity [1]. For example, knowledge of the degree of generalized joint laxity is valuable in assessing a flatfoot or a dysplastic hip. The examination of the back is an essential part of an evaluation of foot deformities. A cavus foot deformity is a common feature of diastematomyelia.

Infant screening Examine the infant on the mother's lap. First, observe the general body configuration. Next, observe the infant's spontaneous movement patterns for evidence of paralysis or pseudoparalysis [2]. Any reduction of spontaneous movements is an important finding. For example, the only consistent physical finding of the neonate with septic arthritis of the hip is a reduction in spontaneous movement of the affected limb. Finally, systematically examine the limbs and back for joint motion and deformity. Always perform a screening hip examination to rule out developmental hip dysplasia.

Examining the child and adolescent The examination requires several steps:

- **General inspection** Does the child look sick [3]? With the child standing in the anatomic position, observe her from the front, side, and back. Look at body configuration, symmetry, and proportions and for specific deformities.



1 Sacral dimple. A midline skin lesion such as a sacral dimple suggests the presence of a congenital spinal dysraphism.



2 Importance of observation. This infant shows reduced spontaneous movement of the left leg and an abducted position of the left hip. The infant has septic arthritis of the left hip.



3 Ill child.

34 Evaluation / Physical Examination

- **Pelvis and back** Place your hands on the iliac crests—are they level? A pelvic tilt usually results from a limb length difference. Next, ask the child to raise one leg at a time. A drop in the pelvis on the opposite side indicates a weakness of the hip abductors found in conditions such as hip dysplasia and cerebral palsy. With the child facing you, assess thoracic and lumbar symmetry for evidence of scoliosis by the forward-bending test. Observe the sagittal alignment of the spine [1].
- **Assessing gait** Ask the child to walk slowly across the room and back first with normal gait and then repeated on her toes and heels. Observe the gait for evidence of asymmetry, irregularity, or weakness. Any abnormal or questionable findings discovered during the screening examination should prompt a more complete evaluation of the problem. For example, a finding of in-toeing should prompt an assessment of the rotational profile.



1 Sagittal alignment. Note the increased lordosis (red arrow) and dorsal kyphosis (blue arrow).



2 Familial joint laxity. Note hyperextension of the knee in both the child and father.

Specific Evaluations

The history and findings of the screening examination serve as guides to more in-depth evaluation.

Joint laxity Joint mobility is greatest in infancy and gradually declines throughout life. Joint laxity, like other traits, varies widely among individuals and is usually genetically determined [2 opposite page]. Extremes in joint laxity are seen in certain disorders, such as Ehlers–Danlos and Marfan syndromes.

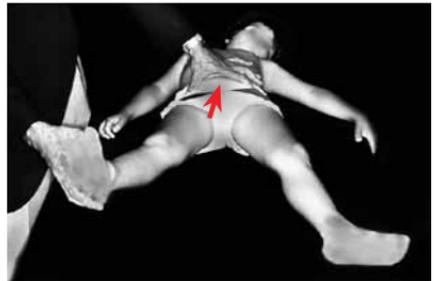
Assess joint laxity by testing the mobility of the ankles, knees, elbows, thumbs, and fingers [1]. Excessive laxity in four or all of the five joints tested occurs in about 7% of children. Joint laxity is a contributing factor in the pathogenesis of hip dysplasia, dislocating patellae, and flatfeet, and it increases the risk of injuries such as sprains. In general, excessive joint laxity suggests the possibility of other problems.

Range of motion (ROM) The normal values of joint motion change with age. Generally, the arc of motion is greatest in infancy and declines with age. Specific joints are affected by intrauterine position. For example, lateral hip rotation is greatest in early infancy and declines during the first 2 or 3 years of growth. In assessing ROM, a knowledge of normal values is helpful. Make certain that the position of the pelvis is determined by palpation when assessing hip abduction [2].

- *Contractures* of diarthrodial muscles are common in children and sometimes require lengthening. For example, contracture of the gastrocnemius and gracilis occur in cerebral palsy. By proper positioning of the joints above and below the contracture, it is possible to differentiate contractures of these muscles from adjacent elements of the same muscle group.
- *Hip flexion motion* is difficult to measure due to compensatory motion of the lumbar spine. Measurements can be made by the Thomas or prone extension tests. The prone extension test has been found to be more reliable. Most ROM measurements of most joints are reproducible within about $\pm 4^\circ$.



1 Finger tests for joint laxity. The ability to approximate the thumb and the forearm and extend the fingers to a parallel relationship with the forearm suggests an excessive degree of joint laxity.



2 Assessing hip abduction. Stabilize the pelvis with one hand (arrow) and abduct the hip with the other. Assess abduction using the anterior iliac spines as points of reference.

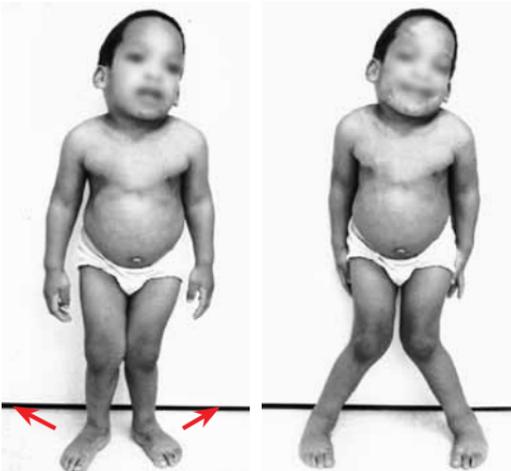
Deformity

Deformity is classified as either functional or structural. Functional deformity is secondary to muscle contracture or spasm-producing fixation of a joint in an abnormal position. For example, a fixed hip adductor contracture elevates the pelvis on the affected side, producing a functional shortening of the limb. This deformity is commonly seen in cerebral palsy and Perthes disease. In contrast, structural deformity originates within the limb. An example is the limb shortening associated with fibular hemimelia.

Assess deformity in reference to body planes with the body in the anatomic position [1]. Frontal or coronal plane deformity is most easily observed and creates the most significant cosmetic disability. Sagittal plane deformity produces problems in the plane of motion. Finally, transverse or horizontal plane deformity is most difficult to visualize and was often overlooked in the past. Currently, CT and MRI studies allow visualization and documentation of this plane and increased the appreciation of transverse plane problems. In assessing and documenting deformity, it is essential that each plane be separated clearly and described independently [2]). For example, in tibia vara, deformity occurs in both the frontal and transverse planes. Failure to clearly separate these planes may result in serious errors if operative correction is undertaken.

Altered Function

Function may be impaired by many mechanisms. The impairment is most obvious when the onset is acute and recent. The parents are aware when the pseudoparalysis is due to their child's "pulled" or "nursemaid's" elbow



1 Differentiate transverse and frontal plane deformity. This child compensates a severe genu valgum deformity by walking with the feet laterally rotated (red arrows). When the legs are placed in the anatomic position, the valgus deformity of the knees becomes apparent.



2 Cubitus varus deformity. This deformity is secondary to a malunited fracture. The child is unaware of any problem.

[1]. Conversely, long-standing changes in function may be overlooked or just considered as an unusual characteristic of the child. A child's bilateral abductor lurch from dislocated hips may go unappreciated for years. Limping of recent origin is usually obvious to the parents. Sometimes the examination is normal, and imaging studies are necessary to establish the diagnosis [2].

Evaluate altered function of recent onset for evidence of trauma or infection. Look for deformity, swelling, or discoloration. Palpate to determine if tenderness is present. Finally, evaluate joint motion for stiffness or guarding. For example, inflammatory and traumatic hip disorders cause a loss of medial hip rotation and guarding of the joint. Evaluate chronic problems for evidence of deformity and an underlying disease. The chronic problem is much more likely to be serious and require a complete and thorough evaluation.

Functional disability is more significant than deformity. Deformity is static; function is dynamic. Deformity is most significant when it adversely affects function. This concept is becoming more universally accepted with time. In the past, handicapped children with conditions such as cerebral palsy were subjected to endless treatments to correct deformity. Often, deformity was corrected at the expense of function. The net effect was harmful.

Some alteration in function is subtle and not readily apparent. For example, a malunited bone forearm fracture may cause a permanent reduction of forearm rotation in the older child. The child compensates for the deformity by rotating the shoulder and may not be aware of any problem. This loss of motion can be detected by physical examination. Determine the degree of disability by functional tests that focus on activities requiring pronation and supination.

Pain

Pain in the child is usually significant. For example, the majority of adults experience back pain but rarely does it require active treatment. In contrast, back pain in children is much more likely to be organic. Pain in the adolescent is more likely to have a functional basis, as is so common in adults. The most common cause of pain in children is trauma. Trauma may result from acute injury or from the so-called microtrauma or overuse syndromes. Overuse syndromes account for the majority of sports medicine problems in children and adolescents.



1 Pseudoparalysis. This child has loss of spontaneous movement of the left arm from a "pulled elbow."



2 Limp. This infant had an obscure limp. The bone scan demonstrated increased uptake over the tibia consistent with a toddler's fracture (arrow).

Point of Maximum Tenderness

The most useful test in establishing the cause of pain is determining its anatomic origin by locating the *point of maximum tenderness (PMT)* [1]. Localization of the PMT, together with the history, often establishes the diagnosis. For example, a PMT over the tibial tubercle in a 13-year-old boy very likely means the boy has Osgood–Schlatter disease [2]. A PMT over the anterior aspect of the distal fibula [1 lower, opposite page], together with a history of an ankle injury, probably points to an ankle sprain. A PMT over the tarsal navicular in a 12-year-old girl suggests the diagnosis of an accessory ossicle [1 opposite page].

The examination to establish the PMT should start distant from the problem. Palpate gently, moving progressively closer to the site of discomfort. Watch the child's face for signs of discomfort. Often a change in facial expression is more reliable than a verbal response. Be gentle. Ask the child to tell you where the tenderness is greatest. With gentleness, patience, and sensitivity, the PMT can usually be established accurately with minimal discomfort.

The PMT is a useful guide in ordering radiographs. A PMT over the tibial tubercle suggests the diagnosis of Osgood–Schlatter disease. If confirmation is necessary, order a lateral radiograph of the knee. Similarly, order oblique radiographs of the elbow if the PMT is over the lateral condyle and the AP and lateral views of the elbow are normal. Fracture of the lateral condyle may be demonstrated only on the oblique radiograph.

The PMT is helpful in evaluating the radiographs. For example, locating the PMT aids in differentiating an accessory ossification center from a fracture. Only a fracture will be tender. To determine if a subtle cortical irregularity in the contour of the distal radius represents a buckle fracture, locate the PMT. If the cortical irregularity represents a fracture, the PMT and the questionable radiographic change will coincide exactly in location.



1 PMT about the hip. The anterior iliac spine (red arrow) and the greater trochanter (yellow arrow) are useful landmarks for determining the PMT about the hip.



2 PMT about the knee. The PMT is easily determined about the knee. The tibial tubercle (red arrow) is tender in Osgood–Schlatter disease. Medial joint line tenderness (yellow arrow) is found with meniscal injuries.

Spondyloarthropathy Seronegative spondyloarthropathies in the incipient stage are associated with a PMT in specific locations. These are referred to as *enthesopathies*. Common sites include the metatarsal heads, plantar fascia, achilles tendon insertion, greater trochanter, and SI joints.

Muscle Testing

Muscle testing is done to determine the strength of muscle groups [2]. Testing is performed for neuromuscular problems such as poliomyelitis and muscular dystrophy. The grades can be further subdivided by a plus or minus designation.

Growing Pains

Growing pains are discomforts of unknown cause that occur in 15–30% of otherwise normal children. Headaches, stomachaches, and leg aches, in that order, are the common pains of childhood. Leg aches characteristically occur at night, are poorly localized, of long duration, and produce no limp or apparent disability. Spontaneous resolution occurs, without sequelae, over a period of several years.

Because the pain of leg aches is so diffuse and nondescript, the differential diagnosis includes most painful disorders of childhood. The conditions a physician must rule out include neoplastic disorders such as leukemia, hematologic problems such as sickle cell anemia, infections such as subacute osteomyelitis, and various inflammatory conditions. The diagnosis of growing pains is one of exclusion, relying primarily on the medical history and the physical examination. Rarely are a CBC and ESR or radiographs necessary. Evaluation and management of growing pains is discussed in greater detail in the Management chapter.



1 PMT about the foot. Because bone and joints of the foot are subcutaneous, the PMT is very accurate and an especially valuable sign. The PMT over the lateral malleolus (red arrow) and over the navicular (yellow arrow) are readily localized.



Grade	Strength	Physical Finding
0	None	No contraction
1	Trace	Palpable contraction only
2	Poor	Moves joint without gravity
3	Fair	Moves joint against gravity
4	Good	Against gravity and resistance
5	Normal	Normal strength

2 Muscle grading. Manual muscle testing is useful in documenting and classifying muscle strength into six categories.

Clinical Tests

Various tests are useful to supplement the general physical examination in children. Some of the more commonly used tests are described below, presented in alphabetical order.

Abdominal Reflex

Stimulate each quadrant of the abdomen [1 opposite page]. Normally the umbilicus moves toward the side being stimulated. This test is commonly used to assess a neurologic basis for spinal deformity (see Chapter 8).

Anvil Test

This tests for the localization of discitis. Percussion on top of the head causes pain at the site of discitis.

Barlow Maneuver

This maneuver is a provocative test for hip instability in developmental hip dysplasia. See page 137.

Coleman Block Test

This tests for hindfoot flexibility. Ask the child to stand on a block positioned under the lateral side of the foot. With weight bearing, the failure of the heel to assume a valgus position is indicative of a fixed deformity.

Ellis Test

This test assesses the tibia-hindfoot length [2 opposite page]. With the patient supine, flex the knees fully. The difference between the knee heights indicates the amount of shortening. This test can also be performed with the child prone. This allows the knees to be flexed to a full 90°.

Ely Test

The Ely test assesses for rectus contracture [3 opposite page]. Place the child prone and flex the knee. If the rectus is spastic or contracted, the pelvis will rise.

Foot-Progression Angle

This test assesses the degree of in-toeing or out-toeing (see Chapter 4).

Forward Bend Test

This assesses the functional and structural stiffness and deformity of the back. While observing the patient from the back and again from the side, ask the patient to bend forward as far as possible. Note asymmetry and stiffness. The normal child should show symmetrical flexion and be able to extend the fingers to at least the knee. The spine should show an even flexion of the thoracic spine and reversal of the lumbar lordosis. The thorax should be symmetrical as viewed from the back and front. Spinal cord tumors, inflammatory lesions, spinal deformity, and hamstring contractures all cause abnormal findings.

Galeazzi Sign

This tests for shortening due to developmental hip dysplasia. Flex both hips and knees to a right angle. Note any difference in apparent length of thighs.

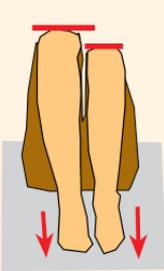
Goldthwaite Test

This test detects lumbar spine inflammation as occurs with discitis. Position prone with hips extended and knees flexed. Moving the pelvis from side to side causes a synchronous movement of the lumbar spine.

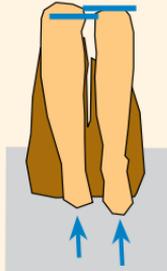


Abdominal reflexes

1 Abdominal reflexes. The abdomen is stroked in all four quadrants. This stimulation causes the umbilicus to move toward the quadrant stimulated. The absence of this response is abnormal.

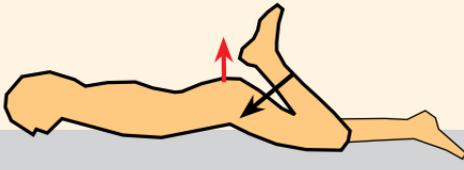


Ribial length
Ellis test



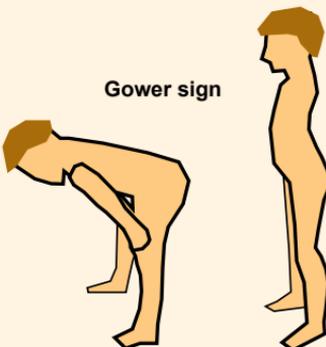
Femoral length
Galeazzi sign

2 Assessing femoral and tibial lengths. Note the difference in tibial and femoral lengths as observed at the flexed knee. With the feet on the table, tibial length differences are apparent (red arrows). With the hips flexed and the feet free, note the differences in femoral lengths (blue arrows).



Rectus contracture
Ely test

3 Rectus femoris contracture evaluation. With this contracture, flexion of the knee (black arrow) causes elevation of the pelvis (red arrow).



4 Gower sign for generalized muscle weakness.

42 Evaluation / Clinical Tests

Gower Test

This tests for general muscle weakness [4 previous page]. Ask the patient to sit on the floor and then stand up without external supports. With trunk weakness, the child uses his hands to climb up his thighs for support.

Hip Rotation Test

The hip rotation test screens for inflammatory or traumatic hip problems [1 opposite page]. Place child in prone position, knees flexed to 90°, and medially rotate both hips. A loss of medial rotation is a positive sign.

Nélaton's Line

This test is useful in clinical assessment of hip dislocation. The tip of the trochanter should fall below a line connecting the anterior iliac spine and the ischael tuberosity.

Ober Test

This tests for tensor fascia contracture [2 opposite page]. Position the patient on one side with the lower knee and hip flexed to a right angle. Abduct and fully extend the upper hip. While maintaining the hip extended, allow the leg to fall into full adduction. An abduction contracture is present if the thigh fails to fall into adduction. The degree of contracture equals the abducted position above the neutral or horizontal position.

Ortoloni Maneuver

This maneuver tests for hip instability in DDH. See page 137.

Patellar Apprehension Sign

This test is for patellar instability. With the knee extended, gradually apply pressure to laterally displace the patella while observing the patient's facial expression. Apprehension indicates previous experience with patellar dislocation.

Patrick Test

This test detects sacroiliac (SI) inflammation [3 opposite page]. Place the ipsilateral foot over the opposite knee. While holding down the opposite ilium, apply a downward force on the flexed knee. Pain at the SI joint is a positive finding.

Pelvic Obliquity Test

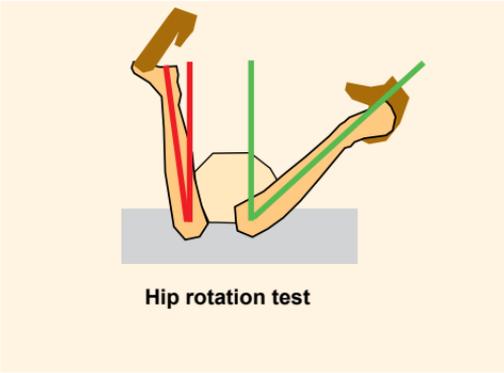
This differentiates suprapelvic from infrapelvic obliquity. Position the child prone with the pelvis on the edge of the examining table, allowing the lower limbs to flex. Windswept positioning of the legs brings the pelvis to neutral if the obliquity is infrapelvic in origin.

Phelps Gracilis Test

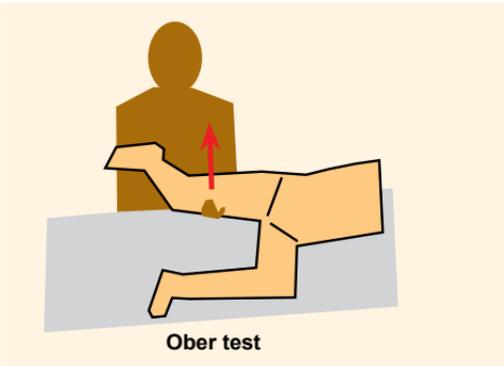
This test is a measure of gracilis spasticity or contracture. Position prone and abduct the hip with the knee flexed. Passive knee extension causes hip adduction if the gracilis is contracted.

Popliteal Angle Measure

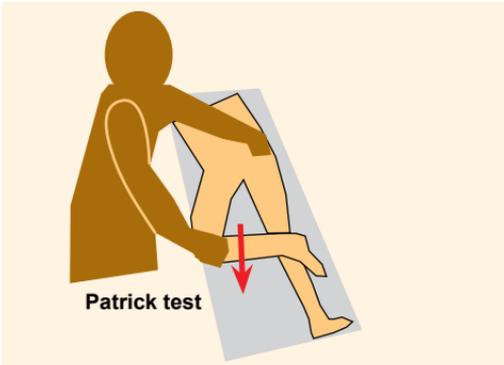
This measures hamstring contracture [4 opposite page]. With the patient supine, flex the hip to a right angle and the knee to a comfortable maximum. The contracture equals the degree of lack of full knee extension.



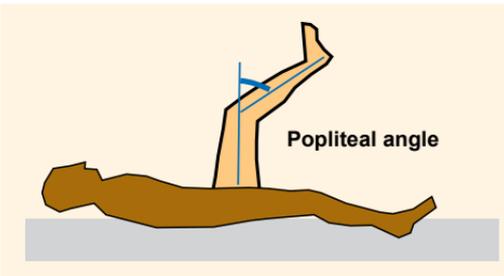
1 Hip rotation test. This screens for traumatic or inflammatory hip problems. A reduction of medial rotation (red angle) is significant, as hip rotation is usually symmetrical in children.



2 Ober test. This tests for tensor fascia contracture. Abduct and extend the leg, then allow it to fall. A failure of adduction is positive for a tensor contracture.



3 Patrick test. This test is performed by positioning the leg across the other and applying downward pressure. This elicits pain in the ipsilateral sacraliliac joint region.



4 Popliteal angle. With the hip flexed, extend the knee. The degrees short of full extension equal the popliteal angle (blue arc).

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Prone Extension Test

This tests for hip flexion contracture [1]. Position the patient prone with the thigh over the edge of the examining table, with one hand on the pelvis and other holding on the leg. Extend the leg until the pelvis starts to elevate. The horizontal–thigh angle demonstrates the degree of contracture.

Thigh–Foot Angle

This is a measure of tibial and hindfoot rotation. See page xx.

Thomas Test

This tests for hip flexion contracture. Flex the contralateral hip fully. The ipsilateral horizontal–thigh angle equals the hip flexion contracture.

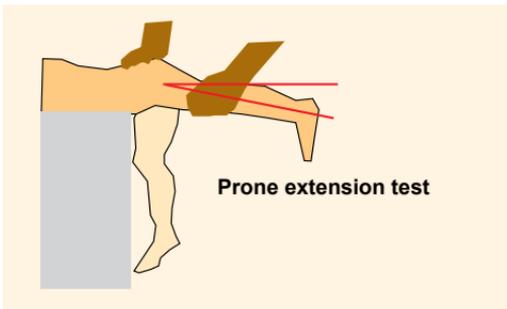
Transmalleolar Angle

This angle is a measure of tibial rotation. See page xx.

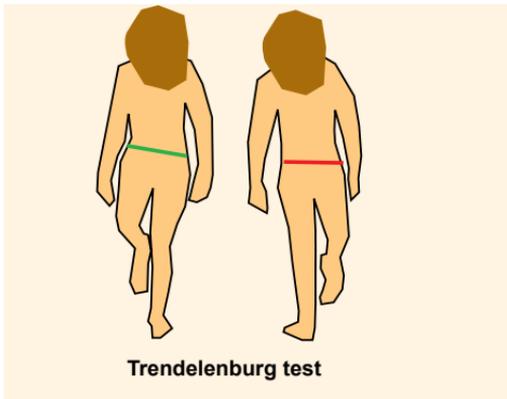
Trendelenburg Test

The Trendelenburg test assesses abductor strength [2]. While observing the pelvis from behind, ask the patient to raise one leg (without holding for support). A drop in the contralateral pelvis indicates weakness of the ipsilateral abductors.

A *delayed* Trendelenburg test is performed by determining the time necessary for the abductors to fatigue, allowing the pelvis to sag. If the elevation of the contralateral pelvis cannot be maintained for 60 seconds, the test is positive.



1 Prone extension test for assessing hip flexion contracture. With the contralateral hip flexed, extend the ipsilateral side to the degree that causes the pelvis to elevate. The degrees short of full extension equal the degrees of contracture.



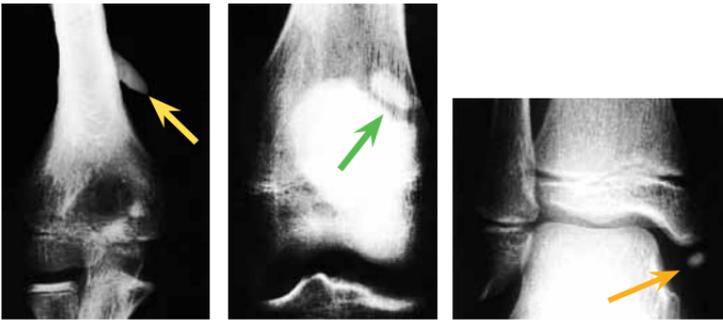
2 Trendelenburg test. Standing on the normal side elicits an elevation in the opposite side (green line). A drop in the opposite side (red line) indicates weakness of hip abduction.

Imaging

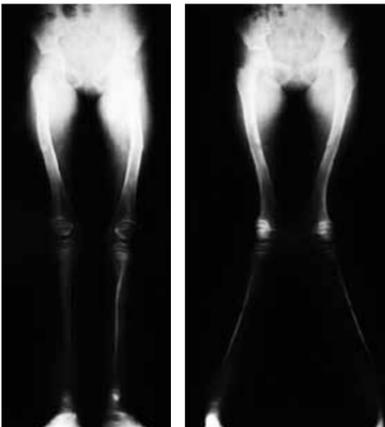
Even after 100 years of experience reading conventional radiographs, we sometimes have difficulty separating disease from normal variability [1]. The lack of experience with new imaging methods makes interpretation even more difficult. Over-reading imaging, such as the MRI, poses risks and may lead to over-treatment. For example, MRI studies of discitis often show extensive soft tissue changes, which might prompt operative drainage if the nature of the disease is not appreciated.

Conventional Radiography

Conventional radiographs are still the mainstay of diagnostic imaging. They are the least expensive, most readily available, and the least apt to be misread. Radiographs show bone, water, fat, and air density well. Bone density must be reduced by 30–50% to show changes on radiographs. Proper positioning of the child is essential. Sometimes the physician needs to position the child. For example, to study genu varum or genu valgum, the child must be placed in the anatomic position with the patellae directed forward. The technician may try to rotate the limbs laterally to fit the legs on the film, creating a deceptive image [2].



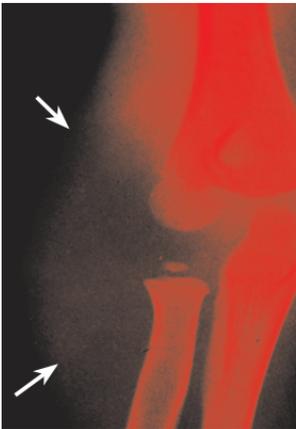
1 Normal variation. The supracondyloid process of the humerus (yellow arrow), a bipartite patella (green arrow), and malleolar ossicles (orange arrow) are uncommon developmental variations of normal.



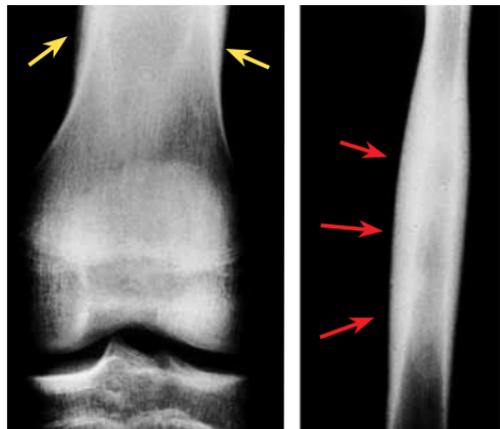
2 Proper positioning for radiographs. This patient had a radiograph made for measuring mechanical axis of the lower limb. The technician rotated the limbs to get the radiograph on one film (left image). A second film was necessary (right image) in which the physician positioned the child in the anatomic position necessary for an accurate measurement.

Limiting radiographs Try to limit radiation exposure by reducing the number of radiographs ordered. The risk of one chest x-ray is considered comparable to smoking 1.4 cigarettes or driving 30 miles. Although the risk is small, it is prudent to limit exposure when possible. Use the following principles to limit exposure to your patients:

1. Shield the gonads when possible except for the initial pelvic image.
2. When possible, order screening radiographs first. For example, if spondylolisthesis is suspected, a single lateral standing spot view of the lumbosacral junction may demonstrate the lesion. AP and oblique studies may not be necessary.
3. Single radiographs are often adequate. For example, a single AP view of the pelvis is usually adequate for evaluating hip dysplasia in the infant or child.
4. Lower extremity and spine radiographs should be taken in the upright position. These standardized views are less likely to be repeated if a referral is necessary.
5. Suggest to primary care physicians that if a consultation is necessary, have the consultant order the studies. Suggest that parents hand carry previous radiographs for consultation, as radiographs are often mysteriously lost in the mail.
6. Order follow-up radiographs only when the information is likely to alter management. For example, ordering a radiograph of a wrist fracture at 3 weeks is generally useless. It is too soon to discontinue immobilization and too late to change position.
7. Finally, the routine practice of ordering a comparative radiograph of the opposite side is often inappropriate.



1 Soft tissue swelling. Soft tissue swelling is an important finding because it suggests that a significant injury has occurred. In this case, the swelling over the lateral condyle was consistent with a lateral condylar fracture. Additional radiographs showed the fracture.



2 Study the edge of the film. Initial radiograph of adolescent complaining of leg pain. The film was read as normal and a diagnosis made of a "conversion reaction." In a later review of the radiograph, periosteal reaction involving femoral distal diaphysis (yellow arrows) was appreciated. Additional radiographs of the whole femur showed extensive sclerosis of the diaphysis (red arrows) due to chronic sclerosing osteomyelitis.

Reading errors Here are some suggestions to avoid reading errors:

1. Study the radiographs in a standardized sequence, starting with the soft tissues [1 opposite page].
2. Study the edge of the film before concentrating on the presumed area of pathology [2 opposite page].
3. If the radiographic and physical findings are inconsistent, order additional views. For example, order oblique radiographs of the elbow if the child has unexplained swelling over the elbow [1 opposite page] and no evidence of a fracture on the initial AP and lateral views. The oblique views will often demonstrate a fracture.
4. Be aware that false negative studies occur in certain situations, such as in the early phase of osteomyelitis and in septic arthritis or developmental hip dysplasia in the newborn.
5. Finally, variations of ossification are often misleading. The accessory ossicles of the foot may be confused with fractures; irregular ossification on the lateral femoral condyle may be misinterpreted as osteochondritis dissecans.

Computerized Tomography Imaging

CT studies provide excellent bone and soft tissue detail [1]. The soft tissue images can be manipulated by computer to enhance tissue separations. This makes the method useful for assessing soft tissue lesions about the pelvis. CT studies can be combined with contrast material for special evaluations, such as CT myelography. Images are obtained in the transverse plane and can be reconstructed by computer with the frontal and sagittal planes or presented as 3-D images for a more graphic display [2]. These studies show relationships well, such as the concentricity of hip reduction and the detailing of dysplasia.

The disadvantages of CT imaging include the need for sedation in the infant and young child, greater radiation exposure, and greater cost than for conventional studies.

Uses for CT Scans

Bone detailing—when conventional radiographs are inadequate
 Spine and pelvic lesions—inflammatory, neoplastic, traumatic
 Complex hip deformity prior to reconstruction
 DDH assessment of reduction in cast
 Physeal bridge assessment
 Complex fractures—such as triplane ankle fractures

1 Uses of CT scans. These are some typical examples of the use of CT scans in assessing musculoskeletal problems in children.



2 Torticollis with plagiocephaly. Asymmetry of the face and skull are demonstrated by 3D CT reconstructions.

Arthrography

Arthrographic studies provide visualization of soft tissue structures of the joints [1]. The contrast is usually provided by air, nitrogen, carbon dioxide, or an iodinated contrast solution. The procedure can be combined with CT or tomography. Arthrography is most useful in evaluating the hip [2] and knee. In septic arthritis, an arthrogram is helpful to confirm joint entry. Arthrography is useful for hip dysplasia and meniscal lesions and in identifying loose or foreign bodies in joints. Disadvantages include the need for sedating younger children and occasional reactions to the iodinated contrast material.



1 Arthrography. Initial radiograph showed a lateral displacement of the upper femoral metaphysis (red arrows) suggesting the possibility of a hip dislocation or subluxation. The arthrogram shows the femoral head to be reduced (yellow arrows) and established the diagnosis of coxa vara.



Uses for Arthrography

- DDH—initial evaluation and when management uncertain
- Perthes disease—to assess shape of cartilagenous femoral head
- Complex trauma—such as elbow injuries in the infant
- Osteochondritis dissecans

2 Arthrography uses. These are typical examples of the use of arthrograms for assessing musculoskeletal problems in children.

Uses for Bone Scans

- Screening—for child abuse
- Limp—localization of site of problem
- Trauma—early stress fractures
- Tumors—localizing lesions, lesion age, differentiating cyst types
- Infections—localizing site or early osteomyelitis, discitis
- Avascular necrosis—LCP disease, osteochondritis staging

3 Uses of bone scans. These are some examples of the use of bone scans for assessing musculoskeletal problems in children.

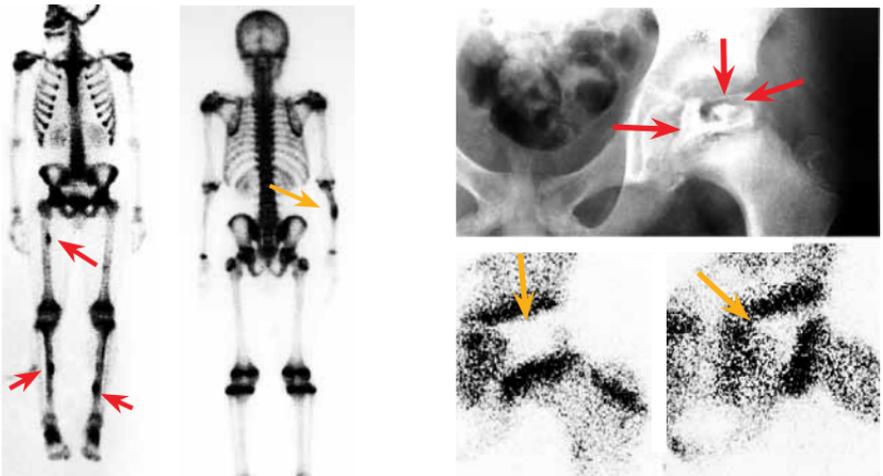
Scintigraphy

Scans utilizing technetium-99m, gallium-67, and indium-111 provide imaging of a variety of tissues. Scintigraphies are more sensitive and show abnormal uptake much earlier than radiographic imaging [3 opposite page]. In addition, bone scanning has a broad scope of applications, including the evaluation of obscure skeletal pain [1] The radiation exposure is equivalent to a skeletal survey with conventional radiographs. Useful options in scanning include a variety of agents, collimator selection, timing of scans, and the use of special techniques.

Collimation "Pinhole" collimation increases the resolution of the image. This is particularly useful for assessing avascular necrosis of the femoral head. Order both AP and lateral views [2].

Agents The vast majority of scans use technetium-99m. This agent has a half-life of 6 hours and, combined with phosphate, is bone seeking. It is highly sensitive, and the images usually become positive in 24–48 hours. Gallium-67 and indium-111 are used primarily for localization of infections. Indium is combined with a sample of the white blood cells from the patient.

Timing Phasic bone scans show the initial perfusion immediately. The soft tissue phase or pooling occurs at 10–20 minutes, and finally, the bone phase is shown after 3–4 hours. Bone scans are not affected by joint aspiration.



1 Bone scans for screening. These screening bone scans demonstrated unsuspected multiple stress reactions in an athlete (red arrows). The other boy (right) has osteomyelitis that is localized to the left ulna (orange arrow).

2 Pinhole collimated bone scan. Conventional radiograph shows avascular necrosis of the femoral head (red arrows). Pinhole collimated scans show reduced uptake in the avascular femoral head (orange arrows).

50 Evaluation / Imaging

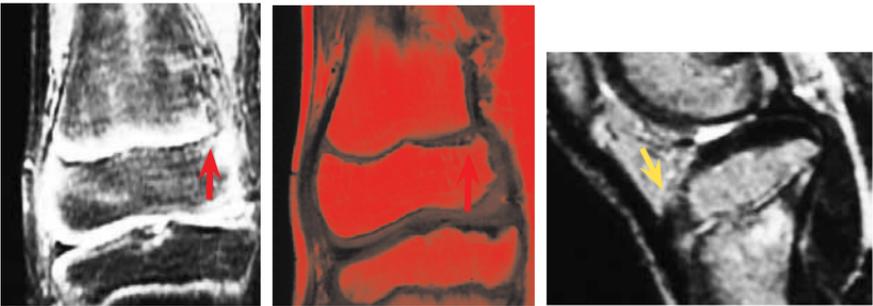
Magnetic Resonance Imaging

MRI provides excellent images of soft tissue [1] without exposure to ionizing radiation. However, it requires expensive, sophisticated equipment and sedation or anesthesia in the infant or younger child for necessary immobilization. Bone imaging is poor, but for soft tissues, MRI is excellent. The interpretation may be difficult because of limited experience, making over-reading a potential problem. Despite these problems, MRIs are proving useful for an increasingly wide variety of conditions [2, 3, and 4].

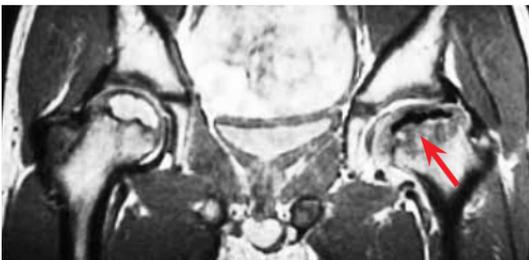
Uses for MRI Studies

Cartilage imaging—meniscal lesion, growth plate injuries
Avascular necrosis—LCP disease, AVN at hip, distal femur
Neural status—spinal cord lesions
Tumors—margins, staging
Infections—soft tissue lesions

1 Uses for MRI. These studies are useful in imaging soft tissue lesions. The usefulness in infants and children is limited by the cost and the need for sedation or anesthesia for immobilization.



2 MRI in physeal injury. Note the defect in the distal femoral physis (red arrows) and the proximal tibial growth plate (yellow arrow).



3 MRI in Perthes disease. The avascular necrosis is clearly demonstrated (arrow).



4 Synovial cyst hip. The cyst (arrow) is clearly seen on this MRI study.

Ultrasound Imaging

Ultrasound applications for the musculoskeletal system are numerous, and the technique is underutilized.

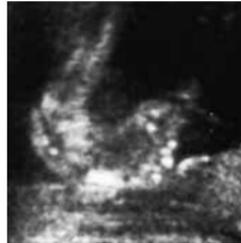
Prenatal ultrasound These studies [1] have the potential of making dramatic changes in orthopedic practice. Here are some useful applications of prenatal ultrasound:

- *Pathogenesis* Improving our understanding of disease in turn improves our ability to prevent or treat diseases.
- *Prenatal treatment* Prenatal treatment, utilizing replacement, substitution therapies, or improving intrauterine environment may correct or improve the problem.
- *Family preparation* Resources can be made available for early postnatal treatment as necessary and preparing families psychologically and educationally.
- *Pregnancy termination* For serious conditions, ultrasound can help determine the need for termination based on the family's choice.
- *Musculoskeletal disorders* The number of these disorders that can be diagnosed by prenatal ultrasound [2] are increasing rapidly with higher resolution studies and greater user experience. False positive studies do occur, however, and may cause considerable unnecessary anxiety in the families.

Clinical uses These studies are highly dependent on operator skill and experience, and in North America, they are usually performed by the radiologist [3]. Ultrasound studies are probably underutilized and could become a practical extension of the physical examination. Ultrasound is safe, potentially inexpensive, versatile, and underutilized in North America.

Uses for Ultrasound—Prenatal

Clubfeet
Skeletal dysplasias
Limb deficiencies
Spina bifida
Arthrogryposis



1 Prenatal ultrasound diagnosis. These musculoskeletal problems can usually be diagnosed.

2 Clubfoot. This clubfoot was identified at 16 weeks of gestation by ultrasound.

Uses for Ultrasound—Postnatal

DDH—evaluation in the young infant
Infections—localization of abscess, joint effusions
Foreign bodies—of the foot
Tumors—especially cystic varieties
Trauma—cartilagenous injuries in young children
Research—measuring torsion, joint configuration

3 Postnatal ultrasound diagnosis. These are typical examples of the use of ultrasound for assessing musculoskeletal problems in children.

52 Evaluation / Imaging

Photography

Medical photography provides an excellent means of documentation [1]. Photographs are inexpensive, safe, and accurate. They are useful in documentation and parent education. The documenting value of photographs is increased by taking certain steps:

Positioning Position for photographs as for a radiograph. Make anterior, lateral, or special views. Position the patient in the anatomic position.

Background Attempt to find a neutral, nondistracting background.

Distance Take photographs as close as possible while including enough of the body to orient the viewer.



1 Clinical photography. Note the cubitus varus deformity of the girl's left arm (red arrow) and the bowing of the boy's right tibia (yellow arrow). The value of each photograph is enhanced by the nondistracting backgrounds, careful positioning, and inclusion of both limbs for comparison. Both photographs document the deformity well and were useful for subsequent evaluation of the effect of growth on severity.

Gait Evaluation

Gait can be evaluated at three levels of sophistication.

Screening Examination

This is part of the standard screening examination and is usually performed in the hallway of the clinic [1].

Clinical Observational Examination

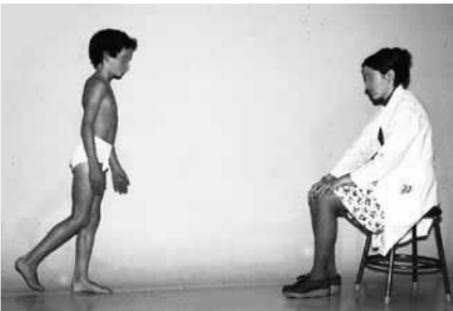
This examination [2 next page] is indicated if (1) the family has reported that the child limps, (2) an abnormality is seen during the screening examination, or (3) the physical findings point to a disease likely to affect gait. In the hallway of the clinic, observe the child walking from the front, behind, and both sides if possible. Look at the child's shoes for evidence of abnormal wear [2]. An abnormal gait often falls into readily identifiable categories:

Antalgic gait Pain with weight bearing causes shortening of the stance phase on the affected side.

In-toeing and out-toeing gaits Assess the foot–progression angle for each side. Average the estimated values and express in degrees.

Equinus gait Toe strike replaces heel strike at the beginning of the stance phase.

Abductor lurch or Trendelenburg gait Abductor weakness causes the shoulders to sway to the opposite side.



1 Clinical observational gait examination. Evaluation of the child's gait is best performed in an open area.



2 Value of observing foot wear. The lack of heel wear (red arrow, left) is evidence of an equinus gait on the left side. Excessive wear on the toes of the shoes is indicative of a more severe degree of equinus (yellow arrows, right) in a child with spastic diplegia.

54 Evaluation / Gait Evaluation

Instrumented Gait Analysis

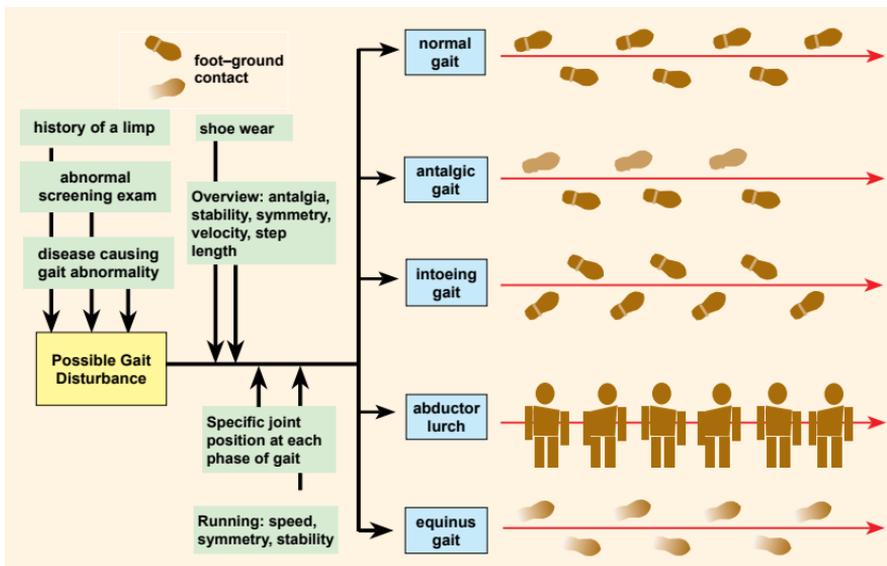
Gait can be assessed by using a video camera to record visual observations. More sophisticated techniques can also be used, including dynamic electromyography to assess muscle firing sequences, kinematic techniques for assessing joint motion, force plate to measure ground reaction forces, and sequence and rate measurements [1]. These values are usually compared with normal values.

Currently, greater attention is being focused on the efficiency of gait by analyzing oxygen consumption and heart rate changes. Over time, we become more concerned about effective and efficient mobility and less about mechanical variations.

The role of the gait laboratory is still controversial. It is clearly an important research tool, but its practicality as a clinical tool remains uncertain.



1 Gait laboratory. The modern gait lab has sophisticated measuring devices to analyze gait.



2 Algorithm for gait assessment.

Laboratory Studies

Laboratory studies provide a limited but useful role in orthopedics. The studies can be combined to reduce the number of needle aspirations.

Hematology

Order a complete blood count (CBC) and erythrocyte sedimentation rate (ESR) and/or C reactive protein (CRP) as part of a screening evaluation to assess the general health of the patient [1], or when infection, neoplasm, or hematologic conditions are suspected.

The ESR is valuable in differentiating infections from inflammation and traumatic conditions. The CRP elevates more rapidly and returns to normal sooner than the ESR. The upper range of value for the ESR is 20 mm/hr. Inflammatory conditions such as toxic synovitis may raise the ESR to the 20–30 mm/hr range, but ESRs above 30 mm/hr are usually due to infection, neoplasm, or significant trauma. Except in the neonate, the CRP and ESR are usually always elevated by infections such as septic arthritis and osteomyelitis. In contrast, a leukocytosis is a less consistent finding.

Chemistry

Serum studies of calcium metabolism are occasionally useful when the possibility of conditions such as rickets is suspected. The normal range of these values is age dependent.

Enzymes

Screen for muscular dystrophy by ordering a creatinine phosphokinase (CPK) determination. Order the test if the young child appears weak, shows a clumsy gait, and has tight heel cords.

Chromosomal Studies

Chromosome studies are indicated for evaluating syndromes with features suggestive of a genetic disorder. These features include multiple system congenital malformations; mental retardation of unknown cause; abnormal hands, feet, and ears; and skin creases.

Bone Mineral Content

Mineral content of bones can be quantitated using several techniques. Cortical measurements can be made by radiography. The second metacarpal is a common standard. Single and dual photon absorptionmetry are other alternatives. These studies are indicated for metabolic diseases, idiopathic osteopenia, and similar disorders.

Condition	Indication for CBC, ESR, and/or CRP
Growing pain	Suspicious features, rule out leukemia
Bone pain	Rule out sickle cell anemia
Stress fracture	Rule out infection
Hip pain	Separate septic arthritis and toxic synovitis
Back pain	Evaluate for discitis
Infection	Follow course of infection

1 Indications for CBC, ESR, and/or CRP. These screening tests are helpful in evaluating a variety of clinical problems.

Electromyography

Electromyography (EMG) is done using either surface or deep electrodes. Surface electrode studies are limited because of artifacts and poor muscle selectivity. The placement of deep electrodes is painful and thus poorly tolerated in children. Furthermore, EMG studies do not show the strength of contraction, only the electrical activity.

EMG is useful in evaluating peripheral nerve injuries, anterior horn cell degeneration, and diseases such as myotonia and myelitis. In peripheral nerve injuries, denervation causes fibrillation potentials 1–2 weeks after injury. During regeneration, the EMG will show polyphasic wave forms. In anterior horn cell degeneration, fasciculations appear.

Nerve Conduction Velocity

Nerve conduction velocity is measured by the time difference shown between the point of stimulation and the recording by EMG. Normal values change with age, from about 25 m/sec at birth to 45 m/sec at age 3 years to about 45–65 m/sec in mid-childhood. The peroneal, posterior tibial, ulnar, median, and facial nerves are usually studied. In children, perform these studies in evaluating peripheral and hereditary neuropathies.

Diagnostic Blocks

Diagnostic blocks are most useful in children for evaluating incisional neuroma and for pain of unknown cause around the foot. By this means, it is possible to localize the site of pain precisely.

Joint Fluid

Joint fluid should be visually examined and also sent to the lab for cell counts, chemistry, culturing, and staining [1]. The joint sugar is usually about 90% serum level and is reduced in infection. In about one-third of cases of septic arthritis, cultures are negative.



Examination	Normal	Septic arthritis	JRA	Traumatic arthritis
Appearance	Straw colored	Grayish	Straw colored	Bloody
Clarity	Clear	Turbid	Slightly cloudy	Bloody
Viscosity	Normal	Decreased	Decreased	Decreased
Total WBC	0–200	50,000–100,000	20,000–50,000	RBCs
PMNS	90+%	Mostly PMNS	Predominate	
Bacteria	None	Seen in about half	None	None
Culture	Negative	Positive 2/3	Negative	Negative
Protein	1.8 g/100 mL	4 g/100 mL	3–4 g/100 mL	Normal
Glucose	20 mg/100,ml below serum	30–50 mg/100 mL below serum	Normal	Normal
Inspection				Fat in aspirate

1 Joint fluid evaluation. Joint fluid differences can be seen among common causes of joint effusions.

Biopsy

The biopsy is an important diagnostic procedure [1] and is not always a simple process. It is preferable for the same surgeon to perform both the biopsy and any reconstructive or ablative procedures. Always remember the old rule: biopsy pus culture biopsy specimens [2]. Needle biopsy for lesions in inaccessible sites – such as vertebral bodies. Plan ahead with the lab to coordinate tissue removal [3 and 4], transfer solutions, frozen sections, and electron microscopic studies.

Tissue	Indication
Muscle	Muscular dystrophy Myositis
Bone	Neoplasms, infections
Skin	Osteogenesis imperfecta
Nerve	Neuropathy

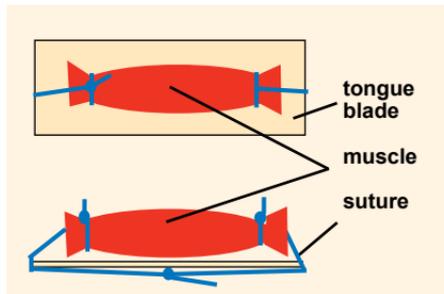
1 Common indications for biopsy. Tissue from bone (left) or other tissues is useful to establish the diagnosis.



2 Osteomyelitis of the clavicle. This lesion is often confused with a tumor. Be certain to obtain biopsy cultures.



3 Biopsy of bone. Biopsies are important procedures that require planning, careful technique, and competent pathologic evaluation.

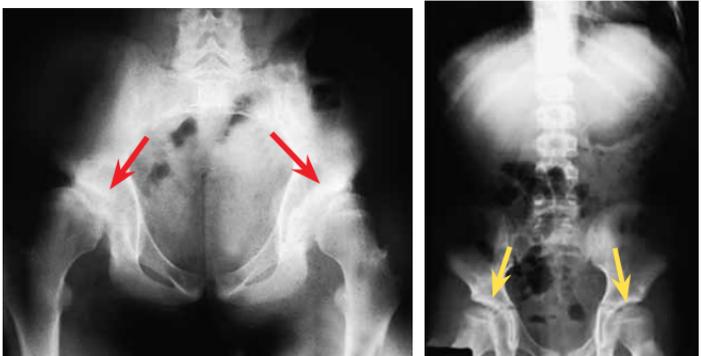


4 Technique of muscle biopsy. Remove a segment of muscle for biopsy (left). Secure the specimen to a segment of a tongue blade with sutures (blue lines, right) to maintain length and orientation during transport and initial fixation.

Time Line

The effect of time and growth on a disorder is called the *time line*. This is also referred to as the *natural history*, or what would happen without treatment. The natural history of many conditions is well known. For instance, we know that nearly all rotational problems resolve with time. Unfortunately, variability from child to child makes the best predictions only estimates. In less common conditions, the course is unknown and the time line is of even greater importance. Sometimes the time line is established by chance [1], but it is usually established by serial radiographs [2 this page, 1 and 2 opposite page] or photographs [3 opposite page]. To establish a time line, the status of the disorder is documented at intervals. During the first visit, obtain baseline studies. The studies are repeated at intervals depending on the disease.

A classic example is in physeal bridge management. If a child sustains a medial malleolar Salter type III or IV injury, it is useful to obtain a baseline full-length radiograph of both tibiae on one film. The same study is made at 3-month intervals. A change in relative lengths of the tibia or a tilting of the articular surface of the ankle is early evidence of a physeal bridge.



1 Chance "time line." This 15-year-old boy was seen for bilateral hip pain. Radiographs demonstrated severe hip dysplasia with subluxation (red arrows). By chance in his old x-ray folder, a KUB was found that was taken when he was 12 years old. His hips showed only mild dysplasia (yellow arrows) at that age.

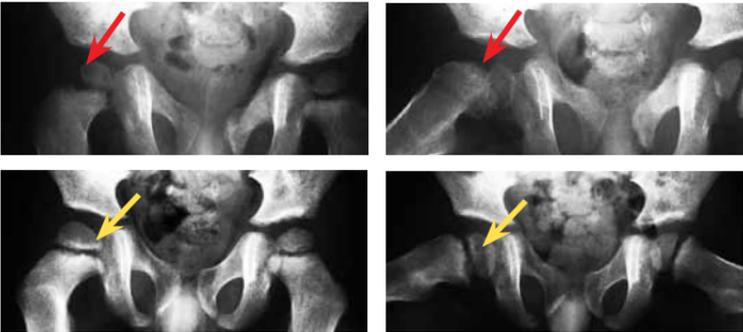


2 Effect of growth. These radiographs show the effect of time and growth when a physeal bridge is present (yellow arrow). Two years later, this 12-year-old boy shows a dramatic increase in valgus deformity of the knee (red arrows).

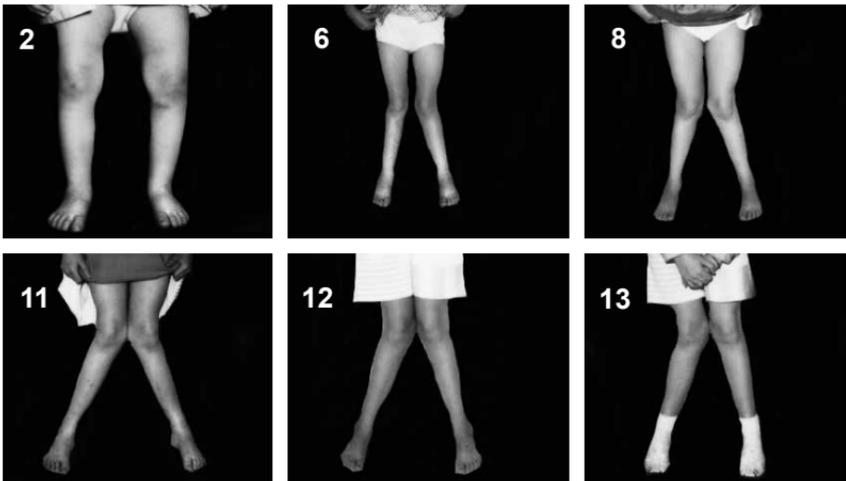


1 Time line using radiographs.

Comparing a sequence of radiographs is a very practical method of assessing the effect of time on deformity.



2 Remodeling. Childhood remodeling of fracture deformity is one of the most graphic demonstrations of the effect of time and growth. This infant sustained a physeal fracture with malunion at 12 months (red arrows). Note the extensive remodeling of the deformity by age 24 months (yellow arrows).



3 Time line using photographs. In this child with vitamin D-resistant rickets, the progression of the genu valgum deformities at ages 2, 6, 8, 11, 12, and 13 years is illustrated. The family and patient elected to delay correction until 14 years of age to avoid recurrence.

Joint Swelling

Joint inflammation is termed *arthritis* [2], whereas joint pain without signs of inflammation is referred to as *arthralgia*. Rheumatologists call pain at ligament and tendon insertions *entheseopathy*. Arthritis occurs in about 2 in 1000 children. The causes of swollen joints in children are numerous [1]. In most cases, the diagnosis [1 opposite page] is established through the approach outlined below.

Differential Diagnosis of Arthritis

Primary

Traumatic

- Direct injury—dislocation, fracture
- Slipped capital femoral epiphysis
- Introduction—foreign body synovitis

Infection

- Bacterial
- Lyme disease
- Tuberculosis

Juvenile rheumatoid arthritis

- Systemic JRA
- Polyarticular JRA
- Pauciarticular JRA
- Spondyloarthopathy

Tumors

- Intraarticular hemangiomata
- Pigmented villonodular synovitis

Vascular

- Legg-Calvé-Perthes disease
- Osteochondritis dissecans

Idiopathic

- Toxic synovitis hip

Secondary

Adjacent inflammation

- Osteomyelitis
- Osteoid osteoma

Systemic disorders

- Leukemia
- Acute rheumatic fever
- Hemophilia with joint effusion
- Acute rheumatic fever
- Systemic lupus erythematosus
- Henoch-Schönlein purpura
- Sarcoidosis
- Postinfectious disorders
- Reflex sympathetic dystrophy

1 Differential diagnosis of joint swelling and pain.



2 Pauciarticular juvenile rheumatoid arthritis. This young girl has little discomfort. Note the swollen right knee.

Approach

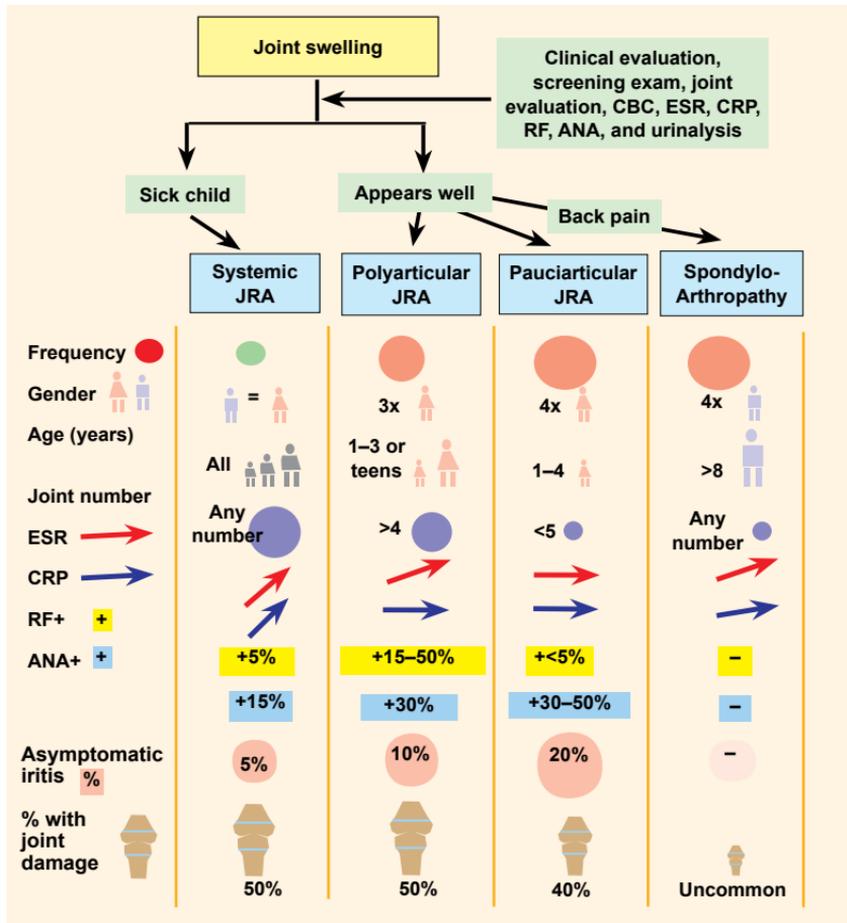
History Ask the patient and family about systemic symptoms, night pain, morning stiffness, other illnesses, family history, duration, severity, and general health.

Examination Perform a careful screening examination. Is the child systemically ill? Carefully examine all extremities to determine if any other large or small joints are involved. Note the degree of inflammation, localization of tenderness, joint range of motion, and any fixed deformities.

Laboratory studies If one suspects juvenile rheumatoid arthritis (JRA), order a CBC, ESR, CRP, ANA, RF, and urinalysis. Order other studies to help separate your short-list differential diagnosis.

Imaging Start with conventional radiographs and add other studies as appropriate.

Joint aspiration Joint aspiration is indicated if an infectious etiology is included in the differential diagnosis.



1 Differential diagnosis of JRA

62 Evaluation / Joint Swelling

Imaging Start with conventional radiographs and add other studies as appropriate.

Joint aspiration Joint aspiration is indicated if an infectious etiology is included in the differential diagnosis.

Clinical Types

Polyarticular JRA occurs in two clinical patterns [1]: young girls and those in adolescence with multiple small and large joint involvement.

Pauciarticular arthritis is the most common form of juvenile arthritis. The patient is most likely a 1 to 4 year old girl [1 previous page]. About a quarter have no pain but are seen because of a swollen joint such as the knee, ankle, and fingers. ANA is positive in 70%, RF negative. About 20% have iritis [2 on page 38]. Early referral of these patients to an ophthalmologist is essential.

Systemic JRA occurs in boys and girls usually between 3 and 10 years. These children are febrile, toxic, have severe myalgias, enlarged nodes, liver, and spleen, and sometimes pericarditis, myocarditis, disseminated intravascular coagulation, and polyarthritis [2]. The course of the disease is variable. Some cases resolve in months, others persist causing joint destruction and disability.

Feature	Early	Late
Age	1–4 years	Adolescence
Sex	Girls	Girls
Joint involvement	Varied and sym.	Large and small, sym.
ANA	Positive	Few
RF	Negative	Half positive
Iritis	Uncommon	Rare

1 Types of polyarticular JRA. This shows two types of presentation of polyarticular JRA. From Sherry (1998).



2 Hand involvement in systemic JRA. Note the swelling of multiple joints.

Seronegative spondyloarthropathies Seronegativity is an absence of rheumatoid factor. These disorders include ankylosing spondylitis, reactive synovitis, Reiter syndrome, and those associated with inflammatory bowel disease and psoriasis. These patients are frequently HLA-B27 positive and are usually adolescent boys. These patients may have low-grade systemic signs of fever, weight loss, and malaise.

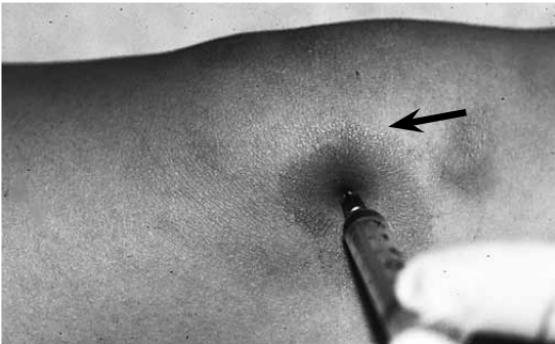
- **Ankylosing spondylitis** This condition is most common in adults but does occur in older children. Inflammation involves the spine, SI, and large joints. Back pain and morning stiffness are common complaints. Stiffness on forward bend test is found. Laboratory findings usually include a mildly elevated ESR, CRP, a positive HLA-B27, and negative ANA and RF. Radiographic changes are late.
- **Reiter syndrome** The triad of arthritis, urethritis, and conjunctivitis are usually found. Painful photophobic iritis can occur. The disease usually follows dysentery or sexually transmitted disease.

Management

Intraarticular triamcinolone hexacetonide (steroid) injections are effective in reducing synovitis and sometimes preventing joint destruction [1].

Systemic agents include ibuprofen, and methotrexate and etanercept. These drugs are best administered by a pediatric rheumatologist.

Joint damage occurs in most types of JRA [1 next page]. Pauciarticular arthritis causes damage but the interval between onset and damage is longer.



1 Joint injections with steroids. Injections in JRA-affected joints dramatically reduce the synovitis and joint destruction.

64 Evaluation / Joint Swelling

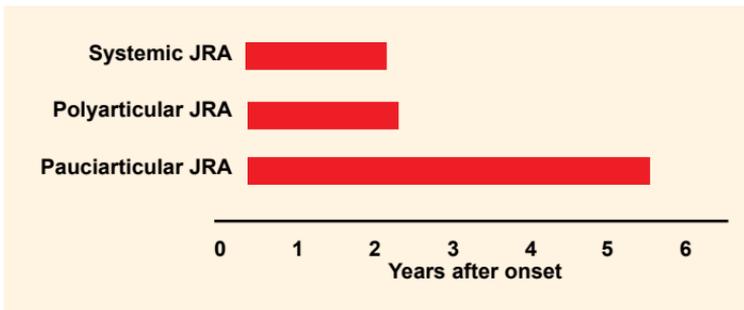
Pitfalls

Confused with trauma A swollen joint is frequently thought to be secondary to an injury. As injuries are a daily occurrence in the life of a child, a history of an injury is common. Swollen joints are seldom the result of an injury. They require evaluation and an accurate diagnosis.

Iritis can accompany pauciarticular and some polyarticular forms of JRA [2]. The iritis is usually asymptomatic and can lead to blindness. Children with these forms of arthritis should be referred to an ophthalmologist for evaluation. The risk of iritis makes an early diagnosis of arthritis of great importance.

Missing septic etiology Permanent joint damage is most likely to occur quickly from septic arthritis. Septic arthritis of the hip is most difficult to differentiate. Monarticular arthritis of the hip is seldom due to JRA.

Missing leukemia Bone and joint complaints are the initial symptoms in 20% of children with leukemia. Leukemia causes bone pain, systemic illness, high ESR, and anemia.



1 Time between onset and joint damage. Mean time between onset of different types of JRA and onset of joint damage.



2 Features of iritis. These photographs show irregularity of the pupil. These are late findings that result from adhesions between the iris and lens. Courtesy DD Sherry.

Limb Deficiencies

Congenital limb deficiencies occur in about 0.1 to 0.2 in 1000 children, or about one-tenth the frequency of clubfeet or DDH.

Causes

Most deficiencies occur in children who are otherwise normal and have no genetic basis. Thalidomide is known to cause multi-limb deficiencies [1]. Most limb deficiencies are sporadic [2 on this page and 2 on next page]. Tibial hemimelia [1 next page] is transmitted as a dominant trait. In other cases, deficiencies are associated with various syndromes such as the radial aplasia–thrombocytopenia syndrome. Acquired amputations result from trauma or treatment of malignant tumors.

Nomenclature

The most widely accepted nomenclature is that of Frantz and O’Rahilly, which divides limb deficiencies into intercalary and terminal types [1 on page 41]. Every type of deficiency is usually classified [All illustration pages xxx and xxx]. These classifications aid in defining severity and indicating treatment methods (see Lower Limb and Upper Limb chapters for details of treatment).



1 Phocomelia.



2 Tibial deficiency. Note the short leg with a normal foot.

66 Evaluation / Limb Deficiencies

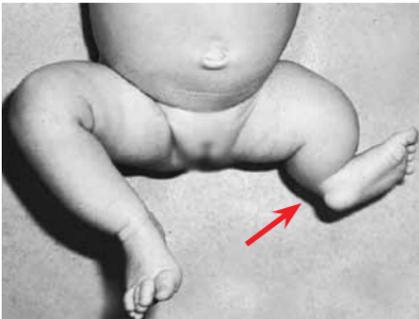
Prevalence

Males outnumber females 3:2, and lower extremities are twice as affected as upper. In 80% of cases, single limbs are involved. Congenital causes are three to four times more frequent than acquired amputations.

Evaluation

Most deficiencies are associated with limb shortening. Order comparative radiographs if a reduction deformity is suspected. Classify the deformity according to the radiographic appearance. Classification is more difficult early on because of lack of ossification. Consider possible associated problems, especially in children with radial deficiencies.

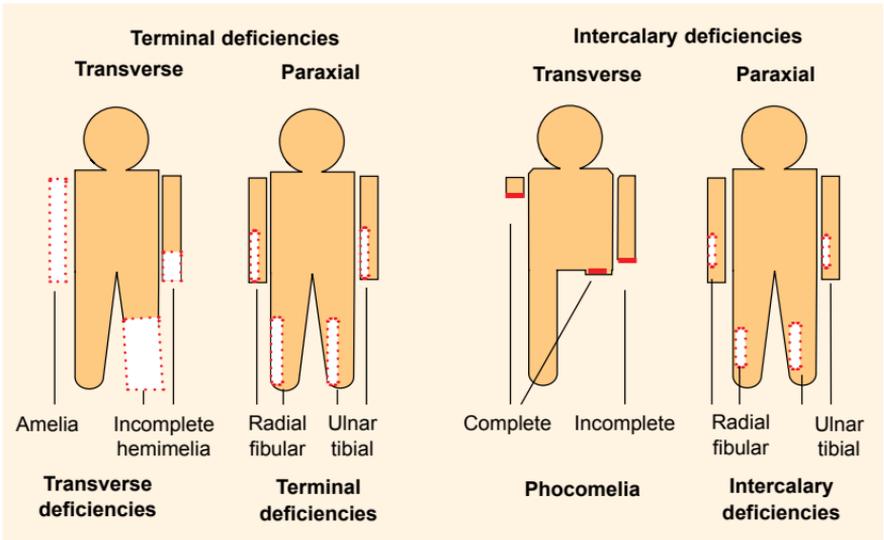
Refer most patients to a limb deficiency clinic. Such clinics provide several important resources for the family: (1) geneticists to evaluate for possible associations and provide family genetic counseling, (2) families to provide support groups, (3) prosthetics to provide often complex fitting problems, and (4) orthopedic surgeons to provide overall management.



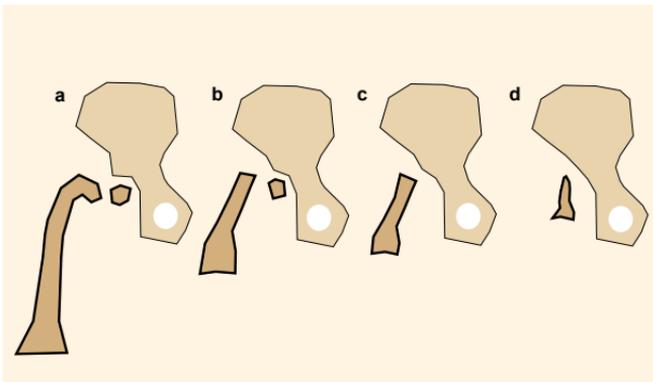
1 Fibular deficiency. Note the shortening, ankle valgus, and hypoplasia of the foot.



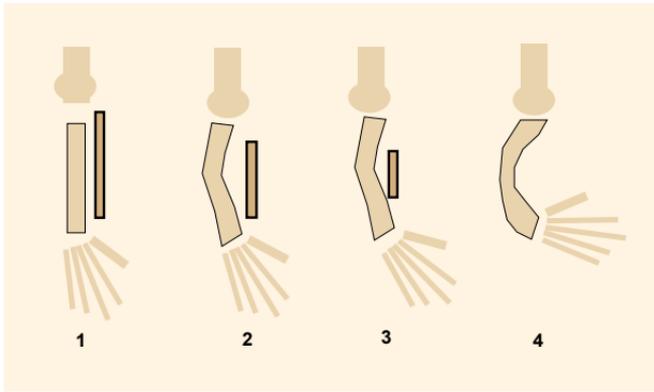
2 Fibular deficiency with bilateral three-rayed feet.



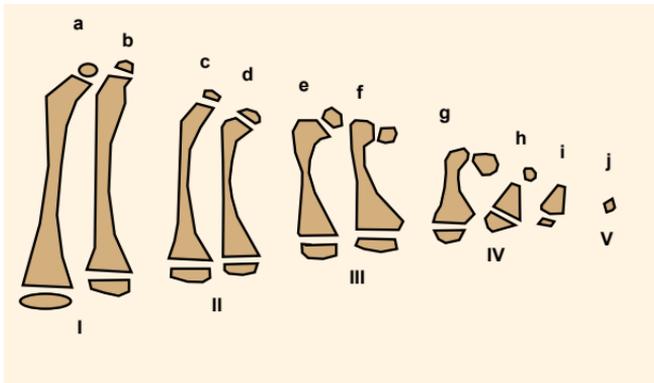
1 Frantz-O'Rahilly classification of congenital limb deficiencies.



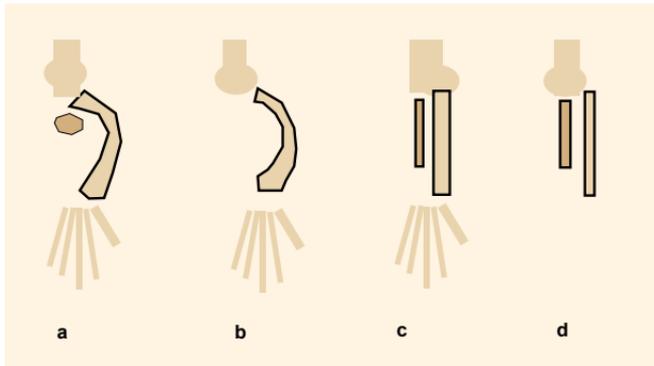
2 Proximal focal femoral deficiency. (a) Good acetabulum, varus deformity of femur. (b) Fair acetabulum, delayed ossification of femur. (c) Poor acetabulum, femoral head absent, femur very short. (d) No acetabulum, femur nearly absent. Based on Aitken (1968).



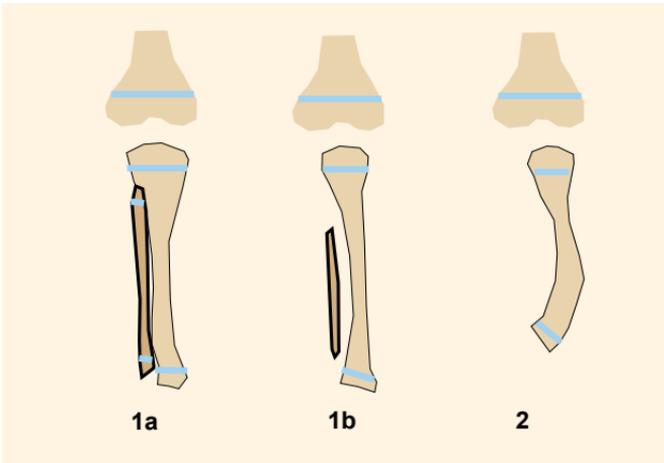
1 Heikel classification of radial deficiencies. (1) Hypoplasia of distal radius. (2) Distal and proximal shortening and ulnar bowing. (3) Varied ulnar shortening with carpal deviation. (4) Radial aplasia. From Heikel, AOS Suppl 39:1 (1959).



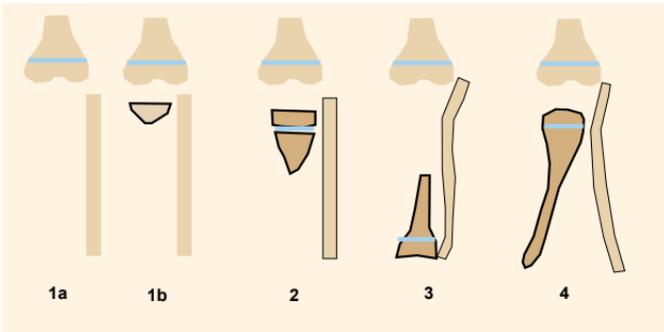
2 Spectrum of the congenitally short femur. This figure shows the wide variation in deformities included in this classification. Based on Hamanishi, JBJS 62B:569 (1980).



3 Swanson classification of ulnar deficiencies. (a) Hypoplasia of the ulna. (b) Absence of the ulna. (c) Fusion of the humerus and radius with hypoplasia of the ulna. (d) Hypoplasia of the ulna with absence of the hand. From Swanson et al., J Hand Surg 9A:658 (1984).



1 Fibular deficiency classification. Type I includes all cases in which some fibula is present. In type 1a, the fibula is short with the proximal fibular physes below that of the tibia and the distal physis above the ankle. In type 1b, the fibula is significantly shortened and not supporting the ankle. In type 2, no fibula is present. From Achterman and Kalamchi, JBJS 61B:133 (1979).



2 Tibial deficiency classification. (1a) The tibia is not seen. (1b) Tibia seen on MRI or US. (2) Distal tibia not seen. (3) Proximal tibia not seen. (4) Diastasis. Based on Jones et al., JBJS 60B:31 (1978).

Chapter 3 – Management

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This chapter covers principles of management. Details are provided in later chapters.

Managing the Family

Skill in dealing with the parents and family is essential in providing optimum care for the child. This requires professional competence, patience, and empathy for child and family. Dealing with parents is often the area of greatest difficulty for the orthopedic resident. Developing appreciation, sensitivity, and skill in communicating with parents and the ability to calm their anxieties are essential skills in dealing effectively with the child's problem.

Child

The child's overall well-being is the primary objective of management. Doing what is best for the child requires respect for the inherent value of childhood as an important time of life [1]. Childhood is more than just a preparatory period of life; it has intrinsic value. Moreover, unnecessary interference with the child's life deprives the child of important life experiences. This concept is especially important in pediatric orthopedics, where the physician often deals with chronic disease; "medicalization" of childhood is a serious risk. We may create what is referred to as the vulnerable child syndrome. These children are often harmed by unnecessary restrictions. Some philosophical and practical guidelines are given here:



1 Play is the occupation of the child. Childhood is the time for varied experiences and has intrinsic value. From Staheli (1986).

1. Resist the pressure to treat the child simply to satisfy the parents or just to “do something.” This is harmful to the child, disruptive to the family, expensive for society, and poor medical practice.

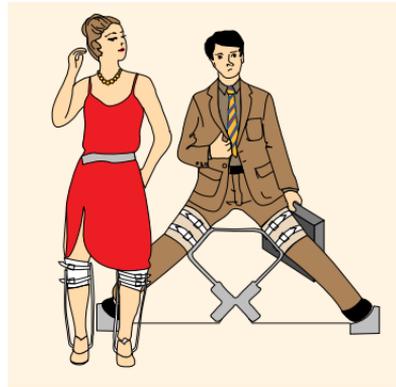
2. Order treatment only when intervention is both necessary and effective. In the past, treatment was commonly prescribed for conditions that resolve spontaneously, such as in-toeing, flexible flatfeet, and physiological bowlegs. Observational management, a policy of monitoring the child’s condition with minimum intervention, provides optimum care for a large percentage of pediatric orthopedic problems. It is least disruptive to the child’s and family’s life and generates a reputation of honesty and competence for the physician.

3. Limit the child’s activity only after thoughtful consideration Play is the primary occupation of the child. Unnecessary restriction denies the child play experiences vital to enjoying childhood and developing critical skills. In some situations the physician may need to curb the parents’ tendency to overprotect the child. It may be in the child’s best interest to risk injury rather than to have long-term constraints on natural activity [1].

4. Avoid medicalization of the handicapped child Overtreatment can further limit the child and overwhelm the family. Excessive numbers of physician’s visits, operations, therapies, braces, and other treatment will result in a large share of the child’s life being expended on treatment that may provide little or no benefit.



1 Integrate treatment with play. Encourage families to have their child participate in physically active play during treatment. These pictures were taken by a mother who achieved this goal.



2 Child’s treatment unacceptable for adults. Treatment that has been commonly prescribed for children such as twister cables for intoeing girls or Perthes disease braces for boys, would never be accepted by an adult patient.

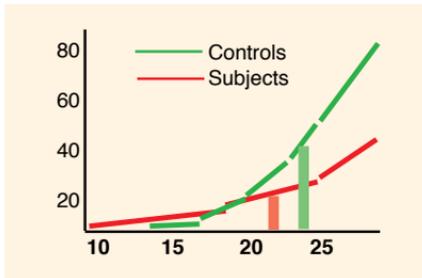
5. Before considering any treatment, consider the child as a whole Treatment methods readily prescribed for children would never be accepted by an adult [2 opposite page]. Orthopedic treatment can be damaging to the individual's self-image [1] and be uncomfortable or embarrassing for the child [3]. Make certain that the anticipated benefits of treatment exceed the harmful psychological, social, and physical effects on the child.

6. Care of the child requires the highest medical standard The results of treating a child, whether good or bad, may remain with the patient for 70 years or more.

Parents

Dealing with parents is an essential part of a pediatric practice [2]. Each family has certain rights, such as privacy, that must be respected, as well as differing needs and values.

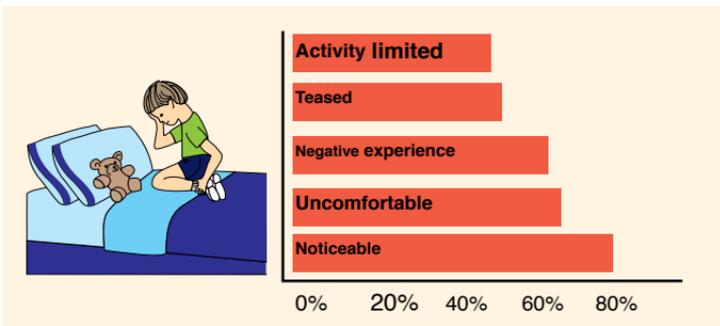
Family coping ability should be respected. Respect the family's resources concerning time, energy, and money. A handicapped child adds stress and complexity for any family. Balance the treatment plan and the family's resources. Consider the well-being of the other children and the health of the marriage; if these are marginal, it may be prudent to order only essential treatment. At different times during management, encourage questions and discuss progress with the family. Being sensitive to the coping ability of the family is part of the physician's responsibility. Demanding more than the family can handle results in noncompliance that may be more the fault of the physician than the family.



1 Orthopedic treatments and self-image. Adults who wore corrective devices as children (red) showed a significantly lowered self-image compared to controls (green). From Driano, Staheli, and Staheli (1988).



2 Parent discussions. Take the families' concerns seriously. Allow enough time to explain the disease and treatment options thoroughly.



3 Recollections. Percentage of adults recalling experience about wearing orthopedic devices during childhood. From Driano, Staheli, and Staheli (1998).

Informed consent should be part of all management, whether surgical or not. The family has the right to know the pros and cons of the management alternatives. The physician's influence is greater with adults as parents than as patients. Most parents are very sensitive to the possibility that the child's current condition may cause some disability in adult life. Certain words such as "arthritis," "crippled," and "pain" have a powerful effect on parents and should be used with caution. For example, in the past, many rotational osteotomies were performed to correct femoral antetorsion under the assumption that the procedure would prevent arthritis of the hip. Although the prophylactic value of the procedure was uncertain, parents readily gave their consent under the presumed threat of arthritis [1]. Several recent studies have shown no relationship between femoral antetorsion and arthritis.

Support and reassurance should be provided for patients and parents. In managing common resolving problems such as intoeing [2], reassurance is the main treatment. With more serious problems, reassurance may take the form of providing information that dispels the parents' fears about the future. In critical conditions, reassurance consists of assuring the family that you will support them throughout the disease. The process of providing effective support and reassurance involves several steps:

- *Make certain that you understand the family's concerns* and take these concerns seriously.
- *Conduct a thorough evaluation of the child.* Pay attention to the family's specific concerns. For example, if they are anxious about the way the child runs, be certain that you observe the child running in the hallway.
- *Provide information about the condition,* especially the natural history.
- *Offer to follow the problem in the future.* Not all positional deformities resolve with time. Offer to see the child again if the family has ad-



1 Complications of rotational osteotomy. This patient underwent an osteotomy for correction of femoral antetorsion. He developed a streptococcal wound infection. This was further complicated by failure of the fixation, nonunion, osteomyelitis, and limb shortening. He had 13 operations to manage complications.



2 Femoral antetorsion improves with growth. This is considered a variation of normal that requires no treatment.

ditional concerns. If the family is obviously apprehensive, or there is someone in the family who is the major source of concerns, such as the grandmother, it may be necessary to provide reassurance repeatedly. Suggest that the grandmother accompany the child during the next visit.

- *If the family is still unconvinced, suggest a consultation.* An offer to refer the child usually increases the family's confidence in the physician. Be certain to communicate to the consultant the family's need for reassurance and not that you are recommending some treatment.
- *Avoid submitting to family pressure for treatment that is not medically indicated.* Performing unnecessary or ineffective procedures because of family pressure is never appropriate.

Procedures are a source of family stress. Whether or not the family should be present during procedures, such as joint aspiration, should be managed individually. Some parents prefer not to be present; others insist on being with the child. Whenever possible, give the family a choice. Be aware that if the parents are present, one of them (usually the father) may feel ill or dizzy and need to lie down. More often, a parent can help calm the child [1]. Moreover, the presence of parents helps to prevent feelings of abandonment in the child [2]. In summary, even though the parents' presence may add a complicating factor for the physician, it may be of benefit to the child.

Litigious problems are fortunately less common in pediatrics compared with other orthopedic subspecialties. However, the legal exposure period for the physician is much longer, because the statute of limitations usually starts at the age of majority. Medical competence, attention to detail, and good rapport with the family are the best protective measures. Additional measures include complete records, generous use of consultants, and avoidance of nonstandard treatments. If an unusual or tragic incident occurs, document the circumstances honestly and thoroughly. Be especially attentive to the family at this time and respond quickly to their concerns.



1 Procedures are less stressful in a supportive environment. Often the mother is best able to comfort the child.



2 Supportive measures during casting. Ongoing support of the child during simple procedures calms and quiets the child.

Religious beliefs may affect the physician's management. Religious beliefs should be respected to the extent that they do not compromise the child's treatment [1]. Discuss the parents' beliefs and concerns openly. Issues regarding blood replacement are common. Alternatives are possible, so do not victimize the child by taking a rigid position against the family. With planning, careful technique, hypotensive anesthesia, and staging if necessary, nearly all orthopedic procedures can be managed without blood replacement. Some families will want a period of time for prayer before giving consent for an operative procedure. Unless time is critical, a negotiated delay is appropriate. Establish a time limit and determine some objective outcome measures in advance.

Family values should be incorporated into the management plan. For some medical conditions, management indications are unclear or controversial. Inform the family of the situation and discuss the choices openly so that the management is consistent with the family's values [2]. Family feelings about operative procedures, bracing, therapy, and other treatment methods vary considerably. The family's feelings and values should be respected but should not supersede the delivery of optimal medical care. Performing an operation that is medically not indicated because of an insisting family is not appropriate.



1 Religious beliefs. Respect the family's right to make or at least influence medical decisions that do not compromise the child's treatment.



2 Families values. Incorporate these values in planning management.

Difficult families may tax the physician's ability to deal with the parents' reaction to their child's illness. The parents may become overprotective or, conversely, may abandon the child. Some parents become abusive toward the physician and staff. Be sure that the parents' behavior does not adversely affect your management of the child. Be understanding but firm, and when appropriate, support abused staff members. Write a note in the chart summarizing the parents' behavior.

Grandparents often accompany the child to clinic [1]. Grandmothers are often concerned about infants' flatfeet, intoeing, or bowlegs. In the grandmother's child-rearing era, positional problems were poorly understood and routinely treated. Overcoming such misconceptions requires a willingness to respectfully explain the reasons for current management.

Unorthodox methods of care by nonphysician practitioners are often considered by parents. Such practitioners usually prescribe treatment, and the treatment often continues over a long period of time. By current standards, such treatments are generally unnecessary and ineffective. Moreover, the treatment may delay necessary treatment. Avoid criticism when discussing these "treatments" with the family; instead, focus on parent education. This is much more effective than criticism. If the parents insist on unorthodox treatment, suggest an objective outcome measure and reevaluate the child later. If appropriate management cannot wait, use a more aggressive approach. Start with the basic facts, obtaining consultations for reinforcement if necessary.

Society

The physician's responsibility to society is seldom addressed. Physicians do have the responsibility to keep health care costs to a minimum by avoiding inappropriate management. We can also choose the least expensive alternatives among equivalent management methods [2].



1 Grandparents in clinic. This grandmother is very well informed and an asset in helping the parents deal with their children's problems.



2 Home traction. Home treatment programs provide cost-effective and convenient management.

Shoes

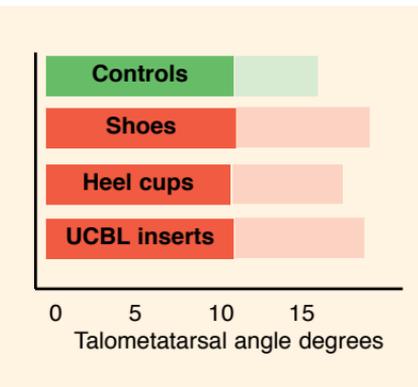
For a long time, shoe modifications were a traditional treatment of infants and children for a wide variety of pathological and physiological problems. Because shoe modifications were usually prescribed for spontaneously resolving conditions, resolution was falsely attributed to the shoe. This led to the concept of the "corrective shoe." Recently, databased studies have consistently shown that natural history, rather than shoe modifications, was responsible for the improvement [1]. We now know that the term "corrective shoe" is a misnomer. Barefooted people have been shown to have feet that are stronger, more flexible, and less deformed than those wearing shoes [1 this page and 1 opposite page]. The feet of infants and children do not require support and do best with freedom of movement without shoes.

Shoe Selection

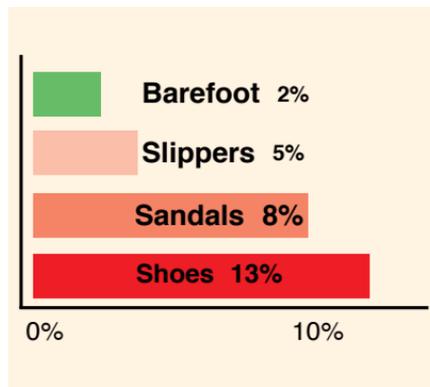
The selection of shoes should be the same as for other clothing. The shoe should protect the foot from injury and cold and be acceptable in appearance. The best shoes are those that interfere least with function and simulate the barefoot state [2 opposite page]. High-top shoes are necessary in the toddler to keep the shoes on the feet. Proper fit is desirable, not to promote support but to avoid falls and compression of the toes. Falls are more common if the shoes are too long or have sole material that is slippery or sticky.

Useful modifications

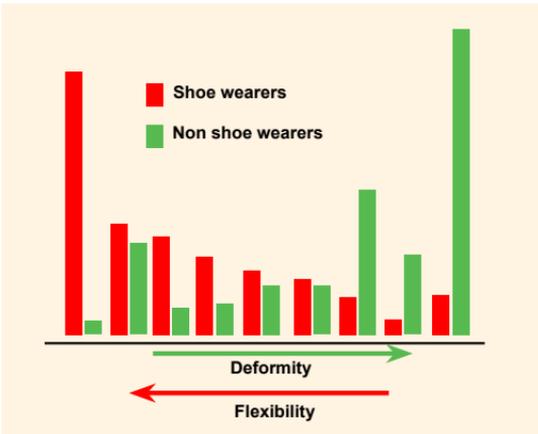
Shock-absorbing footwear may be helpful for the adolescent in reducing the incidence of overuse syndromes [4 opposite page]. Some shoe modifications are helpful [3 opposite page]. These are not for correction but to improve function or provide comfort. Shoe lifts may be useful if leg length difference exceeds 2.5 cm. Orthotics are effective in evenly distributing loading of the sole of the foot.



1 Effect of shoe modifications in flatfeet. This prospective, controlled study compared arch development with various treatments. No difference was found. Talar-metatarsal angles before (light shade) and after treatment (dark shade). From Wenger et al. (1989).



2 Effect of shoe wearing on incidence of flatfoot in adults. In a survey of adults, the percentage with flatfeet was related to footwear during childhood. Note that flatfeet were least common among barefoot children. From Roe and Joseph (1992).



1 Effect of shoe wearing on the incidence of deformity and flexibility in adults. In a survey of Chinese adults, those who wore shoes had more deformity and less flexibility than nonshoe wearers. From Simfook and Hodgson (1958).

Feature	Purpose
Flexible	Improve mobility and strength
Flat	Distributes weight evenly
Foot Shaped	Noncompressive
Friction	Prevents slipping & sticking
Appearance	Acceptable for the child
Cost	Acceptable for the parent



2 Characteristics of a good shoe. The best shoes are those that allow normal function of the foot.

Problem	Modification
Short Leg	Shoe lift for differences >1"
Rigid Deformity	Orthotic to equalize loading
Heel Pain	Elevate heel
Overuse Syn.	Shock absorbing features
Bunions	Stretch shoe over bunion



3 Useful shoe modifications. These modifications are useful to improve the mechanics of load bearing.

4 Cushioned shoes. Shoes have cushioned heels and soles (arrow) that may reduce the incidence of overuse syndromes.

Surgery

Operative Indications

Operative procedures are the most definitive mode of treatment in pediatric orthopedics. The outcome of the procedure is largely determined by the judgment and skill of the surgeon. Children have both greater healing potential and fewer complications than adult patients, increasing the chances for a successful outcome.

Indications for operations include pain, limited function, unsatisfactory appearance, and as a means of preventing future disability. Prophylactic operations are appropriate only when the natural history is known and serious disability is relatively certain. In some, the need for surgery is well accepted [1]; in others, persisting deformity is unexpected [1 opposite page].

Management options Orthopedists can choose from among a wide range of treatment options. First, try a nonoperative treatment, if there is a reasonable chance of success. If the patient has a tarsal coalition and pain, immobilize the foot in a cast for several weeks as a trial. If the pain recurs, excise the bar. If the family is seeking multiple consultations, it is often wise to start with a nonoperative approach. If the family is uncertain, and waiting will not jeopardize the outcome, simply delay the needed operative correction until the family is ready to make a decision.

Conservative management Sometimes an operation is the most conservative of the treatment options. If so, proceed with this treatment first. This approach benefits both the family and child. There are a number of classic examples where delays in operative correction harms the child. A

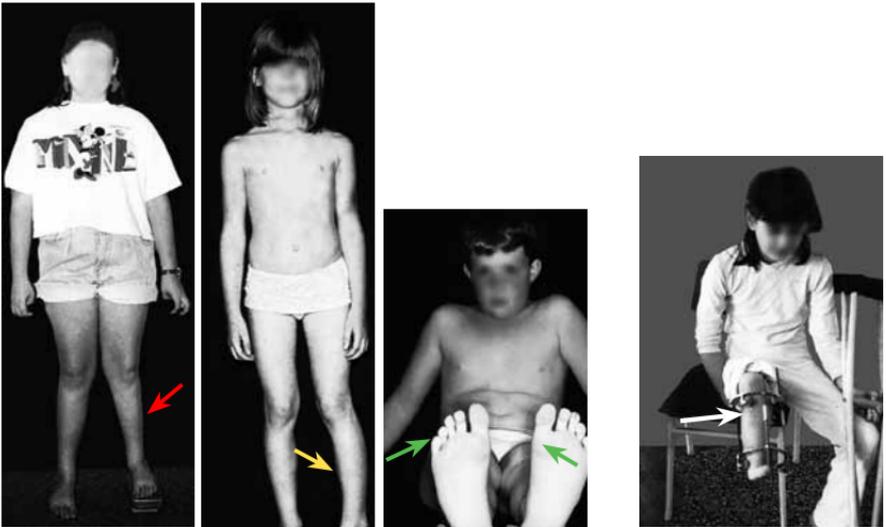


1 Limb deficiencies. Fibular hemimelia (red arrow) causes moderate disability, whereas this child with bilateral tibial hemimelia (yellow arrows) is severely disabled.

12-year-old girl with a 50-degree right thoracic scoliosis is given a trial of treatment with a brace and physical therapy for several years, only to have the 60-degree curve instrumented and fused at age 15. This unfortunate girl experienced the hardships of 3 years of unnecessary brace treatment. Long-term brace treatment is not a benign option; it is often a psychologically damaging experience. In another case, a child with cerebral palsy and a subluxation of the hip is managed by physical therapy. Subluxation progresses to dislocation. An early adductor lengthening or transfer would have been the conservative approach.

Cosmetic disability A cosmetic disability may justify operative correction. Unsightly genu varum or valgum may be corrected by a hemiepiphysiodesis. An abductor lurch may be corrected with a trochanteric transfer. A severe kyphotic deformity may justify instrumentation, correction, and fusion. Each treatment carries risks. Weighing the risks and benefits is often difficult.

Families Role Provide factual information regarding risks and benefits of each alternative, and then allow the family to choose among the medically acceptable options. Be aware that issues concerning body image peak during early adolescence. What is bothersome at age 14 may become acceptable at age 17. Delay the correction of marginally disabling deformities until it is clear that the concern is lasting. Performing an unnecessary operation just because the parent wishes to do something is not appropriate [2]. First and foremost, the orthopedist is the advocate of the child.



1 Uncommon outcomes. Normal variability allows atypical outcomes. In these children the severe genu valgum (red arrow), tibial torsion (yellow arrow), and persisting forefoot adductus (green arrows) are rare outcomes of conditions that nearly always resolve spontaneously.

2 Inappropriate lengthening. This child is undergoing a lengthening of the tibia without a functional foot. The child should have had a Syme type amputation and prosthetic fitting.

Preoperative Planning

During the final preoperative evaluation, think through each step of the procedure to make certain that the preoperative planning is complete. Be certain that special tools or implants will be available.

Anticipate Possible Complications Look for problems that may complicate the procedure. The most common problems are respiratory infections and skin lesions. Check the temperature, evaluate the ears, throat, and lungs. Examine the skin about the operative site for inflammation. The decision regarding respiratory status is generally made by the anesthesiologist. It is usually wise to reschedule the procedure if the child has an unexplained fever, a respiratory infection, infected or inflamed skin lesions in the operative area, or documented exposure to a contagious disease, such as chickenpox or measles. Make certain the family understands the procedure and follow-up plans. Use a model or a skeleton to explain the operation to the family.

Discharge Plans Make discharge plans at this time. Arrange for adaptive equipment necessary for home care [1]. If the child will be using crutches or splints [2] after the procedure, make fittings before the operation. Plan transportation home and anticipate special needs [1 opposite page]. Plan for home teaching if the child will be away from school for more than 2 weeks. Make certain someone will be available to care for the child at all times.



1 Adaptive equipment. Order devices such as wheelchairs in advance. Devices to help in management of children with casts are created by the families and are very effective such as this support that provides stable seating for his daughter in a spica cast (white arrow).



2 Preoperative trials. Sometimes it is useful to try splints in advance to prepare the child for standing after surgery.

Preparing the Child Prepare the child for the operation with a simple and honest explanation. Describe the procedures in terms appropriate to the child's or adolescent's age, and detail what he or she is likely to experience. Use the material in the reference section of this book and use models [2]. A teddy bear in a spica cast is a useful model for young patients. Let the child make choices wherever it is possible; for example, choosing the color of the cast. Arrange for the child to tour the hospital. All of these measures will help to reduce fear and to build a positive attitude toward the experience and the doctor.

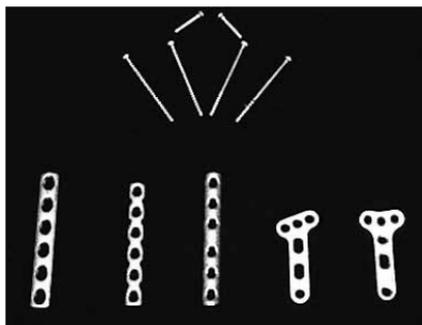
Special Operative Needs Be certain fixation, bank bone graft, special tools, or implants will be available [3].



1 Mobility home. This child will be traveling home by plane. Make arrangements for special seating in advance.



2 Preparing the child. Dolls are very useful in preparing the child for various types of treatment.



3 Special operative needs. Special fixation devices are often needed for children. Order bone graft material well in advance.

Anesthesia

Anesthesia in children has advanced dramatically during the past two decades. Subspecialization, new techniques, a focus on pain management and preparing children psychologically [1] provide improved care.

Preoperative Problems

Respiratory infections The average younger child has 4–5 upper respiratory infections per year. These infections complicate operative planning. Such infections pose operative hazards by increasing the risks of laryngospasm, bronchospasm, and coughing. Coughing during inductions increases the risks of regurgitation and aspiration. These problems can lead to a reduction in oxygen saturation during and after surgery.

- *Elective surgery* If the child has an upper respiratory infection, cancel surgery if the infant is less than a year of age, if signs of viremia or bacteremia are found, if scheduled for a long or complicated procedure, or if findings suggest a lower respiratory component is present.
- *Reschedule* Allow about 2 weeks after cessation of symptoms following a URI and 4–6 weeks after a lower respiratory infection.

Oral intake restrictions

- *Infants under 6 months* Allow feeding breast milk or formula to 6 hours before surgery and clear liquids to 3 hours before surgery.
- *Older infants and children* Allow feeding to 8 hours before surgery and clear liquids until 3 hours before surgery.



1 Well-prepared girl. This girl and her mother were well prepared for surgery.

Disease	Anesthetic concerns
Achondroplasia	Limited cervical spine mobility, restrictive airway disease
Arthrogyrosis	TM joint and C spine stiffness, GE reflex, postop airway obstruction, difficult IV access
Cerebral palsy	Gastroesophageal reflux, airway obst postop
Duchenne muscular dystrophy	Respiratory insufficiency, cardiomyopathy, malign. hyperthermia, hyperkalemia, hyperthermia, carditis, pulmonary dysfunction
JRA	TM joint ankylosis, cervical stiffness or instability

2 Anesthetic problems by disease. Childhood disorders carry certain anesthetic risks that should be understood before surgery.

Perioperative Fluid Management

Fluids Management usually requires replacement and maintenance. Maintain using a balance salt solution (BSS) such as lactated Ringer's solution.

Fluid requirement Calculate fluid requirements on the basis of 4 ml/kg/hr for the first 10 kg of body weight, plus 2 ml/kg/hr for the next 10 kg and 1 ml/kg/hr above that.

Estimated blood volume for children Calculate the EBV in ml/kg at 90 for newborns, 80 for infants in first year, and 70 for older children.

Indications for intraoperative blood replacement In the healthy child replace acute volume losses of 25–30%. The most reliable signs of hypovolemic shock in children are a tachycardia, diminished pulse pressure, and prolonged capillary refill time. Generally, replacement is indicated if the hematocrit drops below 21–25%.

Special Anesthesia Problems

Certain diseases carry special risk requiring special consideration [2 opposite page].

Meningomyelocele Start blood replacement early [1].

Myopathies This group of patients presents special risks. They should be referred to the anesthesiologist in advance of the procedure for an evaluation, which often includes an EKG, chest X-ray, and pulmonary function studies. These patients are prone to develop a malignant hyperthermia syndrome, cardiac rhythm dysfunction, and postoperative respiratory problems.

Cervical spine abnormalities Be concerned if the patient has dysproportionate dwarfism, Down syndrome, Goldenhar syndrome, Klippel–Feil syndrome, systemic JRA, or neck trauma. These patients should have preoperative evaluation and require special precautions especially during induction.

JRA Children may have TM joint ankylosis, cervical spine stiffness, and/or instability.

Latex allergy Latex allergy includes a spectrum of dermatitis, rhinitis, asthma, urticaria, bronchospasm, laryngeal edema, anaphylaxis, and interoperative cardiovascular collapse.

- **High risk patients** Include those with spina bifida, and those with repeated procedure and exposure to latex and who show varied allergic reactions.
- **Current status** Suspect and send for preoperative anesthesia consultation. Plan a latex-free surgical environment during the operation.



1 Kyphosis in myelodysplasia. Correction is difficult technically and blood loss can be substantial.

Anesthetic Risks

Anesthesia is generally a greater risk than most orthopedic procedures. Still, the risk of anesthesia is small. Fatal complications of anesthesia occur in 3–4 per 100,000 procedures. Most complications can be prevented by close monitoring, maintaining adequate oxygenation, and careful control of the level of anesthesia. Greater sophistication and cooperation between the surgeon and anesthesiologist [1] can reduce these risks. Complications include laryngeal and bronchospasm, aspiration, and cardiac arrhythmia and arrest.

General anesthesia is the standard for infants and children [2]. The induction technique is determined by the age of the child and the planned procedure. Rectal induction is used for some infants (over 6 months) and for simple procedures such as spica cast changes. Intervenuous induction is appropriate for the adolescent when the IV is placed before the procedure. In children, induction is often provided by inhalation anesthesia. The IV is then started quietly, with the child asleep. The preferred sites are hand or foot, antecubital veins, scalp, or external jugular vein. Avoid the femoral vein. In infants with small veins and abundant subcutaneous fat, IV insertion can be very difficult.

Regional anesthesia Occasionally, local or regional anesthesia will be used for upper extremity fracture management. This may be achieved by a hematoma block, intravenous anesthesia, or axillary or peripheral nerve blocks.

Sedation Some procedures are planned with sedation only and anesthesia ready if needed. This is suitable for early spica cast application for femoral shaft fractures, dressing changes, or minimally painful procedures.



1 Surgical team. Cooperation between the surgeon and anesthesiologist reduces risks.



2 General anesthesia. Provides excellent control of airway.

Blood Replacement

Order blood preoperatively if replacement is a reasonable possibility. Generally, major procedures done without a tourniquet may require replacement. Families are extremely concerned about the risks of AIDS when the possibility of transfusion is considered. The risk depends in part on the competence of the blood bank. Statistically, when transfused, the risk of receiving infected blood is roughly equivalent to the risk of receiving the anesthetic. Special situations may complicate blood replacement. Transfusions may be restricted by religious beliefs or the fear of AIDS. Minimizing blood loss may be necessary in difficult cases, such as spinal fusions and limb salvage procedures. Hypotensive anesthesia may be adequate. The mean blood pressure is maintained between 50 and 60 mm. Hypothermia is seldom indicated in orthopedics. Hemodilution is a technique by which blood is removed just prior to surgery and replaced at the end of the procedure. Finally, autologous blood donation and cell-saving techniques are becoming more widely available. Blood substitutes will be available in the future.

Postoperative Pain Management

Postoperative pain management can begin before the procedure with placement of an epidural catheter [1]. Epidural blocks are contraindicated when there is a need to monitor postoperative pain, as when a compartment syndrome is a possible postoperative problem. Problems with epidural blocks include itching and urinary retention [2]. Consider injecting marcaine into the wound edges at the end of the procedure to provide comfort in the early postoperative period. Patient-controlled analgesia (PCA) is a valuable technique for the child over 7 years of age. Morphine or meperidine are useful agents. Provide oral pain control by codeine or oxycodone.



1 Epidural anesthesia.

Epidural Anesthetic Complications

- Nausea, emesis
- Masking compartment syndrome
- Pressure sore – heel ulcer
- Urinary retention
- Prolonged hospitalization
- Itching

2 Epidural anesthetic complications. Consider these problems when making the anesthetic choice and when following the child after surgery.

Surgical Preparation

Positioning

Position for ease of access [1]. Use prone position for clubfoot surgery and for release of knee flexion deformity. Positioning at the end of the table when possible allows greater freedom to maneuver around the patient during various stages of the procedure. Positioning on the child's side allows procedures to be done from the front and back of the child without the need for redraping. If an intraoperative image is planned, place the X-ray cassette under the patient before draping.

Mark Sites for Skin Incisions

To minimize incisional length, consider marking the exact sites for surgical procedures using the imaging intensifier [2].

Skin Preparation

Shave if excessive hair is present [1 opposite page]. Perform a surgical prep. 1% iodine in alcohol is effective and efficient, but has no commercial promoters, so its use is seldom appreciated [2 opposite page]. We have used iodine successfully for thirty years. Apply one coat of the iodine solution and allow it to dry. This provides a sterile field and enhances the adhesion of the plastic film used for draping. For open wounds, a bacteriostatic soap solution is used. Shaving is usually not necessary in children. Shaving may cause superficial skin lacerations and irritation causing discomfort during the postoperative period.



1 Positioning. Thoughtful positioning facilitates surgery.



2 Mark the operative sites before draping. Marking the exact surgical site sometimes allows for the shortest possible operative incision.

Draping

Draping should provide adequate exposure for the surgical incision, a sterile barrier, and, if needed, free movement of the limb [3]. The margins, or the entire operative field, may be secured with an adhesive plastic film. Without clips, radiographs are less cluttered. Drape to provide wide exposure of the operative field. This allows the surgeon to extend the incision if more exposure is required.

For reconstructive procedures that require intraoperative alignment, drape one or both extremities free. This allows the surgeon to make certain the alignment is correct for lower limb alignment osteotomies, hip fusions, and similar procedures. Prep and drape both limbs, then use a sterile tourniquet.



1 Shaving. The need is minimal in children.



2 Surgical prep with iodine solution. Mechanical support may simplify the surgical prep.



3 Drape to allow full access. Plan for access well above and below the operative sites to allow intraoperative evaluation of joint motion.

Operative Scars

Plan the location and length of the incision as carefully as the fixation. The fixation is temporary, the scar permanent. Children are often embarrassed about their scars [1] even when they become adults. Not infrequently, scars from orthopedic procedures cause children to avoid sports like swimming, running, or basketball that require clothing that exposes the scar. Make the scar as inconspicuous as possible by limiting its length and making the incision in the least noticeable location that allows adequate access for the procedure. Try to avoid incisions that are known to cause bad scars.

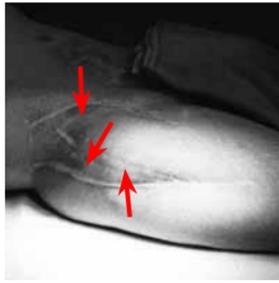
Minimizing Scars

Several techniques will be useful in reducing the disability from operative scars.

Shorten scars Make a short incision exactly over the site of the procedure. Before the skin prep, mark the position for the osteotomies using fluoroscopy.

Least conspicuous position Use the axillary approach for the upper humerus, the anterior approach for draining the hip, etc.

Avoid incision that cause wide scars For example, avoid making the vertical portion of the Smith–Peterson approach. To achieve the same exposure, extend the incision more medial in the *bikini line*. This provides adequate access with a much more cosmetic outcome. Likewise, avoid incisions over the clavicle. Make the approach below the clavicle even if a longer incision is necessary. Consider the anterior midline longitudinal incision for major knee procedures. The scar is more cosmetic, and if additional procedures are required later, the same approach can be used avoid multiple knee incisions.



1 Orthopedic scars. Operative scars may be unsightly and cause permanent embarrassment. Note the numerous scars on this boy's hip (red arrows). The improved appearance of the transverse scar on this girl's knee (green arrow) and the disfiguring scar (yellow arrow) on the adolescent girl. Every day she planned her clothing in an attempt to hide this scar.

Bilateral procedures Mark the position and scar length before preparing the skin [1]. Anticipate that the patient and family will compare the scars later. Asymmetrical scars for the same procedure creates doubt about the precision of the surgeon.

Short incision with mobilizing skin allows satisfactory exposure with minimal incision length [2].

Skin Closure

Close the skin with subcutaneous absorbable sutures [3]. Skin closure can be done quickly. Place a few subcutaneous sutures, and close the skin with subcuticular 3-0 absorbable suture. Supplement this closure with skin tape while approximating the skin edges by applying traction on both ends of the suture. The suture ends are cut off flush with the skin. This closure technique is rapid and definitive.

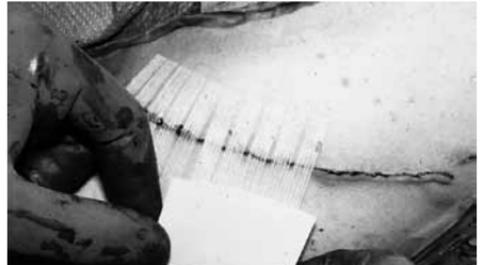
For incisions that are or will be under tension, close with 4-0 interrupted nylon sutures placed relatively close to the incision. Remove the sutures in 7 days. Avoid wide sutures to minimize the scar.



1 Mark position for incisions. Make symmetrical incisions for bilateral procedures. Whenever possible avoid scars over prominences such as malleoli or heel-cords.



2 Make minimal incision and mobilize. Often a short incision with subcutaneous skin mobilization will minimize the final scar length.



3 Subcuticular closure. Close the skin with absorbable subcuticular sutures. Supplement with skin tapes if necessary.

Fixation

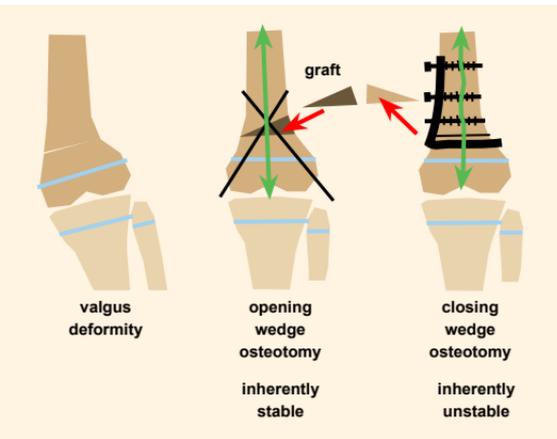
Fixation of fractures or osteotomies in children take many forms [1 opposite page]. The options of fixation in children are many. Take into consideration the child's age, location, inherent stability [1], and the likely time for healing. Internal fixation is usually necessary for bony procedures. When fixation is supplemented with a cast, the internal fixation need not be rigid. Often minimal fixation supplemented with a cast is adequate for children.

Plates

Plates have a limited role in fixation in children. Plates have inherent disadvantages. They require broad exposure, produce stress risers through end screws, and their removal is a major procedure. Plate fixation is useful for such procedures as the repair of congenital pseudarthrosis of the clavicle.

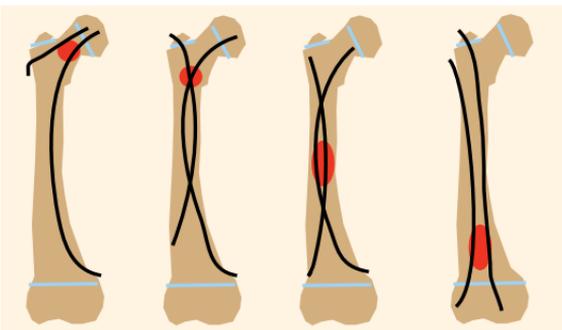
Intramedullary Fixation

Intramedullary fixation (IM) has many advantages in children. Flexible, small diameter, IM fixation is adequate for children, making reaming and large nails unnecessary. Adequate fixation is provided by pins, Rush rods, or special purpose devices. Because of their length and shape, IM rods seldom migrate long distances. Make certain the fixation extends well above and below the site to be fixed [2] and pins may traverse the growth plate.



1 Stability and osteotomy type.

Opening wedge osteotomies are more stable and require less rigid fixation than closing wedge procedures.



2 Intramedullary fixation by level.

The configuration of IM rods is in part determined by the level (red circle) of the lesion or fracture.

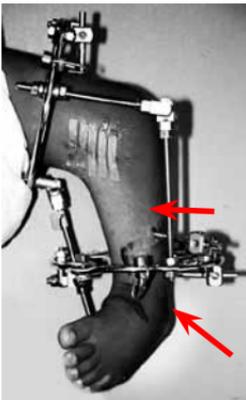
For tumors, plan to leave fixation until the lesion is healed. For conditions that permanently weaken bone, fixation is best left in place indefinitely. When pins remain for long periods, make certain the ends are deep enough to avoid skin irritation.

External Fixators

External fixators of a variety of types are suitable for children. Pins fixing bone may be stabilized externally with casts, frames, or special devices. They are used for stabilizing fractures and osteotomies, and for correcting deformities involving both bone and soft tissues. External fixators provide exceptional versatility, allowing changes in alignment, apposition, and length [2]. The disadvantages include the risk of pin tract infections, multiple scars, and the prolonged need for close medical attention.



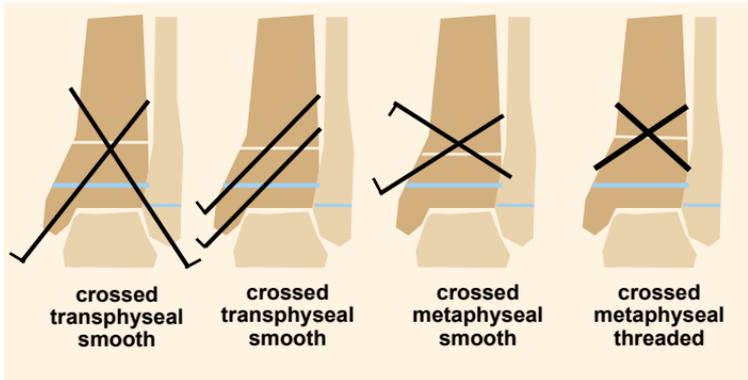
1 Varied fixation in children orthopedics.



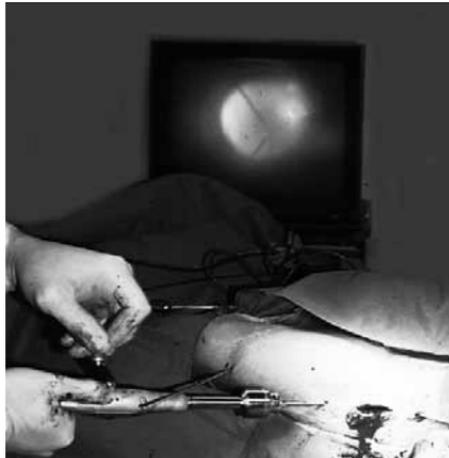
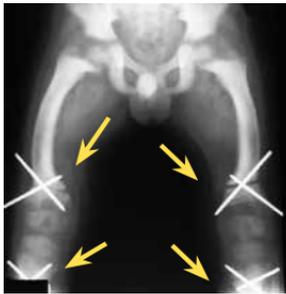
2 External fixation. These fixators are useful for correcting deformities in arthrogyposis (red arrow) or for the tibia following a release for a compartment syndrome (yellow arrow).

Pins

Pins can be for the orthopedist what nails are for the carpenter. Pins may be placed with varied configurations [1]. Pins are versatile, inexpensive, and rapidly applied and removed. Osteotomies fixed with crossed pins require small skin incisions [2]. Generally, smooth pins are most useful; they may traverse growth plates and are left outside the skin for removal in clinic. Threaded pins may be cut off just beyond the cortex, may not require removal, and should not be placed across the physis. Pins provide adequate fixation for bony procedures in nearly all infants, most children, and some adolescents. The absence of commercial promotion leaves the usefulness of pins often unappreciated.



1 Types of pin fixation. Varied uses of cross pins may be made based on the osteotomy level, age of patient, and indications for pin removal.



2 Cross pin fixation. Pins simplify fixation as a minimal incision is often adequate (red arrow) with the pins placed percutaneously. Usually crossed pin configuration is used as appropriate in rickets (yellow arrows) or for a double level osteotomy (green arrows) in Perthes disease.

Grafts

Tissue grafting involves autogenous and banked bone, fat, fascia, and cartilage [1]. Autologous organ transplants include bone, physéal plates, muscle, blood vessels, and nerves. Organ transfers require microsurgical techniques.

Autogenous Grafts

Bone Autografts are widely used, safe, rapidly incorporated, osteogenic, and readily available.

- *Local grafts* Harvest bone from the site of the primary procedure when possible. Calcaneal bone for subtalar fusions, bone iliac bone for acetabular shelf procedure, cranial bone for upper C spine fusions, etc.
- *Iliac grafts* Small amounts of bone can be removed percutaneously using a curette. Bicortical grafts in children are rapidly filled in.
- *Vascularized grafts* Complexity and donor site problems limit the usefulness of vascularized grafts of bone joints or growth w plates.

Soft tissue Free fat grafts are commonly used to replace defects in bone following physéal bridge resections.

Organ grafts Composite grafts are used to cover traumatic defects, for toe to finger transfers, etc.

Allografts

Bone Cadaver bone is convenient, carries a small risk of AIDS transmission, and incorporates more slowly than autogenous grafts. Such grafts are useful in procedures such as calcaneal lengthenings and bone replacement following resection of malignant tumors.

Osteochondral grafts are used for replacing joints for management of malignant tumors or trauma. The survival of cartilage is poor. Bone typing may improve results.



1 Allografts. These may be from a femoral head (arrow) or packaged commercially.



Postoperative Care

During the immediate postoperative period, the child is usually seen once or twice a day depending upon the magnitude of the procedure. Fortunately, most children have few postoperative problems and recovery is rapid.

Postoperative Orders

General orders include positioning, activity, vital signs circulation monitoring and other special considerations [1 and 2].

Fluid management A common error during and after surgery is prescribing too much fluid – especially dextrose in water. Most patients require little or no potassium because of tissue damage. Limit intake the first 24 hours after usual maintenance doses [1 opposite page].

Pain Management

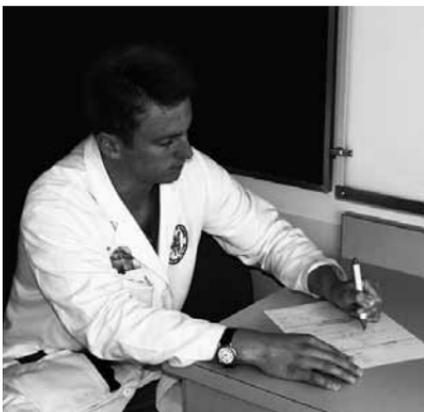
IV analgesia Administer morphine 0.1–0.2 mg/kg as a loading dose and continuous infusion of 0.01–0.03mg/kg/hr as necessary to maintain comfort [2 opposite page]. This is reduced by 10% every 12–24 hours.

Oral analgesia The patient is switched to oral analgesic when oral intake is allowed. Several agents are acceptable.

Patient controlled analgesia (PCA) In children older than about 5–6 years of age, this method is useful. Administer a loading dose of about 0.2 mg/kg. Allow doses of 0.02–0.03 mg/kg be given every 5–10 minutes with a maximum of 0.75–0.1 mg/kg per hour.

Epidural anesthesia This neuraxis analgesia is administered by the caudal route in children under years of age 6 or by lumbar route. Higher level administration may be necessary in selected cases.

Children with epidural pain management after surgery may be more comfortable, but be aware that epidural anesthesia may mask compartment syndromes.



1 Postoperative care. Work out a plan that is comprehensive and includes follow-up and home care.

Physician Inpatient Orders

- Position
- Vital signs
- Fluids
- Pain meds
- Other meds
- Intake
- Activity

Discharge Orders

- Follow-up
- Medication
- Activity
- Special equipment

2 Postoperative orders. Include these categories of needs.

Postoperative Problems

Fever Fever (temperature $>38^{\circ}\text{C}$) is seen in most children following surgery [3]. Be concerned if the fever is severe, the child looks more ill than expected, or if the child has positive physical findings suggesting a pulmonary or urinary problem.

Vomiting Nausea and vomiting following anesthesia are twice as common in children compared with adults. Causes of vomiting include agents such as nitrous oxide anesthesia and morphine. A history of motion sickness also increases the risk.

Special needs

Continuous passive motion (CPM) is a valuable technique in restoring motion [1 next page]. The excursion is set at the range achieved by the interoperative releases. Continue the CPM for about 6 weeks.

Splints or braces may be fitted and completed during hospitalization [1 on page 99].

Water (ml/kg/day) per body weight

First 10 kg	100
Second 10 kg	50
Each additional kg	20

1 Standard fluid maintenance for children.

Narcotic Medication

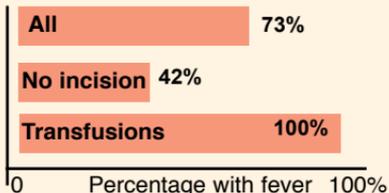
Usual Dosage

Acetaminophen + codeine elixir	Acetaminophen 15 mg/kg + Codeine 1 mg/kg/dose PO q 4–6 hrs
Acetaminophen + codeine tablets	1–2 tablets PO q 4–6 hrs prn
Morphine sulphate oral solution	0.3 mg/kg PO q 4 hrs prn
Acetaminophen (325 mg) + oxycodone (5 mg) <i>Percocet</i>	1–2 tablets PO q 4–6 hrs prn
Hydrocodone (5 mg) + acetaminophen (500 mg) <i>Vicodin</i>	1–2 tablets PO q 4–6 hrs prn

Anti-inflammatory Drugs

Ibuprofen <i>Motrin</i>	8 mg/kg PO q 6 hrs for 24–48 hrs
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2 Oral analgesic for children. From Ezaki (1998).



3 Factors associated with fever following orthopedic procedures in children. Fever is common following orthopedic procedures. Fever is related to the duration and magnitude of the procedure. From Angel et al. (1994).

Time for Discharge

Observing the child's general appearance and behavior are valuable methods of monitoring recovery. Be concerned if the child is not becoming progressively better. As the child becomes comfortable and smiling, the parent's concerns diminish. At this point, the child is ready for discharge.

Scheduling Follow-up Visits

Order the follow-up clinic visits thoughtfully. Make certain each visit has a specific purpose. Plan the postoperative visits to approximate the time when the child is at risk for a complication or when some change in management is anticipated. Most operative complications, such as infections, loss of reduction, or position or pressure sores will occur within the first week.

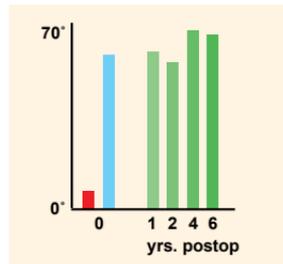
Time follow-up visits to coincide with timing for orthotic, physical therapy, or other postoperative visits. Being thoughtful about the family's resources will be appreciated.

Activity Status

Compliance with orders to limit activity is poor. If necessary ensure compliance by immobilization in a cast. Avoid burdening the family with the duty to enforce activity restriction; it is an impossible task and an unfair assignment.

Physical Therapy

Crutch training is best done preoperatively. Mobilize the child [2 nex page] when possible before discharge. Therapy is necessary for special situations such as for children with muscular dystrophy following contracture release procedures. Physical therapy is not necessary for routine postoperative care.



1 Continuous passive motion for knee stiffness. CPM is effective in restoring knee motion in children. Children with stiff knees had operative release and postoperative CPM. Preoperative motion (red) showed substantial improvement (blue) with maintenance of motion over the next several years (green). From Cole and Ehrlich (1997).

Hardware Removal

Generally smooth pins may be removed in the clinic. Often threaded pins and fixation hardware require removal under anesthesia. Most hardware removals are performed 3–9 months following surgery.

Indications In the past, removal of fixation devices was routine. As hardware requires an anesthetic, operative exposure, and sometimes added complications, routine removal is not appropriate. Indications for hardware removal include:

- *Prominent hardware* Hardware that alters the body contour and may cause discomfort such as proximal femoral fixation for varus osteotomies.
- *Fixation complicating known future procedures* Large fixation devices that are buried in bone about the hip may complicate total hip replacement.
- *Fixation causing stress risers* in the femur may require removal.
- *Hardware that extends into a joint.*
- *Metal reaction or infections* are relative indications.

Contraindications of hardware removal include:

- *Fixation that reduces the risk of pathologic fracture* should be permanent. This includes IM fixation of benign tumors, or fractures through osteopenic bone. This permanent fixation strengthens the bone and usually prevents refracture.
- *Fixation that is likely to be very difficult to remove* The difficulty of hardware removal is classically unappreciated. Complication rates can be significant. Make certain the benefit is worth the risks.



1 Making splints for postoperative care. The postoperative period in the hospital is an ideal time for fabrication of splints, orthotics, or adaptive equipment for home use.



2 Mobilizing the child following discharge. Make certain braces are constructed before the procedure (red arrow). Sometimes the child can be mobilized in the brace (yellow arrow) or with support.



Complications

Avoiding complications is a major operative objective. Orthopedists have been divided into *risk avoiders* and *risk acceptors*. Be a *risk avoider* when managing children. Complications are fewer in the pediatric age group because the child is more physiologically resilient. For example, thrombophlebitis and cardiopulmonary problems are uncommon in children. Complications sometimes cannot be prevented whereas others are secondary to poor technique [1]. Others are due to technical problems, such as inadequate correction, loss of position or fixation, or pressure sores from a tight cast.

It is usually possible to avoid complications if the risks are identified prior to the treatment and preventive measures to taken in advance. For example, when planning a proximal tibial osteotomy be aware that the procedure is associated with peroneal nerve palsies and compartment syndromes. These complications can often be prevented by (1) altering the level of osteotomy, performing rotational osteotomies in the distal rather than proximal level; (2) performing a prophylactic fasciotomy; (3) splitting the cast and, (4) careful postoperative monitoring.

As another example, when pinning a slipped capital femoral epiphysis, be aware of the risks of joint penetration and postfixation fracture. Fixing with a single central pin reduces the risk of joint penetration. Making the entry point proximal reduces the risk of postop fractures. Entry points at or below the lesser trochanter are associated with a risk of fracture through the site of pin penetration of the lateral cortex.

Other high-risk procedures include procedures using external fixation, or major procedures such as spine operations, extension osteotomies of the distal femur, unstable slipped capital epiphysis cases, procedures on myelodysplastic or poorly nourished children, etc.



1 Fractures around fixation. Sometimes the problem is preventable, as in this osteotomy, which was performed too low (red arrow). In others, the problem is not preventable, as in this bent rod in a child with osteogenesis imperfecta (yellow arrow).



2 Skin breakdown over foot. This skin breakdown was due to postoperative swelling.

Wound Infections

Wound infections are less common in children than adults. The risk of infection is greatest in operations of long duration; infections are most serious if the procedures involve bones and joints.

Prophylactic antibiotics can often prevent infection and should be administered intravenously at the beginning of the operative procedure. The single dose is adequate unless the procedure is prolonged.

Clinical signs of wound infections develop as early as 24 hours in streptococcal infections. The more common staphylococcal infections appear after 3-4 days. The child with a wound infection often shows systemic signs of fever and malaise, and the wound becomes more swollen, warm, and erythematous. The fever from infection must be differentiated from the common benign postoperative fever that commonly occurs after most procedures. Such fever resolves in a day or two. Be concerned if a secondary temperature elevation occurs. It should be considered a sign of an infection until proven otherwise.

If the child shows systemic signs of infection, search for the cause. Examine the ears and throat. Listen to the lungs. Window the cast and examine the wound. Perform a urinalysis. Culture the urine, blood, and wound. If the child is ill, start antibiotic therapy while the cultures are incubating. If the wound is infected, it should be opened, cultured, and drained under sterile conditions in the operating room. Close the wound after the infection is controlled.

Skin Problems

Skin problems may be due to irritation under the cast [2 opposite page] and is common in infants immobilized for hip dysplasia [2]. Prevent skin problems by keeping casts as dry as possible. The inflammation resolves once the cast is removed.



1 Tourniquet burns. This skin breakdown occurred under a tourniquet.



2 Skin irritation under a spica cast. This severe irritation is due to poor home care.

Tourniquet Irritation

Tourniquet "burns" [1 previous page] can usually be prevented by avoiding prep solution from seeping under the tourniquet. Treat as a burn.

Pressure Sores

Pressure sores are most common in children with neuromuscular problems. Anesthetic skin and the child's difficulty in communicating are major contributing factors. Take added precautions in patients with myelodysplasia and cerebral palsy. Pressure sores occur in characteristic locations: the heels [1], trochanters, sacrum, and other bony prominences [2]. They can usually be prevented by careful casting technique. Apply thick padding, avoid a snug cast, and window the cast over the heels if necessary. During the postoperative period, rotate the patient frequently, and inspect areas at risk, such as the sacrum, often. In communicative patients with intact sensation, ask about localized pain, such as over the lateral malleolus. Cast pressure pain is often described as a burning pain. Detecting pressure sores is sometimes difficult. Be concerned if a foul odor is present. The stench of devitalized tissue is different from the usual fecal-urine odor. *Sniffing* the cast or wound is a simple and effective test. Operative related pressure sores will heal if the wound is kept clean and protected from pressure [1 opposite page] or abrasion. Identify pressure sores early so they can be healed by the time the cast is removed.

Stiffness

Joint stiffness as an operative complication is uncommon in children. Simple postoperative stiffness is temporary and resolves as the child resumes activity. This makes physical therapy unnecessary. Persistent stiffness usually results from joint damage due to compression, ischemia, or infection. Manage joint stiffness by active range-of-motion exercises and observation. If improvement plateaus and the stiffness produces disability, consider performing an arthrolysis and employing continuous passive motion or special splints to maintain the correction gained. Generally expect to retain about half of the range of motion obtained intraoperatively.



1 Heel ulcer. This ulcer is due to excessive pressure from the cast.



2 Skin breakdown in cerebral palsy. These are common sites of skin breakdown under the spica cast in a child with cerebral palsy.

Pin Migration

Smooth pins may migrate long distances in the body. They have been found in the mediastinum and in the heart. Migration is best prevented by bending the end of the pin. Bend the protruding end of the pin to a right angle and cut the pin off about 1 cm from the bend.

Pin Tract Infections

Pin tract infections are usually due to motion around the pin, or tension on the adjacent soft tissue [2], producing necrosis. Both of these problems are usually preventable. After placing the pin, if the skin is tented, incise the skin to relieve the tension. Stabilize the pin-skin junction. Immobilize the limb in a cast. Pin tract infections should be cultured and treated with antibiotics. Open drainage is seldom necessary.

Cast Syndrome

The cast syndrome, or superior mesenteric artery syndrome refers to a spectrum of disorders caused by compression of the second portion of the duodenum between the superior mesenteric artery and aorta. This is sometimes referred to as the "nutcracker effect." The clinical manifestations vary from partial duodenal obstruction to bowel infarction. Predisposing factors include hyperextension of the trunk, supine positioning, and poor nutritional status. Each tends to increase the nutcracker-like squeeze of the artery and aorta on the duodenum. This syndrome may develop in a hyperextended body cast or prolonged supine positioning. Treat by removing the cast, prone positioning, and increased caloric intake.



1 Skin breakdown in myelodysplasia. The heavily padded cast protects the skin and allows healing.



2 Pin tract infection. This infection around a percutaneous pin was due to excessive tension on the skin.

Motor Regression

Most children experience some temporary motor regression following immobilization after operative procedures. In children with neuromuscular disorders, the regression is much more profound [1]. In severely disabled adolescents, recovery may be incomplete, and full return to the preoperative motor level never achieved. For example, the adolescent with cerebral palsy who is a marginal walker preoperatively may not return to walking after a long recovery from a triple arthrodesis, hip, or spine procedure. Regression can be minimized by upright positioning, an active exercise program, and shortening the period of immobility.

Compartment Syndromes

Compartment syndromes, or ischemia from tight casts [2] should be promptly diagnosed. Following any major procedure distal to the elbow or knee, the cast should be prophylactically bivalved and spread. Perform a prophylactic anterior and lateral compartment release whenever a proximal tibial osteotomy is performed.

Pathological Fractures

Postoperative pathologic fractures are most common in children with reduced sensitivity and flaccid paralysis. The risks are greatest in the child with myelodysplasia [1 opposite page]. Fractures occur most commonly at the distal femoral level following cast removal. These fractures are difficult to prevent. Minimize the period of immobilization, load the limb by standing the child in the cast, and use special caution in applying physical therapy after cast removal.



1 Motor regression. This child lost the ability to walk following surgery. Recovery took many months.



2 Volkmann ischaemic contracture. This complication resulted from management of supracondylar fracture of the humerus.

Deep Vein Thrombosis

Deep vein thrombosis is most common in spinal surgery, spinal trauma with paralysis, and in children with predispositions such as those with protein C deficiency, vascular malformations, etc. External compression devices on limbs during surgery reduce the risk.

Toxic Shock Syndrome

Toxic shock syndrome (TSS) is a rare but catastrophic complication that occurs usually 2–3 weeks following surgery. TSS is a reaction to toxins from staphylococcal and streptococcal infections. About half occur in menstruating girls. Sudden onset of fever, vomiting, diarrhea, rash, and hypotension are common. Multisystem organ failure may occur. Fatality rates in orthopedic patients are about 25%.

Hypertension

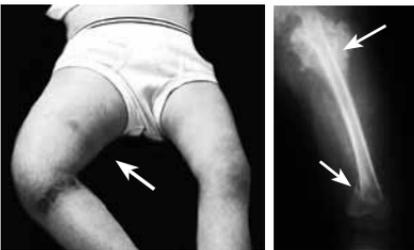
Hypertension is common following certain orthopedic procedures that cause stretching or lengthening of extremities. In these procedures monitor the child's blood pressure.

Avascular necrosis

Avascular necrosis (AVN) is a serious complication that may follow treatment of DDH [2], acute slips, traumatic dislocated hips, displaced transcervical femoral fractures, lateral condylar fractures, radial neck fractures, and other problems. With this known risk, warn the parents and document in the chart an awareness of the risk and measures to avoid AVN before undertaking management. In many cases no effective prevention is available. In others, such as for lateral condylar fractures, careful operative technique avoiding excessive soft tissue dissection may prevent the complication.

Arterial Injury

Injuries to veins and small arteries can usually be controlled by pressure and time. In contrast, major arterial injuries may be limb-threatening. Unless you are skilled in vascular repair, ask a colleague with this expertise for assistance. If excessive pulsatile bleeding occurs, control by local pressure, pack the wound, and wait several minutes. While waiting, improve exposure, optimize the lighting, and have suction ready. If bleeding continues, the injured vessel can usually be found and ligated. For more severe injuries, proximal control and arterial repair may be necessary. Arteriography may delay the repair. Less common are aneurysms following operative procedures [1 next page]. Aneurysms may become evident weeks or months postoperatively.



1 Pathological fractures in myelodysplasia. These fractures causes extensive soft tissue swelling and callus formation.



2 Partial AVN complicating DDH management. This type 2 AVN causes a deformity of the upper femur.

Bad Scars

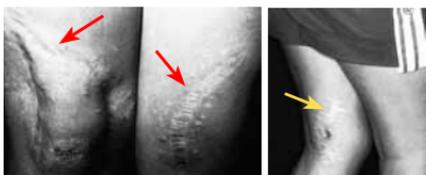
Ugly scars are far too common following procedures in children [2 and 3]. Poorly placed, excessively long and sloppily closed operative scars last a lifetime. Bad scars embarrass children, limit their selection of clothing and restrict activities. Most are preventable.

Lack of Compliance

Children often exceed limits placed on postoperative activities. Families may not return for scheduled clinic visits. Often the child or family is blamed for the complication; however, it may be due to poor medical care [4]. Take precautions based on the assumption that the child will do whatever is possible. Be creative. Use fixation unlikely to cause problems with removal. Make casts excessively strong or activity-inhibiting by design. Employ a *tickler system* to automatically generate recall action if families miss appointments.



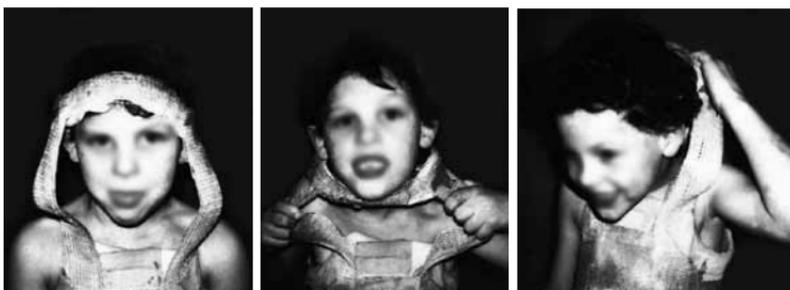
1 Aneurysm of femoral artery. The artery was injured during an operative procedure.



2 Ugly scar about the knee. Note the bad scars in front of the knee from patellar realignment (red arrows) and behind the knee (yellow arrow) from a hamstring lengthening procedure.



3 Note the unnecessarily long scars from femoral osteotomies.



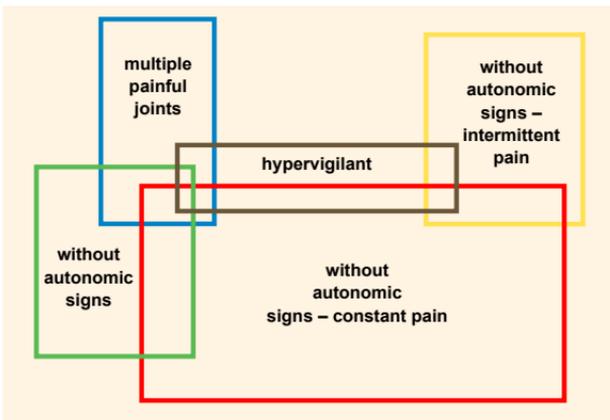
4 Flimsy casts are no match for children. This cast was applied to hold the head in a neutral position following torticollis surgery. It was inadequate and quickly broke down. The child was not *noncompliant*, he was simply behaving like a normal child.

Amplified Musculoskeletal Pain Syndromes

Included in this category of syndromes include reflex sympathetic dystrophy (RSD) or reflex neurovascular dystrophy (RND), idiopathic pain syndrome, and fibromyalgia.

Scope

These pain syndromes are varied and may be associated with autonomic signs [2]. The patients typically present features of disability out of proportion to the trauma history or clinical findings. These patients are often seen first by the orthopedist because the pain is musculoskeletal and frequently follows minor injury.



1 Common features of amplified pain syndrome. Based on Sherry (2000).

Common Features of Amplified Pain Syndrome

- Most common in preadolescent or adolescent girls
- Increasing pain after minimal or no trauma
- Significant disability
- Crawls around house or on stairs
- Discomfort with light touch – clothing, bed sheets, etc.
- Autonomic changes – cold, color, clammy, edema
- Worse or not better with cast immobilization
- Unsuccessful previous treatment
- High-level athletes or dancers
- Personality features – mature, excels at school, perfectionistic, etc.
- Recent major life change – move, school, friends, divorce, etc.
- Mother speaks for the child
- Incongruent affect for degree of pain or disability
- La belle indifference about pain
- Compliant when asked to use the limb
- Autonomic signs especially after use
- Pain not restricted to dermatome or peripheral nerve distribution
- Negative neurological examination

2 Overlap of pain amplification syndromes. Note the varied patterns. From Sherry (2000).

Diagnosis

The patients present with a wide variety of clinical features [1 previous page]. The findings may show considerable variations from autonomic features [1] to dynamic or fixed deformity [2]. Usually the evaluation elicits a sense of disparity. The pain or disability is exaggerated beyond any signs of underlying disease.

Management

These patients are difficult to manage. The psychological or functional underlying problem is often clear, but the family will often be offended if that possibility is presented as the primary problem.

Referral When available, consider referring the child to a pediatric rheumatologist to manage care.

Active treatment is usually successful. Order functional aerobic training using the involved limbs such as drills, running, play activities and swimming for a period of 5 hours daily. Desensitize skin with towel rubbing. Refer for psychological evaluation and provide psychotherapy as appropriate. This intensive treatment may require inpatient care with a follow-up home program for another month.

Outcome 80% cured, 15% improved, 5% unimproved; relapse 15%; 90% doing well at 5 years.



1 Autonomic features of right foot. Note the discoloration and swelling of the leg and foot (white arrow) and increased uptake on bone scan (blue arrow).



2 Severe fixed deformity from RSD. This 15-year-old girl developed a fixed equinovarus deformity of the foot (red arrow) over a period of many months. Correction requires soft tissue releases and casting (yellow arrow).

Traction

Traction still has a role in management. Although less than in the past, specific indications have replaced standard management.

Common Indications for Traction

Temporary stabilization Skin traction is commonly used for femoral shaft fractures before cast immobilization or operative fixation and for preoperative management of unstable proximal femoral epiphyseal slips.

Home traction Home traction programs have been used for preliminary traction in DDH management [1] and in home management of femoral shaft fractures in young children.

Fracture management The common uses of traction include supracondylar humeral fractures, femoral shaft [2] and subtrochanteric fractures.

Overcoming contracture Traction is sometimes used to improve motion in Perthe's disease or chondrolysis of the hip. Whether the improvement is due to the traction or simply the enforced bed rest and immobilization is unclear.

Spine problems Complex spine problems such as congenital and neuromuscular deformities are sometimes managed by a combination of traction and surgery.

Traction Cautions

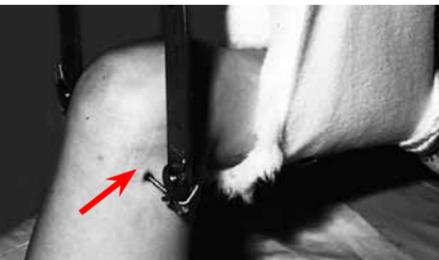
Inflammatory hip disorders Avoid traction in inflammatory hip disorders such as toxic synovitis or septic arthritis. Traction often positions the limb in less flexion, external rotation, and abduction, resulting in increased intraarticular pressure and possible avascular necrosis.

Overhead leg position Avoid Bryant traction in patients weighing more than 25 pounds as the vertical positioning may result in limb ischaemia. This traction is seldom used today. It is preferable to reduce the flexion of the hips from 90° to about 45° to reduce the risk of ischaemia.

Proximal tibial pin traction Avoid proximal tibial skeletal traction as distal femoral traction [2] provides greater safety. Reports of recurvatum deformity and knee ligamentous laxity make this treatment risky.



1 Skin traction for DDH. Sometimes prereluction skin traction is used to overcome contractures to facilitate reduction and possibly reduce the risk of AVN.



2 Distal femoral traction. This is the preferable site for traction when treating femoral shaft fractures.

Complications of Traction

Skin irritation This is common under skin traction [1]. Prevent this problem by avoiding excessive traction or compression. Frequent inspection of the skin reduces this risk.

Nerve compression This most commonly involves the peroneal nerve [2] from skin traction. Avoid excessive pressure over the upper fibula.

Vascular compromise This complication is most commonly associated with overhead traction for femoral fractures in infants over 25 lbs.

Physeal damage from pins This complication has been most common from upper tibial pin traction.

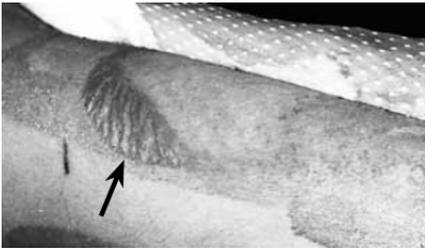
Cranial penetration of halo pins The thin calvarium in children makes inadvertent penetration a risk [3]. The risks are reduced by using more pins with less compression. Preapplication CT studies may be helpful in determining proper sites for pin placement.

Superior mesenteric artery syndrome This serious complication may result from long periods of supine positioning in poorly nourished individuals.

Hypertension The mechanism is unknown.

Procedures

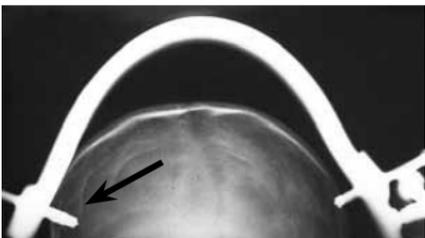
Traction procedures are detailed on pages xxx and xxx.



1 Skin irritation under traction tapes. This skin blister is due to excessive pressure and traction.



2 Peroneal nerve palsy. The excessive pressure resulted in a loss of function.



3 Halo pin penetration. This is prevented by using multiple pins and limiting penetration pressure during application.

Casting

Casting is useful for immobilization, control of position, correction of deformity, and sometimes to ensure compliance with treatment. Cast treatment is relatively safe, inexpensive, and well tolerated by children. Casts may be made of plaster or fiberglass. Plaster casts are least expensive and readily molded. Fiberglass casts are expensive, lightweight, water-resistant, radiographically transparent, and less messy and provide many color and decorating options [1]. Sometimes the materials are combined in treating deformities such as clubfeet.

Categories of Casts

Casts are remarkably versatile and take many forms. They may be circumferential or applied as splints.

Cast Problems for Children

When applying casts for children, keep in mind the unique problems that may be encountered.

Compliance Children are less compliant than adults. They may not hold still for cast application, may allow their cast to become wet, or damage their cast in play.

Communication Infants or young children may not be able to communicate the pain that precedes the development of pressure sores over bony prominences.

Sensation The child with myelodysplasia or cerebral palsy has poor sensation and is at risk for pressure sores.



1 Colorful casts. Allowing the child to choose the color makes the experience less threatening. Casts can be decorated.



2 Padding. Hold the desired position the same throughout the casting process. Apply the stockinette then the padding.

Cast Application

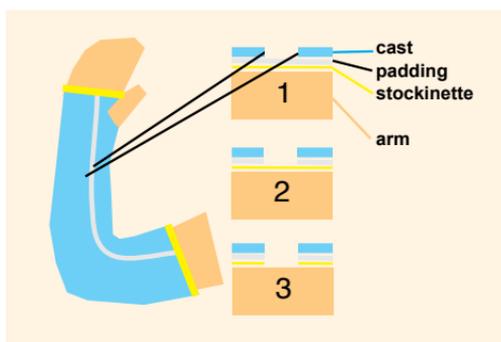
Positioning First, make certain the child is comfortable and the limb is held in the position desired after the cast is completed. For cylinder casts or body jackets, the child should be standing. For long-leg casts, it is helpful to apply the short-leg section first; after it has hardened, extend it to the thigh. Include the toes in children's casts to provide protection. Apply the cast only when the patient is comfortable and the limb immobile. Make certain the assistant holding the limb maintains the proper position until the cast has hardened.

Padding Apply at least two layers of padding [2 previous page]. The first is the tubular stockinette that allows a neat trim for the cast edges. The material is usually cotton or dacron. The second layer is the padding. Apply extra padding over bony prominences, if the child is likely to move during cast application, or if the child is at risk for pressure sores.

In applying the cast [1], start at one end and proceed in an orderly fashion to the other end of the cast. Apply with a 50% overlap by rolling rather than pulling on the material. The techniques for application of plaster versus fiberglass are different. Tucks are taken in plaster casts to make a neat application.



1 Cast application. Apply the first layers, turn the stockinette back for trim. Apply the final layer to trim the cast and add the desired color.



2 Stages of cast splitting. Casts can be split to different levels. Note that in cross-section only the cast is divided (1); cast and padding are divided (2); or all layers are divided (3).



3 Cast technique with fiberglass. Unroll a length and then lay the material down without tension.

Fiberglass application Fiberglass rolls must be guided to maintain control of direction. *When applying fiberglass, free a segment of material from the roll, the apply it smoothly and without tension* [3 opposite page]. Plaster has a definite time of crystallization and hardens rather abruptly. Fiberglass hardens slowly. The ideal thickness of most casts is three layers. Apply extra layers over sites of greater stress, such as the hip in spica casts or the knee and ankle in long-leg casts.

Early Cast Care

Bivalving Bivalving or splinting casts may be by degree [2 opposite page]. Be aware that padding is often not elastic and may create as much compression as cast material. For complete relief of pressure, it is necessary to divide all layers of the cast on both sides.

Pressure relief If sensation is poor or communication limited, consider relieving pressure over bony prominences. Cut a rectangle of cast or cut a X over the site for relief. Elevate the cast edges and leave the padding intact. To make the child in a spica cast more comfortable, consider flaring the thoracic edges and creating a *stomach hole* [1].

Trim cast edges To save operative room time, consider trimming the cast in the recovery room or on the ward. Provide a generous amount of space around the perineum.

Cast Care

When the child bathes or plays in the rain, cover the cast with a plastic bag to keep it dry. Even fiberglass casts are uncomfortable when wet. In infants, spica casts pose a special problem. Instruct staff and parents to change the infant's diapers frequently and to avoid tucking the diaper under the cast. Skin irritation is best managed by exposure to air and light. Avoid criticizing the child for the appearance of the cast. Often a worn cast is evidence of success in incorporating the treatment in the play activity of the child [2].



1 Spica cast. Note the large abdominal window (red arrow) and extra space around the thorax (yellow arrow).



2 "Well-used" cast. Ideally the child will be very active in the cast.

Cast Removal

Cast removal is often the most risky phase of cast treatment [1]. Cast saws can cut the skin if the contact is made under pressure. Cast saw lacerations are most likely over bony prominences such as the malleoli. Plaster casts may be soaked off by the parents prior to clinic. The crying, struggling child is at special risk. Try to reassure the child by placing the moving blade gently against your arm to show that it only vibrates and normally does not cut skin. Compare the saw noise to an airplane. Have the mother comfort the child as well.

Avoid dragging the saw; use consecutive in-and-out movements to cut the cast. Try to avoid cutting directly over the bony prominences. Insist that the inexperienced assistant learn to remove casts on adolescents or adults and not infants or children.

Hair grows more rapidly under casts. The adolescent girl is often shocked by the amount of hair on her leg following cast removal. Reassure her that in a month or so the hair growth will return to normal.

Orthotics

Orthoses are used to control alignment, facilitate function, and provide protection. They include braces and splints [1 opposite page]. Often the distinction between braces and splints is poorly defined.

Splints provide static support or positioning and often encompass only half of the limb. They are often worn only part-time.

Braces are usually more elaborate and worn while the child is active [2 opposite page]. Braces are sometimes divided into passive and active types.

- *Passive braces* are those that simply provide support such as some scoliosis braces in children with neuromuscular disorders.
- *Active Braces* are those that facilitate function. Such braces may promote active correction as seen in scoliosis braces that incorporate pads.



1 Cast removal. Reassure the child by applying the moving blade to the hand (upper left). Cut the cast with in and out movements (upper right). Cut both sides of the cast, spread the cut edges, and divide the padding with a sissors. This allows the cast to be removed.

Goals

Be realistic about the goals of bracing. Bracing will not correct static deformity or scoliosis. At best, braces prevent progression. Orthotics do not correct physiologic flatfeet or torsional deformity. Although radiographs taken with the orthotics in place may show improvement, this correction is not maintained after the brace is removed. Unbraced radiographs can be made to assess real correction.

Naming Orthoses

The name of the device is determined by which joints are involved. An AFO includes the ankle and foot; a KFO adds the knee; a HKFO includes the hip, knee, and ankle. Special braces are often named by city of origin.

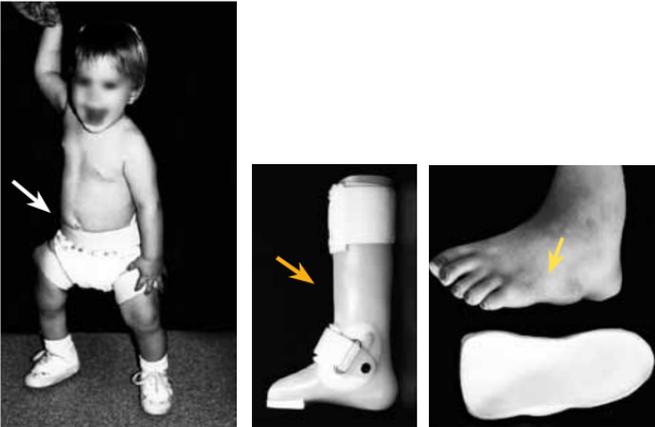
Ordering Orthoses

The prescription should include several components: the extent, material, joint characteristics, and closure types [1 next page]. Order orthoses thoughtfully as any orthoses is a burden for the child.

Minimizing the Orthotic Burden

Attempt to reduce the burden to the child.

Effective? Many orthoses are ineffective and should not be used. Examples include all orthoses for developmental deformities that occur in normal children. These include orthoses for flatfeet, twister cables for torsional problems, or wedges for bowlegs.



1 Common orthotics. Hip abduction (white arrow), foot orthosis (yellow arrow), and ankle-foot orthosis (orange arrow) are most common braces.



2 Functional bracing. This child with arthrogyposis has special braces that incorporate light weight, heel elevations, knee flexion, and internal rotation components.

Perform the child test For children with neuromuscular problems, orthoses such as AFOs are frequently ordered to improve function. If the brace truly improves function, the child will usually prefer to use the brace. If the braces causes more trouble than benefit, the child will prefer to go without. Make certain the brace is comfortable and fitting properly. If the child prefers to go without a comfortable, well-fitting orthoses, it generally means that the brace is a functional liability. In most cases the unwanted brace should be discontinued.

Minimum duration Duration of bracing is critical to success and acceptance. The effectiveness of bracing to arrest progression of a deformity depends upon two factors: the amount of corrective force applied and the duration this force is applied (based on a 24-hour day). The effectiveness of bracing increases with duration. The psychological and physiological costs also increase with duration. Balancing the benefit and cost is a challenge. Nighttime bracing is least "costly" for the patient because bracing does not interfere with play, is convenient to use, and causes little effect on the child's self-image. The duration of bracing can vary from full-time (allow an hour free), to nighttime, or part-time. Part-time bracing is commonly ordered for 4-, 8-, or 12-hour periods per day. Negotiate with the child to make certain that the precious free hours coincide with the child's priorities, such as school or specific athletic or social activities. This will improve compliance.

Minimal length The longer the brace the greater the disability. Extending braces to the pelvis is seldom necessary. Likewise shoe lifts for leg length inequality may be prescribed that are less than needed to completely level the pelvis. Usually allowing up to 2 cm undercorrection is acceptable to reduce the weight, instability, and unsightly appearance of a higher lift.

Prescribing Orthoses

Length

- AFO
- KFO
- HKFO

Material

- Molded
- Leather
- Polypropylene

Hinges

- Free
- Right angle stop
- Spring loaded



Upright

- Single – Double
- Steel – Aluminum



Closures

- Velcro
- Buckles

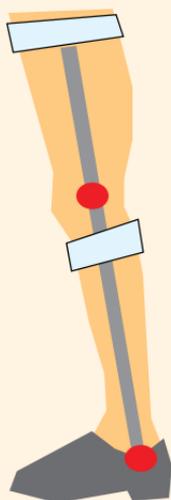


Special Features

- Stress – valgus or varus
- Pads – location

1 Orthosis

prescription. Specify each element of the brace.



Prosthetics

Prostheses are artificial substitutes for body parts. Most prostheses in children are designed to replace limb deficiencies secondary to congenital, traumatic, or neoplastic problems.

Naming Prostheses

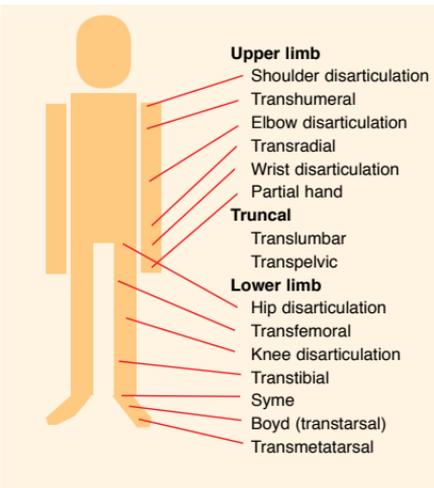
Name the prosthesis based on the level of the deficiency or type of amputation [1].

Prescribing Prosthesis

Detail each element of the limb [2].

Special Needs of Children

Children have special prosthetic needs. Children grow, making prosthetic adjustments necessary 3–4 times a year. The prosthesis must be rugged and simple in design. Because multiple limb deficiencies occur in up to 30% of congenital losses and 15% of acquired losses, customized prosthetic management is often necessary.



1 Amputation levels.

Upper Limb Prosthesis

Type

- Infant
- Child

Harnessing

Control Motion

Terminal Device

Components

- Wrist
- Elbow
- Shoulder

Motor

- Body
- Myoelectric

Special Features

- Partial amputations

Lower Limb Prosthesis

Type

- Immediate
- Early
- Preparatory
- Definitive

Design

- Endoskeletal
- Exoskeletal

Suspension

- Transfemoral
- Transtibial

Socket Liners

Components

- Knee
- Ankle
- Foot

Special Features

- Compensate deformity
- Include foot

2 Prostheses

prescription. When ordering prostheses describe each of these features.

Age for Fitting

Lower limb Fit lower limb prosthetics when the child first pulls up to stand, about 10 months. Initially the knee may be omitted to keep the limb simple, light, and stable [1]. Delay bilateral amputee fitting a few months.

Upper limb Fitting upper limb deficiencies is controversial. Some fit at about 6 months of age. Others prefer to wait until a need is recognized by the child, which usually occurs in mid-childhood.

Acceptance

Lower limb prostheses are well accepted as they clearly enhance function and appearance [2]. Stability and symmetry required for walking are readily provided by the prosthesis.

Upper limb prostheses are less well accepted. Some find the artificial limb to be a burden without sufficient compensation in improved function to justify the trouble. The lack of sensibility limits function. Children learn to function well with one hand. Children seldom use the prehensile function of upper limb terminal devices. Cosmetic hands are useful in adolescence.

Myoelectric Power

Powered limbs have the advantage of slightly improving appearance but the disadvantages of being more complex, heavier, and slower. The results are mixed.



1 Toddler with nonarticulated prosthesis. First prosthesis.



2 High below knee prosthesis. This boy with tibial deficiency has a short stump with strong quads. He is fully active in sports.

Therapy

Therapy utilizes the treatment methods of physical medicine, including manipulations, exercises [1], positioning, stimulation, massage, and application of cold and heat. The role of the pediatric therapist is much broader than that of the general therapist, requiring knowledge of growth and development [2].

The current emphasis on function has improved the effectiveness of therapy programs. Emphasis on effective mobility, independence skills, and communication focuses therapeutic energy and resources on an outcome that optimizes the child's quality of life.

Physical Therapy

In pediatric orthopedics, the primary focus of physical therapy is on lower limb function and mobility.



1 Hydrotherapy. This child with arthrogryposis with knee flexion contractures is given her first walking experience.

Expanded role of the pediatric therapist

- Assesses function
- Educates family
- Provides psychological support for the family
- Explores uses of adaptive equipment
- Documents management and research

2 Role of pediatric therapist. The therapist's role is considerably broader than simply providing exercises and manipulation.

Effective mobility A child needs independent and efficient mobility [1]. Without this capability, the child's psychosocial and educational experiences are significantly limited. The level of mobility should be appropriate to the child's mental age. The method of mobility is not critical.

Whether mobility is provided by walking or by the use of adaptive equipment [2], the method of mobility should be manageable by the child himself, conserve the child's energy, and be functionally practical. Options range from the use of an electric wheelchair [3] to unassisted walking. The objective of management is to provide effective mobility by whatever means necessary while helping the child progress toward a realistic mobility objective. The objective should be an optimistic target within a realistic range. The therapist's accurate appraisal of the child, knowledge of mobility potential for the disease, and periodic assessments of progress help protect the child from disappointment, frustration, and wasted efforts. With time, objectives may change, depending upon the rate of progress.

A major objective of therapy is support and education of the family. Often the family has unrealistic expectations that create an additional burden for the child. The family's major concern is often "Will my child walk?" A better objective would be "Will my child be independent and happy?" Assisting the family and guiding their concepts and expectations is an important role for the therapist.



Objective	Device
Mobility	Walkers Wheelchairs Motorized vehicles
Self-Care	Lifts Ramps Special toilets
Communication	Communication boards Computers

1 Effective mobility. The mobility of this bright child with severe arthrogryposis is provided by an electric wheelchair.



2 Integration. Mainstreaming or integration into a normal environment is very helpful for social development.



3 Effective mobility. This electric wheelchair allows this child self determined mobility.

Infant stimulation programs Helping the parents to interact positively with the child is a vital role of therapy. Parents may be uncomfortable with the infant, and this strained relationship further limits the child. Interactive play therapy, taught to the parents [1] by the therapist, provides the positive physical contact infants need for optimal emotional and intellectual growth. Infant stimulation programs are effective in promoting cognitive, motor, language, emotional development.

Neurodevelopmental therapy Neurodevelopment therapy (NDT) focuses on motor development. NDT is more effective than the original passive treatment methods but is being replaced by therapy with a broader focus.

Accepting disability Accepting the disability and working around it, using adaptive equipment, is often the most effective management strategy for the child. Usually the physician or therapist cannot cure the disease, but can minimize the disability.

Adaptive equipment is useful to help the child become more independent and functional. Adaptive equipment is useful for the child’s mobility, self-care skills, and communication, and often enhances care of the child by the caregiver.



1 Infant stimulation. This infant is provided play experience which is necessary for intellectual development

Form	Indication
Isotonic	Contraction through an arc
Isometric	Static contraction
Active Range of Motion	Maximum range of motion by patient unassisted
Assistive Range	Therapist-assisted maximum range of motion

2 Forms of exercises for children.

Several forms of exercise are available. Passive exercise that causes pain is contraindicated in children, as it often increases stiffness by injuring the joint.



3 Stretching. Stretching exercises should be done carefully to avoid fractures and causing pain.

Exercises are not very useful for the young child because the child lacks the interest and discipline to perform the exercises. Fortunately, children have little need for exercises, as muscle strength and function usually recover spontaneously. Moreover, assistive or stretching exercises can be harmful. In posttraumatic stiffness, stretching often increases stiffness by adding new injury and scarring. Exercise should not be painful. Exercises take on a variety of forms [2 previous page]. Chronic passive motion is a new technique for maintaining joint motion following operative release or injury. The joint is moved slowly and continuously through a range of motion during healing.

Stretching is a traditional treatment for contracture [3 previous page]. Flaccid contractures respond best to stretching. The prolonged effects of spasticity, as in cerebral palsy, cannot be controlled by intermittent stretching. To prevent contracture, the elongated or stretched position must be maintained for about 4 hours in each 24-hour day. This requires bracing or splinting. Stretching beyond the child's pain threshold is not advisable; overstretching causes further injury and scar formation.

Therapy at home, with a parent acting as therapist and the therapist as a consultant, is effective and practical when the family is willing and able. Home therapy programs reduce stress on the family by making the treatment more convenient and less expensive. The therapy can often be incorporated into the daily routines, increasing frequency and improving outcomes. Home therapy may also have a bonding effect on the family. This requires parent education and periodic visits to the therapist to assess technique and progress.



1 Occupational therapy. Teaching families how to provide play activities for each child is essential for development.



2 Adaptive equipment. Selection or development of special devices to facilitate activities of daily living is very valuable in developing independence.

Treatments of doubtful value include massage, thermotherapy, injections, and diathermy. These “treatments” are not helpful in pediatric orthopedics.

Occupational Therapy

Occupational therapy focuses on upper extremity function [1 opposite page] and activities of daily living [2 opposite page], including independence skills and correction of deformity [1]. This aspect of therapy plays a broad role in managing childhood disabilities because modern management places greater emphasis on assessment and self-care skills. Physical and occupational therapists often work together, especially for children with long-term disabilities, as part of a management team.

Self-care skills are taught to increase independence in feeding, dressing, and toileting. Self-care can be achieved by learning special techniques from the therapist, using adaptive equipment, or making the environment more easily livable for the child. Independence learned in childhood enhances the individual’s self-respect and happiness and reduces the burden for the family and the costs for society.



1 Hand splints. These resting splints are invaluable in preventing recurrence of deformity in children with muscle imbalance, burns, or following surgery.

Chapter 4 – Lower Limb

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This chapter covers problems of one or more lower limb segments and includes some of the most common problems in children's orthopedics [1 and 2].

Leg Aches

Leg aches or growing pains, are idiopathic, benign discomfort of extremities, which occur in 15–30% of children. The pains are most common in girls, usually occur at night, and primarily affect the lower limbs. The condition produces no functional disability or objective signs and resolves spontaneously without residual. The cause is unknown. Undocumented speculation on cause includes genetic, functional, or structural (hypermobility) etiology. Leg aches follow headaches and stomach aches as the most common sites of pain during childhood.



1 Rotational deformity. This girl has medial femoral torsion. Note the medial rotation of the patellae.



2 Physiologic bowing. This symmetrical bowing is typical of this benign form of bowing.

Clinical Features

The differential diagnosis of leg aches includes most of the painful conditions involving the musculoskeletal system in children. The diagnosis is made by exclusion [1].

History The pain from leg aches is typically vague, poorly localized, bilateral, nocturnal, and seldom alters activity. This condition does not affect gait or general health. A history of long duration is most consistent with the diagnosis of leg aches. This long duration is helpful in separating out more serious problems, which over a period of time will usually produce objective findings.

Screening examination Does the child appear systemically ill? Is deformity or stiffness present? Does the child limp?

Tenderness Systematically palpate the limbs and trunk for tenderness.

Joint motion Is joint motion guarded or restricted? Check for symmetry of medial rotation of hips.

Differential Diagnosis

Night pain may also be due to a tumor, such as ostoid osteoma, osteogenic, or Ewing sarcomas. Tumor pain is more localized, often associated with a soft tissue mass, progressive, and usually occurs later in childhood than growing pains.

Management

If the history is atypical for leg aches or signs are found on examination, imaging and laboratory studies are required. If the findings are negative, a presumptive diagnosis of growing pains or leg aches is made. Provide symptomatic treatment with heat and an analgesic. Reassure the family about the benign, self-limited course of the condition, but advise them that if the clinical features change, the child should be reevaluated.

Feature	Growing Pain	Serious Problem
History		
Long duration	Often	Usually not
Pain localized	No	Often
Pain bilateral	Often	Unusual
Alters activity	No	Often
Causes limp	No	Sometimes
General health	Good	May be ill
Physical Examination		
Tenderness	No	May show
Guarding	No	May show
Reduced range of motion	No	May show
Laboratory		
CBC	Normal	± Abnormal
ESR	Normal	± Abnormal

1 Differentiating growing pains from more serious problems. The features of growing pains are usually so characteristic that special studies are seldom required.

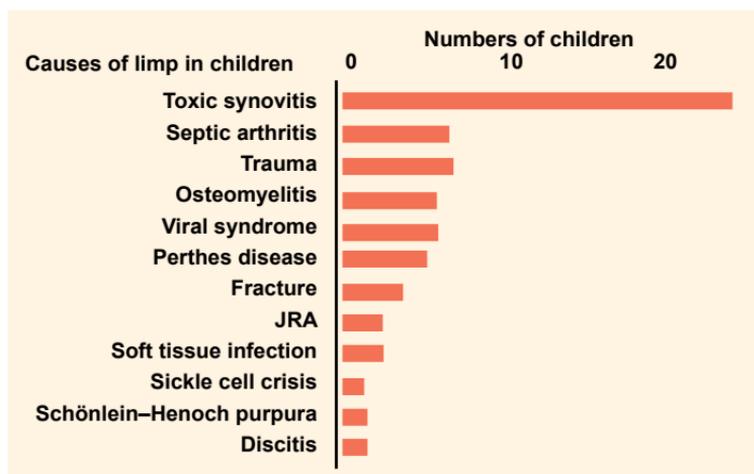
Limp

A limp is an abnormal gait that is commonly due to pain, weakness, or deformity. A limp is a significant finding and the cause should be established [1].

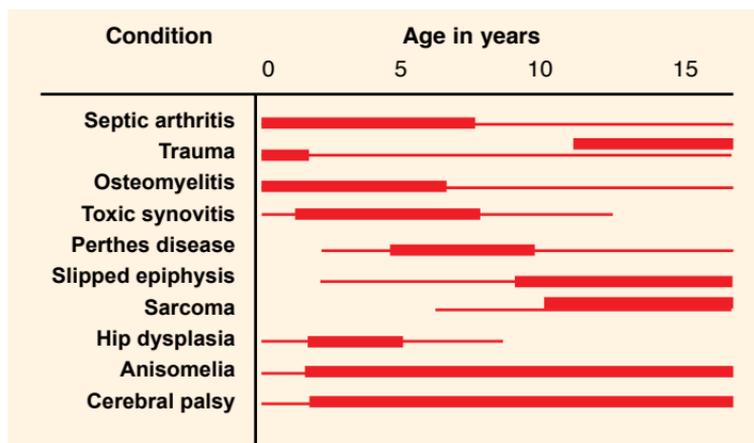
Evaluation

A presumptive diagnosis can usually be made by the history and physical examination. Age is an important factor to consider during evaluation.

History First inquire about the onset [2]. When was the limp first noted? Was the onset associated with an injury or illness? Was it gradual or abrupt? If the limp has been present since infancy, inquire about developmental history, because children with neuromuscular disorders have delayed motor development.



1 Causes of limp in 60 young children. Data from Choban and Killian (1990).



2 Causes of limp by age. The causes of limp are related to the age of the child. The fine red lines show the range and the heavy lines the most common age range of involvement.

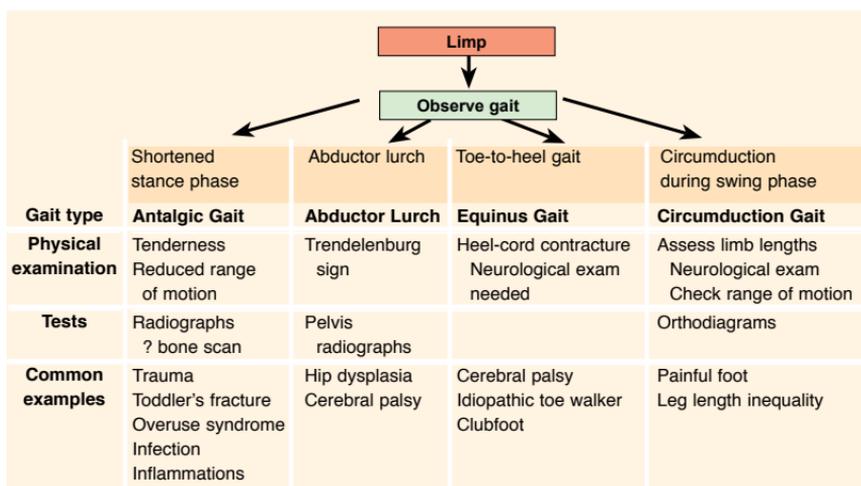
Observation The type of limp can usually be determined by observation. Remove outer clothing to allow the full view of the legs. Watch the child walk in the hallway of the clinic [1]. Observe in three phases: (1) Overview. Look for obvious abnormalities. Which side seems abnormal? Is the stance phase on each side equal in duration? Is lateral shoulder sway present? Is circumduction seen? (2) Study each leg *individually*. Look for more subtle changes. Is the normal heel-to-toe gait pattern present? Does the knee approach full extension during stance phase? How is the hand carried? Elevation of the arm is seen in hemiplegia. (3) Make a presumptive diagnosis, and then make a final observation to be certain that this diagnosis is consistent with the characteristics of the limp.

Types of Limps

The common types of limping may be classified into four groups [2]. The hip is the most common site for the problem [1 opposite page].



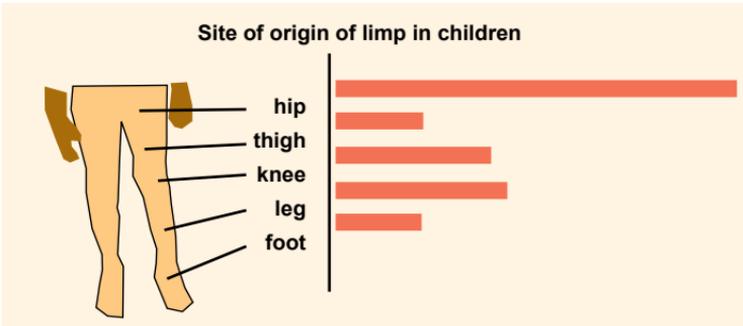
1 Hallway observation. Evaluate the limp by studying the child's gait while the child walks in the clinic hallway.



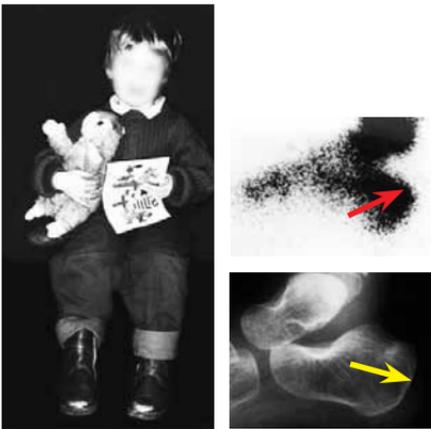
2 Algorithm for evaluation of limping. The major causes of limping are shown. A general categorization is first possible by observation. The exact causes are established by the physical examination and laboratory studies.

Antalgic limp This is a painful limp. The most prominent feature is a shortened stance phase on the affected side. To minimize discomfort, the time of weight bearing on the affected side is shortened. The child is said to "favor" one side or the other. The term "favor" is ambiguous because it may be used to describe either the affected or unaffected side. Find the anatomic location of the problem by determining the site of tenderness, joint guarding, or limitation of motion. Follow-up with radiographs. Often a CBC and ESR or CRP are helpful. If the radiographs are negative, order a bone scan to localize the problem [2 this page and 1 next page].

Equinus gait An equinus gait is due to a heel-cord contracture, which is usually due to cerebral palsy, residual clubfoot deformity, or idiopathic heel-cord tightness. Regardless of the etiology, the contracture causes a "toe-to-heel" sequence during stance phase on the affected side. In the young child, equinus is often associated with a "back knee" or recurvatum deformity of the knee that occurs during stance phase. Document the deformity by evaluating the range of dorsiflexion of the ankle with the knee extended. The ankle should dorsiflex more than 10° . If an equinus deformity is present, a thorough neurological examination is required.



1 Origin of limp in 60 young children. Data from Choban and Killian (1990).



2 Role of bone scan in limp evaluation. This child had an antalgic limp with a negative physical examination, radiographs, and ESR. The bone scan showed increased uptake over the calcaneus (red arrow). This suggested a stress injury to the calcaneus. This was confirmed by a radiograph 2 weeks later showing evidence of a stress fracture (yellow arrow). The child had been stressed by being taken on long walks in a shopping mall.

Abductor lurch results from weakness of the abductor muscles, usually due to hip dysplasia or a neuromuscular disorder. An abductor lurch is characterized by lateral shoulder sway toward the affected side or sides. In normal gait, the abductor muscles contract during stance phase to maintain a level pelvis and a linear progression of the center of gravity of the body. If the abductors are weak, during stance, the pelvis tilts and falls on the unsupported side. To maintain the center of gravity over the foot, the shoulder shifts toward the weak side. This shift is referred to as an *abductor lurch* or a *Trendelenburg gait*. Weakness of the abductors is demonstrated by the Trendelenburg test or sign. The test is positive if the pelvis drops on the unsupported side during single leg standing. The cause of the abductor lurch is usually established by a standing radiograph of the pelvis and a neurological examination.

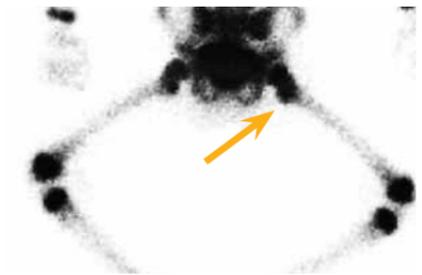
Circumduction allows a functionally longer limb to progress forward during swing phase. Circumduction is often due to a painful condition about the foot or ankle because circumduction requires less ankle movement, making walking more comfortable.

Management

The limp may be caused by something as simple as a stone in the shoe, or by something as serious as leukemia or osteogenic sarcoma. Thus, generalizations regarding management cannot be made. Sometimes the cause of the limp cannot be determined. Should the diagnosis be unclear, reevaluate the child weekly until the problem resolves or a diagnosis is established.



1 Obscure limp from osteomyelitis. This 2-year-old complained of night pain and showed a subtle limp during the day. Radiographs were negative, the bone scan showed slight increased uptake in the upper femur (orange arrow). CT scan demonstrated an intracortical defect (red arrow). The differentiation from ostoid osteomy was made by a resolution with antibiotic treatment.



Torsion

Torsional problems, in-toeing, and out-toeing often concern parents and frequently prompt a variety of treatments for the child. Management of torsional problems is facilitated by clear terminology, an accurate diagnosis, a knowledge of the natural history of torsional deformity, and an understanding of the effectiveness of management options.

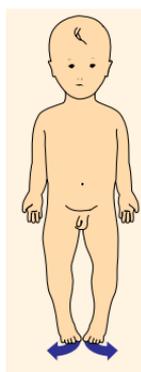
Terminology

Version describes normal variations in limb rotation [1]. Tibial version is the angular difference between the axis of the knee and the transmalleolar axis. The normal tibia is laterally rotated. Femoral version is the angular difference between the transcervical and transcondylar axes. The normal femur is anteverted.

Torsion describes version beyond ± 2 standard deviations (SD) from the mean and is considered abnormal and described as a “deformity.” Internal femoral torsion (IFT) or antetorsion and external femoral torsion, (EFT) or retrotorsion describe abnormal femoral rotation. Internal tibial torsion (ITT) and external tibial torsion (ETT) describe abnormal tibial rotation.

Torsional deformity may be simple, involving one level, or complex, involving multiple segments. Complex deformities may be additive or compensating. Thus, internal tibial torsion and internal femoral torsion are additive. External tibial torsion and internal femoral torsion are compensatory.

Level	Normal	Deformity
Terms	Version within ± 2 SD mean	Torsion >2 SD from mean
Tibia	Tibial version	Tibial torsion Internal (ITT) External (ETT)
Femur	Femoral version Anteversion Retroversion	Femoral torsion Internal (IFT) External (EFT)



2 Lower limb laterally rotates with age (left). Both the femur and tibia laterally rotate with growth. Femoral anteversion declines and tibial version becomes more lateral.

1 Terminology of rotational variations. Normal variations and deformity are described by different terms.



3 Internal femoral torsion affecting mother and child (right). Often evaluation of the parent reveals a rotational pattern similar to that present in the child.

Normal Development

The lower limb rotates medially during the seventh fetal week to bring the great toe to the midline. With growth, femoral anteversion declines from about 30° at birth and to about 10° at maturity [2 previous page]. Values for anteversion are higher in the female and in some families [3 previous page]. With growth, the tibia laterally rotates from about 5° at birth to a mean of 15° at maturity. Because growth is associated with lateral rotation in both the femoral and tibial segments, medial tibial torsion and femoral antetorsion in children improve with time. In contrast, lateral tibial torsion usually worsens with growth.

Evaluation

While the diagnosis of torsional deformities can be made by the physical examination, the history is helpful in excluding other problems and assessing extent of disability.

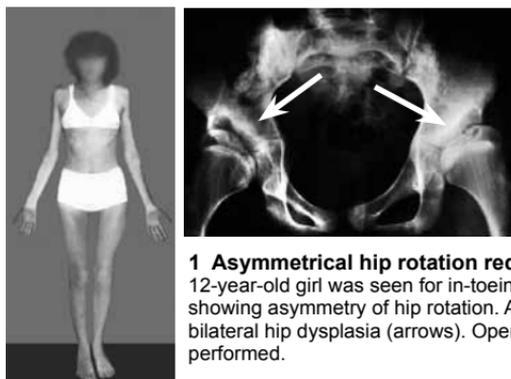
History Inquire about the onset, severity, disability, and previous treatment of the problem. Obtain a developmental history. A delay in walking may suggest a neuromuscular disorder. Is there a family history of a rotational problem? Often rotational problems are inherited and the status of the parent foretells the child's future.

Screening examination Screen to rule out hip dysplasia and neurological problems such as cerebral palsy.

Rotational profile The rotational profile provides the information necessary to establish the level and severity of any torsional problem. See the larger charts on page 419. Record the values in degrees for both right and left sides. Evaluate in four steps:

1. *Observe the child walking and running.* Estimate the *foot progression angle (FPA)* during walking [1 opposite page]. This is the angular difference between the axis of the foot and the line of progression. This value is usually estimated by observing the child walking in the clinic hallway. The average degree of in-toeing or out-toeing is estimated. A minus value is assigned to an in-toeing gait. In-toeing of -5° to -10° is mild, -10° to -15° moderate, and more than -15° severe [3 previous page]. Ask the child to run. The child with femoral antetorsion may show an "eggbeater" running pattern with the legs flipping laterally during swing phase.

2. *Assess femoral version* by measuring hip rotation [1 on page 10]. Measure external (ER) and internal rotation (IR) with the child prone, the knees flexed to a right angle and the pelvis level. Assess both sides at the same time. Internal rotation is normally less than 60° – 70° . If hip rotation is asymmetrical, evaluate with a radiograph [1].



1 Asymmetrical hip rotation requires further evaluation. This 12-year-old girl was seen for in-toeing. The rotational profile was abnormal, showing asymmetry of hip rotation. A radiograph of the pelvis showed severe bilateral hip dysplasia (arrows). Operative correction of the hip dysplasia was performed.

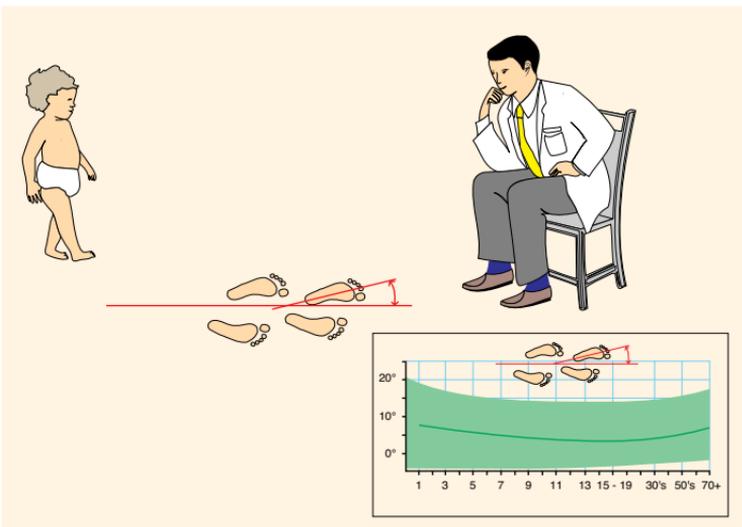
3. *Quantitate tibial version* by assessing the *thigh-foot angle* [1 on page 11]. With the child prone and the knee flexed to a right angle, the TFA is the angular difference between the axis of the foot and the axis of the thigh. The TFA measures the tibial and hindfoot rotational status. The TMA is the angular difference between the transmalleolar axis and the axis of the thigh. This is a measure of tibial rotation. The difference between the TMA and the TFA is a measure of hindfoot rotation. The normal range of both the TFA and TMA is broad, and the mean values increase with advancing age. For these measurements, positioning of the foot is critical. Allow the foot to fall into a natural position. Avoid manual positioning of the foot as this is likely to cause errors in assessment.

4. *Assess the foot* for forefoot adductus. The lateral border of the foot is normally straight. Convexity of the lateral border and forefoot adduction are features of metatarsus adductus. An everted foot or flatfoot may contribute to out-toeing. Include both in the rotational profile.

From the screening examination and rotational profile establish the cause of the torsional deformity [2 next page].

Special Studies

Order special imaging studies if hip rotation is asymmetrical or if the rotational problem is so severe that operative correction is being considered. In general, special imaging to document rotational problems is not very useful. Before operative correction, image severe antetorsion to rule out hip dysplasia and to measure the degree of femoral antetorsion. Measurements can be made by CT scans or biplane radiographs. Usually antetorsion exceeds 50° in children whose condition is severe enough to require operative correction.



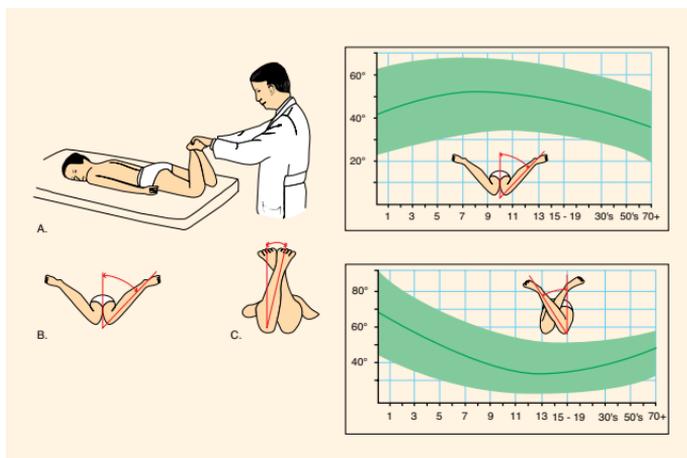
1 Foot progression angle. Foot progression angle is estimated by observing the child walking. The normal range is shown in green.

Management Principles

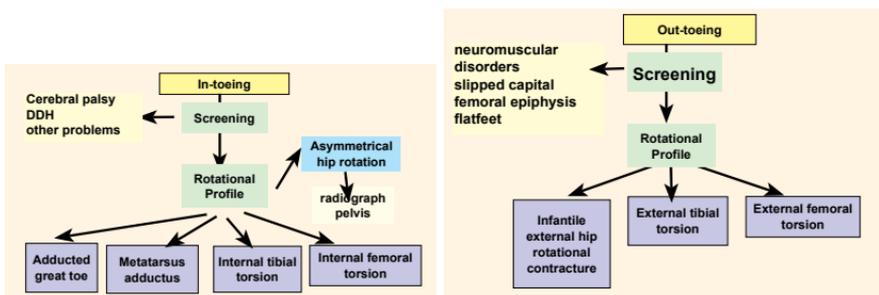
The first step is establishing a correct diagnosis. In managing rotational problems, the most common management challenge is dealing effectively with the family. Because the lower limbs laterally rotate with time, in-toeing spontaneously corrects in the vast majority of children. Thus, simply waiting to allow this spontaneous resolution is best for the child. Attempting to control the child’s walking, sitting, or sleeping positions is impossible. Such attempts only create frustration and conflict between the child and parent.

Shoe wedges or inserts are ineffective [2 opposite page]. Likewise, day-time bracing with twister cables only limits the child’s walking and running activities [3 opposite page]. Night splints that laterally rotate the feet are better tolerated and do not interfere with the child’s play, but probably have no long-term benefit.

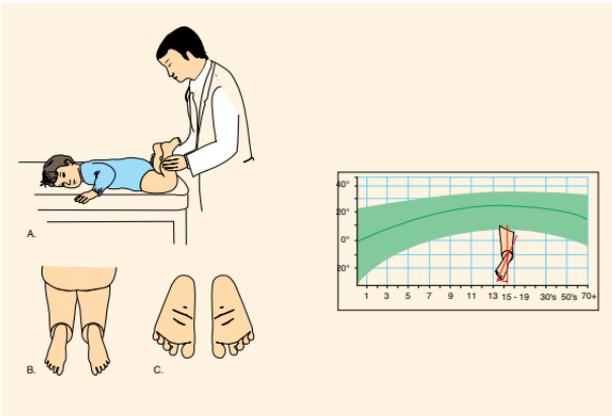
Thus, observational management is best. The family needs to be convinced that only observation is appropriate. This requires careful evaluation, education, reassurance, and follow-up. The family should be informed that only rarely does a torsional problem persist. Less than 1% of femoral and tibial torsional deformities fail to resolve and may require operative correction in late childhood. The need for rotational osteotomy is rare and the procedure is effective.



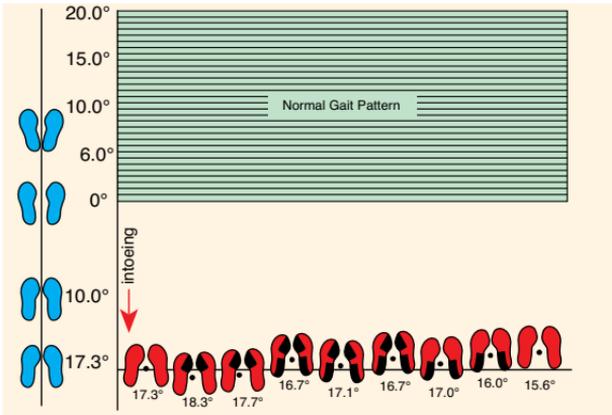
1 Hip rotation. (A) Hip rotation is assessed with the child prone. (B) Internal rotation and (C) external rotation are measured. Normal ranges are shown in green.



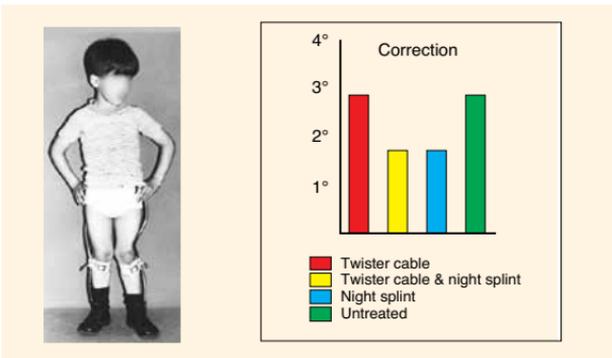
2 Flow-charts for assessment of in-toeing and out-toeing. By the screening examination and the rotational profile, the diagnosis can usually be readily established.



1 Assessing rotational status of tibia and foot. The rotational status of the tibia and foot are best assessed by evaluating the child in the prone position (A) allowing the foot to fall into a natural resting position. (B) The thigh-foot axis and (C) shape of the foot are readily determined. The range of normal is shown in green.



2 Ineffectiveness of shoe wedges. Various wedges were placed (shown in black). Mean values for intoeing for each wedge are shown compared with the unwedged controls. Redrawn from Knittle and Staheli (1976).



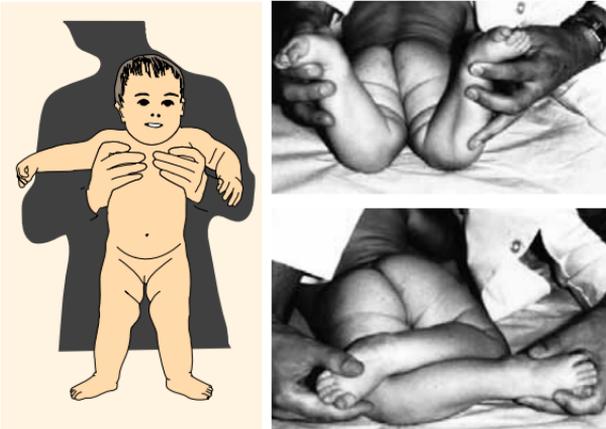
3 Lack of effectiveness of twister cables. The chart compares the effectiveness of various "treatments" and the untreated child with antetorsion. These interventions made no difference in the measured femoral anteversion before and after treatment. From Fabry et al. (1973).

Infant

Out-toeing may be due to flatfeet with heel valgus or more commonly due to a lateral rotation contracture of the hips. In-toeing may be due to an adducted great toe, forefoot adductus, or internal tibial torsion.

Lateral hip rotational contracture Because the hips are laterally rotated *in utero*, lateral hip rotation is normal. When the infant is positioned upright, the feet may turn out [1]. This may worry the parents. Often only one foot turns out, usually the right. The turned out foot is the more normal one. The opposite limb, the one that is considered normal by the parents, often shows metatarsus adductus or medial tibial torsion.

Adducted great toe The adducted great toe has been described both as a spastic abductor hallucis and as a “searching toe.” This is a dynamic deformity due to a relative overpull of the abductor hallucis muscle that occurs during stance phase [2]. This may be associated with adduction of the metatarsals. The condition resolves spontaneously when maturation of the nervous system allows more precision in muscle balance around the foot. No treatment is required.



1 Physiologic infantile out-toeing. Out-toeing in early infancy is usually due to a lateral rotation contracture of the hips. In this infant, medial rotation is limited to about 30° (upper photograph), whereas lateral rotation is about 80° (lower photograph). This results in a lateral rotation of the limb (drawing), which resolves spontaneously.

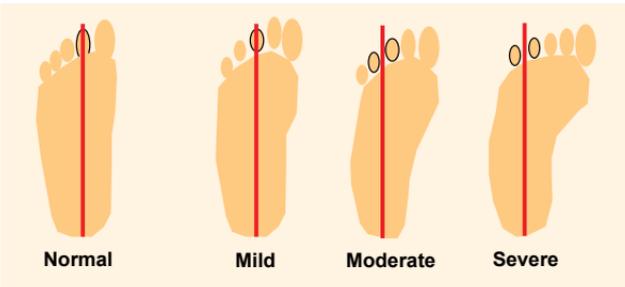


2 Searching toe. This is a dynamic deformity due to overactivity of the abductor hallucis muscle.

Forefoot adductus describes a spectrum of foot deformities characterized by a medial deviation of the forefoot of different degrees [1]. The prognosis is clearly related to stiffness. The condition is detailed in the next chapter.

Metatarsus adductus Flexible deformities are deformations that occur from intrauterine crowding. Like other deformations, they resolve spontaneously with time. Most resolve within the first year and the rest over childhood. Manage with observation and reassurance. If the deformity persists after the second year, resolution may be hastened with bracing that holds the foot abducted and the leg laterally rotated.

Metatarsus varus Rigid forefoot adductus tends to persist. The deformity is characterized by stiffness and a crease on the sole of the foot. The natural history is for incomplete spontaneous resolution. The deformity produces no functional disability and is not the cause of bunions. It produces a cosmetic problem, and when severe, a problem with shoe fitting. Be sure to distinguish the rare skewfoot. Recall that the skewfoot occurs in loose-jointed children and is characterized by marked forefoot adductus and hindfoot valgus. Most parents want the deformity corrected. As described on page 98, correct by long-leg braces or casts [2].



1 Grading severity of forefoot adductus. Project a line that bisects the heel. Normally it falls on the 2nd toe. The projected line falls through the toe 3 in mild, between toes 3–4 in moderate, and between toes 4–5 in severe deformity. From Bleck (1983).



2 Metatarsus varus. Stiff or persisting deformity can be corrected with long-leg splints (yellow arrows) that abduct the forefoot (red arrows).

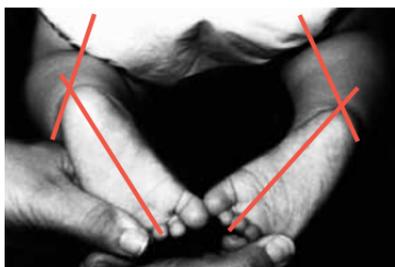
Toddler

In-toeing is most common during the second year, usually noticed when the infant begins to walk. This in-toeing is due to internal tibial torsion, metatarsus adductus, or an adducted great toe.

Internal tibial torsion Internal tibial torsion (ITT) is the most common cause of in-toeing. ITT is often bilateral [1]. Unilateral ITT is most common on the left side [2]. Observational management is best. Fillauer or Denis Browne night splints are commonly prescribed, but probably have no long-term value with resolution occurring with or without treatment [3]. Avoid daytime bracing and shoe modifications because they can slow the child’s running and may harm the child’s self-image.

Child

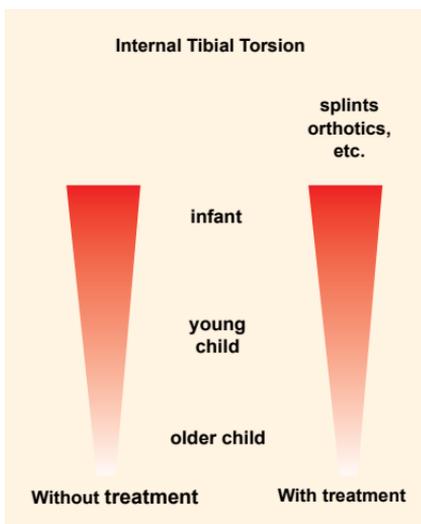
In-toeing in childhood is commonly due to femoral antetorsion and rarely to persisting internal tibial torsion. In late childhood, out-toeing may be due to external femoral torsion or external tibial torsion. The natural history is to externally rotate with growth, often correcting internal tibial torsion and making external tibial torsion worse [1 opposite page].



1 Bilateral internal tibial torsion. The thigh-foot angled are negative (red lines) for both legs.



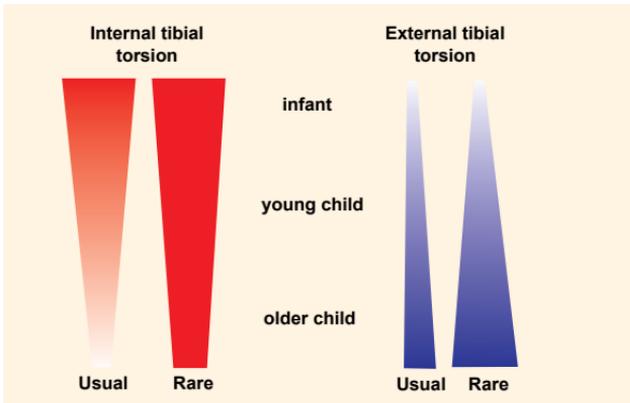
2 Unilateral internal tibial torsion. Medial tibial torsion is often asymmetrical, usually worse on the left side (arrow).



3 Management of internal tibial torsion. Management with or without intervention gives the same excellent results.

Internal tibial torsion is less common than external tibial torsion in the older child. ITT may also require operative correction if the deformity persists and produces a significant functional disability and cosmetic deformity in the child over 8 years of age [2]. Operative correction may be indicated if the thigh-foot angle is internally rotated more than 10° .

External tibial torsion Because the tibia normally rotates laterally with growth, ITT usually improves but ETT becomes worse with time [2 this page and 1 on page 17]. ETT may be associated with knee pain. This pain arises in the patellofemoral joint and is presumably due to malalignment of the knee and the line of progression. This malalignment is most pronounced when ETT is combined with IFT. The knee is internally rotated and the ankle externally rotated, both out of alignment with the line of progression, producing a "malalignment syndrome." This condition produces an inefficient gait and patellofemoral joint pain.



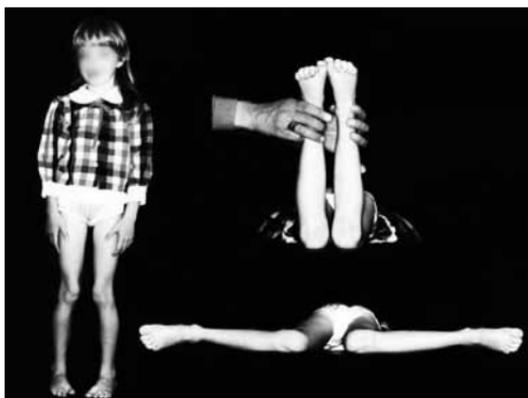
1 Comparison of natural history of internal and external tibial torsion. As the tibia laterally rotates with growth, internal torsion improves and external torsion may worsen. Torsion severe enough to require tibial rotational osteotomy is more common with lateral torsional deformities.



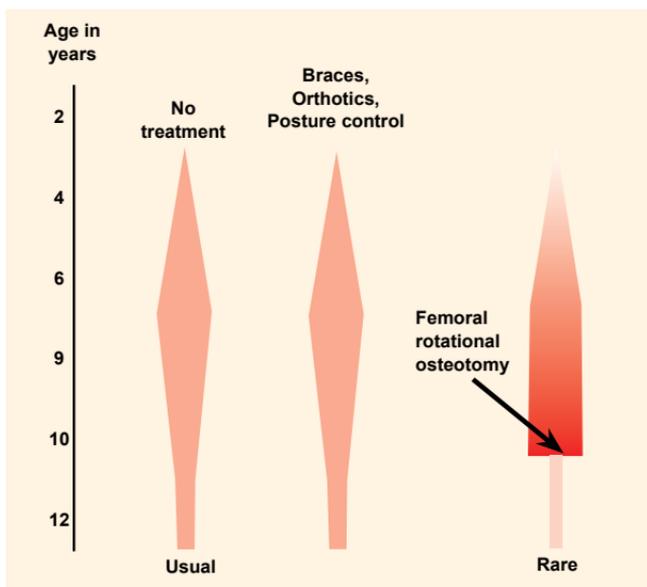
2 Persisting tibial torsion. Rotational deformities do not always resolve with time. These girls show persisting tibial torsion (arrows), which caused enough disability to require tibial rotational osteotomy for correction.

Femoral antetorsion or internal femoral torsion is usually first seen in the 3–5 year age groups and is more common in girls [1]. Mild residual eformity is often seen in the parents of affected children. The child with MFT sits in the “W” position, stands with the knees medially rotated (“kissing patella”), and runs awkwardly (“egg-beater”). Internal hip rotation is increased beyond 70°. IFT is mild if the internal hip rotation is 70°–80°, moderate if 80°–90°, and severe if 90+°. External hip rotation is reduced correspondingly, as the total arch of rotation is usually about 90°–100°.

Femoral antetorsion usually is most severe between 4 and 6 years of age and then resolves [2]. This resolution results from a decrease in femoral anteversion and from a lateral rotation of the tibia. In the adult, Femoral antetorsion does not cause degenerative arthritis and rarely causes any disability.



1 Medial femoral torsion. This girl has medial femoral torsion. Her patella face inward on standing. Her lateral rotation is 0° (upper) and her internal rotation is 90° (lower).



2 Clinical course of femoral antetorsion. Femoral antetorsion becomes more clinically apparent during infancy and early childhood. The deformity is usually most severe between about 4 and 6 years of age. Resolution occurs regardless of common treatments. Rarely, the deformity is severe and fails to improve and requires rotational osteotomy for correction.

Femoral antetorsion is unaffected by nonoperative treatment. Persistence of severe deformity after the age of 8 years may necessitate correction by a femoral rotational osteotomy.

Femoral retrotorsion may be of greater significance than commonly appreciated. Retrotorsion is more common in patients with slipped capital femoral epiphysis. Presumably the shear force on the physis is increased. Retrotorsion is associated with increased degenerative arthritis and an out-toeing gait. The gait problem is not sufficiently severe to warrant operative correction.

Operative Correction

Rotational osteotomy is effective in correcting torsional deformities of the tibia or femur [1 on page 20]. Osteotomy is indicated only in the older child, over age 8–10 years, who has a significant cosmetic and functional deformity, and with a single deformity 3 SD above the mean or combined deformity 2 SD above the mean. The child’s problem should be sufficiently severe to justify the risks of the procedure. These procedures should not be considered “prophylactic.”

Femoral correction Femoral rotational osteotomy is best performed at the intertrochanteric level. At this level healing is rapid, fixation most severe, scarring least obvious and should malunion occur, least noticeable. Usually rotational correction of about 50° is required. See page 401.

Tibial correction Tibial rotational osteotomy is best performed at the supramalleolar level [2]. Correct rotation to bring the thigh-foot angle to about 15°. See page xxx.



1 External tibial torsion. External tibial torsion is often unilateral. When asymmetrical it is usually worse on the right side (arrow). Note that the thigh-foot angle is more external on the right (red lines).



2 Rotational osteotomy. Rotational osteotomies are usually performed at the supramalleolar tibial or intertrochanteric femoral levels (arrows). Fix tibial osteotomies with crossed transcutaneous pins and a long leg cast. Fix femoral osteotomy with pins and a cast or a nail-plate.

Rotational Malalignment Syndrome

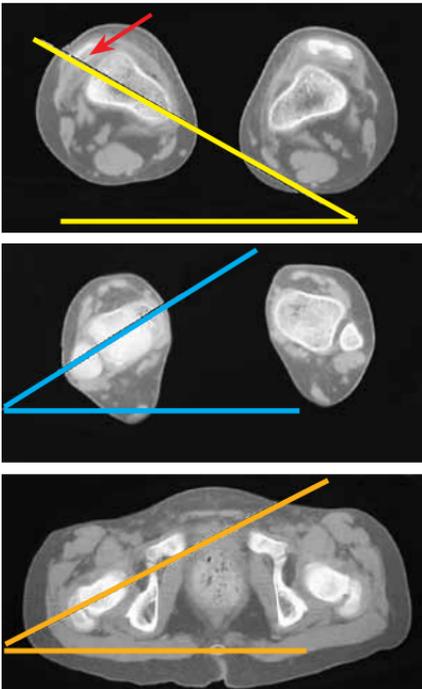
This syndrome usually includes external tibial and internal femoral torsion. The axis of flexion of the knee is not in the line of progression. Patellofemoral problems of pain and, rarely, dislocation follow.

Manage most conservatively. Very rarely operative correction is necessary. Correction is a major undertaking as it usually requires a 4 level procedure (both femoral and tibia). The site of the tibial osteotomy may be distal (most safe) or proximal. Proximal osteotomy just above the tibial tubercle has been reported.

Rarely, rotational malalignment is associated with severe patellofemoral disorders such as congenital dislocations [1]. Correction is complex and may require both osteotomy and soft tissue reconstructions.

Prognosis

The effects of adults with internal femoral torsion show little or no functional disability. Mild internal tibial torsion may facilitate sprinting by improving push-off. Degenerative arthritis of the knee has been associated with femoral antetorsion and of the hip with femoral retrotorsion.



1 Rotational malalignment with patellar dislocations. This CT study of a child with a dislocated patella (red arrow) shows torsional malalignment. Note the 40° lateral tibial torsion (blue lines), the 40° medial rotation of the axis of the knee (yellow lines), and 30° femoral anteversion (orange lines).

Leg Length Inequality

Leg length inequality or anisomelia may be structural [1] or functional. Functional anisomelia is secondary to joint contractures producing an apparent discrepancy in length. Structural discrepancies may occur at any site in the limb or pelvis. Often only discrepancies of the tibia or femur are measured. The height of the foot and pelvis should be included in calculating the total disparity. Discrepancies of 1 cm or more are considered significant.

Etiology

The causes of anisomelia are numerous [1 next page]. Minor discrepancies are seen in clubfeet, hip dysplasia, and Perthes disease. Major differences are seen in tibial or femoral agenesis.

Natural History

The course of anisomelia is determined by the cause. The inhibition or acceleration that causes progressive forms of anisomelia varies according to the etiology. Growth inhibition from congenital defects is usually constant and makes predicting the final disparity feasible. Inhibition or acceleration from vascular, infectious, or neoplastic disorders are variable. For example, growth acceleration may be associated with chronic diaphyseal osteomyelitis. The acceleration occurs only when the infection is active.

Gait

The effect on gait depends on the magnitude of the discrepancy and the age of the patient. Children compensate for discrepancies by flexing the knee on the long side or by standing in equinus on the shortened limb. These compensations level the pelvis. Discrepancies are compensated by altered function. The long limb may be circumducted during swing phase or by "vaulting" over the long limb during stance phase. This vaulting results in a rise and fall of the body and consumes more energy than normal gait.

Adverse Effects

The adverse effects of anisomelia have been overstated. Limb length difference in childhood does not lead to an increased risk of structural scoliosis or back pain in adults.



1 Leg length differences. The boy has overgrowth of the right leg (red arrow) due to Klippel-Trenaunay syndrome. This girl has a short right leg due to weakness secondary to poliomyelitis (yellow arrow).

Evaluation

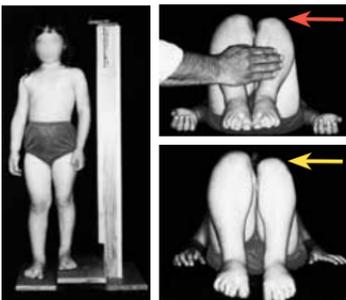
During the evaluation, calculate the projected height of the patient and the degree of shortening at skeletal maturity if untreated. This evaluation requires a screening examination, a search for the cause, clinical and radiographic assessment of severity, and a determination of bone age. Serial evaluations are necessary during growth to improve the accuracy of the evaluation. From the history, determine if the child has been injured or experienced any musculoskeletal diseases.

Screening examination Note any asymmetry and alterations in body proportions. Does the asymmetry involve only the lower limbs? Is the long side the normal or abnormal side? Sometimes overgrowth makes the long side the abnormal one. Is it a hemihypertrophy or hemihypoplasia? Hemihypertrophy [1 previous page] is important to recognize because it is sometimes associated with Wilm’s tumor. The finding of hemihypertrophy should prompt an abdominal ultrasound evaluation. Hemihypoplasia is usually due to hemiparesis from cerebral palsy. Often these underlying problems are more significant than the length discrepancy itself. Observe the child walking. Is equinus, vaulting, circumduction, or abductor lurch present? Assess the abnormal limb to determine the site or sites of the discrepancy. Are the feet of equal length? Are the tibial and femoral segments equal? Are the forearms of equal length? Are any associated abnormalities present? Is joint motion symmetrical? Assess to determine whether the difference is in the femur, tibia, or combined [2].

Clinical measures of discrepancy The limbs can be measured from the medial malleolus to the anterior iliac spine with a tape measure. This is usually accurate within about 1 cm. A more practical method uses blocks. Blocks of known thickness are placed under the short side until the pelvis is level. The patient will often sense when symmetry is established. By this method, all segments including foot and pelvis are assessed.

Category	Shortening	Lengthening
Congenital	Aplasia Hypoplasia Hip dysplasia Clubfoot	Hyperplasia
Neurogenic	Paralysis Disuse	Sympathectomy
Vascular	Ischemia Perthes disease	AV fistular
Infection	Physeal injury	Stimulation
Tumors	Physeal involvement	Vascular lesions
Trauma	Physeal injury Malunion	Fracture stimulation Distraction

1 Causes of limb length discrepancy. The common causes of limb shortening and lengthening are shown.



2 Leg length inequality. In this child the right lower leg is hypertrophied. Note that the tibia is longer and the diameter greater. The left leg is more proportional in size to the rest of the body. The left limb is shorter both in the femur (red arrow) and tibia (yellow arrow).

Imaging methods Image to measure discrepancies and determine any associated bone or joint deformities. Radiographic measures include the teleroentgenogram with a single exposure or orthodiagrams requiring multiple exposures on the same film. The orthodiagrams may be full length on a 36-inch film or telescoped on a 17-inch film [1]. For the infant and young child, order a teleroentgenogram because it provides an excellent screen for other problems such as hip dysplasia, it requires only one exposure, and it does not require patient cooperation. Enough serial studies should be made to provide adequate documentation to time the epiphysiodesis. These need not be done yearly. If the discrepancy is detected in the infant, obtain the baseline study early and repeat at about 3, 6, and 9 years of age.

Bone age Bone age is the most inaccurate of the measurements. Often measurements are given with a ± 2 year qualification. The standard for assessment is the Gruelich and Pyle atlas. It is wise to sample bone ages over a period of several years and average any differences from the chronological age to improve reliability.

Body height at skeletal maturation The projected height at skeletal maturation is sometimes useful in planning correction of anisomelia. Shortening is more feasible for the tall individual, whereas lengthening may be more acceptable for those of short stature. The estimation can be made by comparing the child's height with the bone age to determine a percentile. This percentile is projected to maturity as an estimate of adult height.

Calculating discrepancy at maturation The discrepancy at maturation is the sum of the current discrepancy and the discrepancy accumulated during the period of remaining growth. The current discrepancy is assessed by clinical and radiographic measures. The discrepancy created by remaining growth must be calculated based on the percentage of growth retardation (or acceleration).

Minimal acceptable height (MAH) The MAH is the shortest stature that would be acceptable to the family. This will be based on racial, social, cultural, individual, and family differences. As a starting point for discussion, set the MAH at 2 SD below the mean value or about 65 inches for men and 59 inches for women [2 next page]. Establishing the MAH involves an integration of complex issues such as the value the family gives to preservation of height and balancing this with the increased risks of lengthening over shortening.



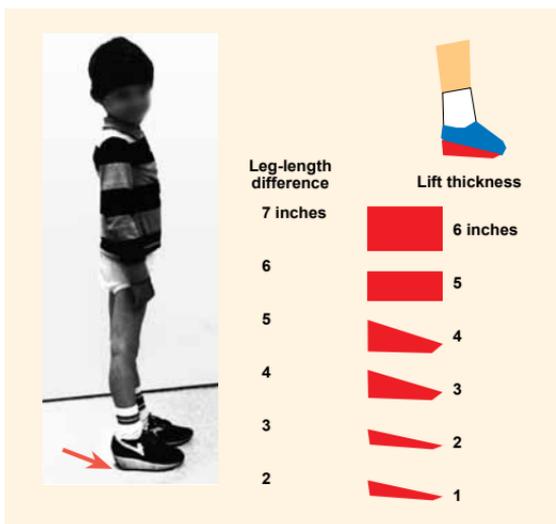
1 Radiographic measures. The long radiograph (red arrow) is best for young children. The orthodiagram is more accurate for older children (yellow arrow).

Management Principles

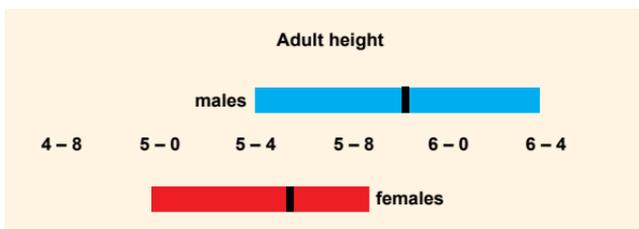
The objective of management is to level the pelvis by equalizing extremity length without imposing excessive risk, morbidity, or height reduction. The severity of the discrepancy determines the general approach to management.

Severity Degrees of shortening can be categorized to aid in planning management. These values are influenced by the minimal acceptable height as determined during evaluation. In general, correct the discrepancy by shortening down to the MAH, then lengthen the limb as required to achieve the MAH.

Lifts Lifts may be useful in discrepancies greater than 2–3 cm [1]. Lifts cause problems for the child. They make the shoe heavier and less stable and are usually a source of embarrassment. Lifts make a clear statement, “I have a disability,” which may be harmful to the child’s self-image and status among peers. Because no immediate or late harmful effect of uncompensated anisomelia has been shown, the lift should improve function enough to compensate for inherent problems of wearing a lift. Walking without the lift will not damage the child. Lifts may be applied inside the shoe or on the heel. Make the lift as inconspicuous and lightweight as possible. More in-shoe correction can be placed in a high-top shoe. Consider placing 1 cm inside and another cm on the heel. Order tapered lifts when possible as the less bulk means a lighter, more stable and less conspicuous lift. To further reduce the lift size, order a lift that will leave the correction about 2 cm less than the disparity.



1 Shoe lifts. Attempt to use wedges to minimize size and weight. For large discrepancies block lifts are necessary. Maintain correction below the actual discrepancy to minimize disability.



2 Distribution of normal adult height. The mean value is shown in black with the range of ± 2 SD shown in blue for men and red for women.

Timing of Correction

The usual objective of management is to correct the leg length discrepancy to within 1 cm of the opposite side. Because of its simplicity, effectiveness, and safety, epiphysiodesis remains the most effective means of correcting discrepancies between 2 and 5 cm.

The timing of epiphysiodesis determines the degree of correction, and 5 methods of timing are commonly used.

The simplistic method is useful for giving a rough estimate of the discrepancy at maturation from discrepancies of congenital origin. This is based on the assumption that the growth retardation is consistent. For example, a child with a congenital discrepancy of 3 cm at age 2 years has reached roughly half of his adult height. Thus, at skeletal maturation, the discrepancy is likely to be about 6 cm.

The arithmetic method is based on average growth rates and chronological age. On average, the distal femur contributes 3/8 inch of growth per year and the proximal tibia contributes 1/4 inch per year [1]. Girls complete growth at 14 and boys at 16 years of age. Use this method for long-term planning.

The growth-remaining method for timing of epiphysiodesis was the standard for many decades [1 next page].

The Paley multiplier method allows prediction of eventual discrepancy by simply multiplying the disparity with an age-adjusted factor [2 next page] to establish the disparity at maturation.

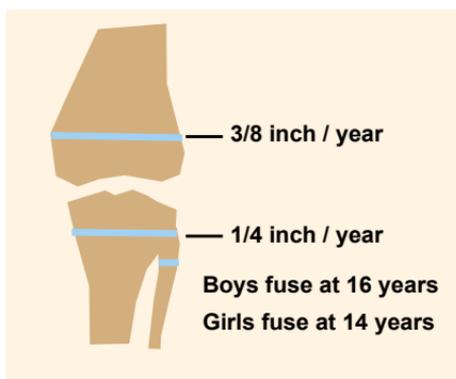
Straight line graph method requires a special graph for each patient [3 next page]. The method is graphic and has the advantage of averaging the bone ages. This method has become the standard method of timing. The timing and procedure are detailed on page 398.

Correction

Plan management based on age of diagnosis, severity, projected height at maturity and any other special factors [2 on page 149].

Techniques of Correction

Bone shortening is a relatively safe and effective method of correcting discrepancies in the patient beyond the age when correction by epiphysiodesis is possible. Closed shortening procedures are now the standard [1 on page 149].

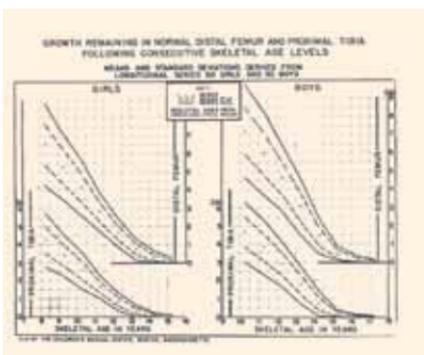


1 Arithmetic method of predicting effect of epiphysiodesis. The growth rate per year for the lower femur and upper tibia are shown.

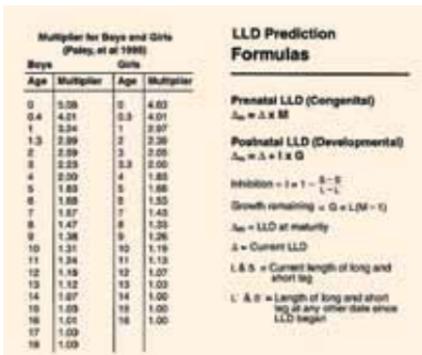
Stapling as a means of achieving an epiphysiodesis is appropriate only when calculating the appropriate timing for an epiphysiodesis is not possible due to difficulties in reading bone age and plotting growth.

Epiphysiodesis is the best method to correct most discrepancies between 2 and 5 cm. The traditional method leaves a long scar. Newer percutaneous methods use either a curette [3 opposite page] or a drill to remove the growth plate.

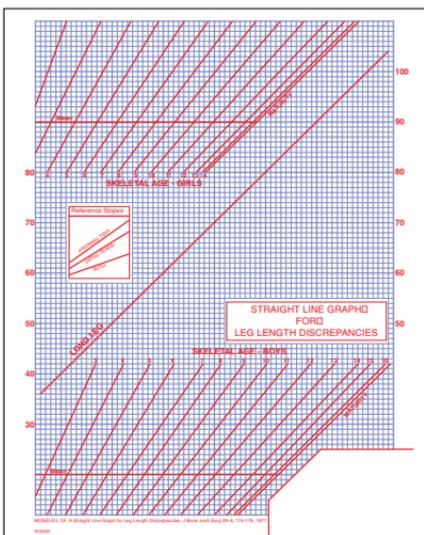
Lengthening as a means of correcting anisomelia has been practiced for 70 years. During the past two decades, new techniques have reduced the risks and made the procedure more effective [4 opposite page]. This increased effectiveness is primarily due to the improved osteogenesis achieved by applying biological principles established through research.



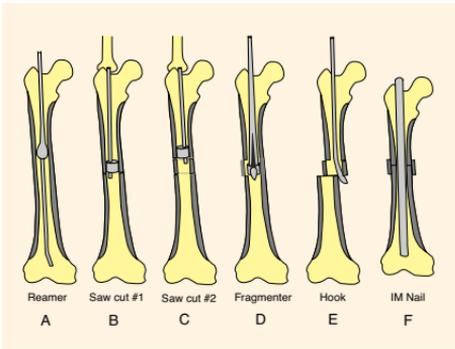
1 Growth-remaining charts for girls and boys. The growth-remaining charts for girls and boys are different. Actual correction is based on growth of the short limb. To use it correctly, the discrepancy at maturity and the percentage of growth retardation of the short limb should be calculated. Redrawn from Anderson and Green (1963).



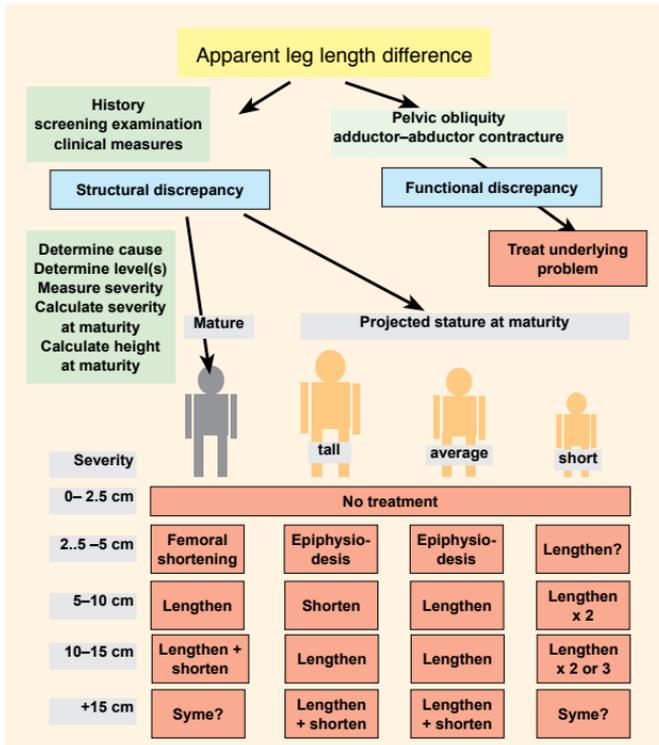
2 Paley multiplier. From the Maryland Center for Limb Lengthening and Reconstruction. This is a simple method of determining the leg length difference at maturation. This is applicable for shortening conditions in which growth retardation is consistent. From Paley et al (2000).



3 Moseley straight line graph. This method utilizes graphic presentation of data to calculate the age for epiphysiodesis. See page 412 for full-size graph.



1 Closed femoral shortening. A segment of femur is removed by a saw placed down the medullary canal from above. A segment is divided, split, displaced, then fixed with an intramedullary nail. From Winquist (1986).



4 Ilizarov lengthening.

3 Epiphysiodesis. The growth plate is being removed by curettage. This causes bony fusion across the growth plate and arrests growth.

Genu Varum and Genu Valgum

Genu varum and genu valgum are frontal plane deformities of the knee angle that fall outside the normal range, ± 2 SD of the mean. Knee angle variations that fall within the normal range are referred to as *bowlegs* or *knock-knees* or physiologic variations [1]. The range of normal for knee angle changes with age [2]. Lateral bowing of the tibia is common during the first year, bowlegs are common during the second year [3], and knock-knees are most prominent between 3 and 4 years [1 opposite page]. Varus or valgus deformities are classified as either "focal" as seen in tibia vara or "generalized" as occurs in rickets.

Evaluation

History Inquire about the onset. Was there an injury or illness? Is the deformity progressing? Are old photographs or radiographs available for review? Is the child's general health good? Does the family provide a normal diet? Are other family members affected?

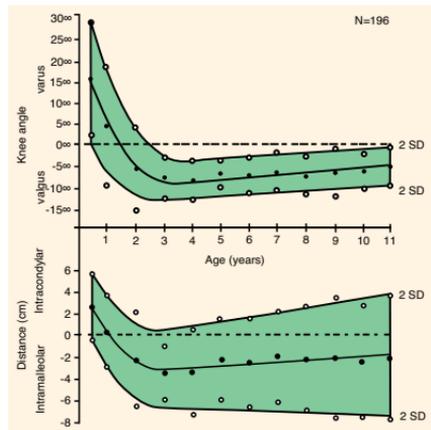
Physical examination Start with a screening evaluation. Does the child have normal height and body proportions? Short stature is common in rickets and various syndromes. Are other deformities present? Is the deformity symmetrical? Is the deformity localized or generalized? Are the limb lengths equal? Shortening and knee angle deformity may be due to epiphyseal injuries or some developmental problems such as fibular hemimelia. Measure the rotational profile. Often frontal and transverse plane deformities coexist; make a clear separation. Measure the deformity. With the patella directly forward, measure the knee angle with a goniometer. Measure the intramalleolar or intracondylar distance. Does the deformity increase when the child stands? If the collateral ligaments are lax, such as in achondroplasia, the varus deformity is worse in the upright position.



1 Physiological bowlegs and knock-knees. These siblings show the sequence with the toddler with bowlegs and the older sister with mild knock-knees.



3 Physiologic bowlegs. This 18-month-old infant has moderate bowing.



2 Normal values for knee angle. The normal values for the knee angle are shown both in degrees and intracondylar or intramalleolar distances. From Heath and Staheli (1993).

Laboratory If the child has a generalized deformity, order a metabolic screen including calcium, phosphorus, alkaline phosphatase, and creatinine, plus a hematocrit.

Imaging If findings suggest the possibility of a pathological basis for the deformity, order a single AP radiograph of the lower limbs [2]. If knee ligaments are loose, make the radiograph with the infant or child standing. Position the child with the patella directly forward [3]. Use a film large enough to include the full length of femora and tibiae. A 36-inch film is often required. Study the radiograph for evidence of rickets, tibia vara, or other problems. Measure the metaphyseal-diaphyseal angle of the upper tibia (page 84). Values above 11° are consistent with tibia vara. Measure the hip-knee-ankle angle. Complete the evaluation with other imaging studies if necessary. For knee deformities, a lateral radiograph is useful. Early tibia vara can be assessed with a bone scan. Uptake is increased in the medial portion of the proximal tibial epiphysis. CT or MRI studies may be useful in identifying and measuring a physeal bridge. Document the deformity by photography. A sequence of photographs provides a graphic record of the effect of time.

Diagnosis

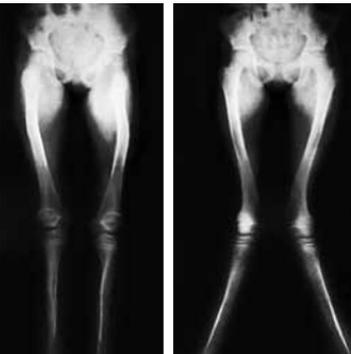
Follow a plan [1 and 2 on page 153]. First make the differentiation between physiologic and pathologic forms [1 and 2 next page]. If a pathologic form is present, consider the various categories of causes [3 next page]. Causes are varied and usually the diagnosis is not difficult.



1 Physiologic knock-knees. This 3-year-old girl has mild physiologic knock-knees.



2 Positioning for radiographs. This girl is being carefully positioned to assure an accurate study.



3 Poor and well positioned radiographs. Patient positioned to get legs on radiographs (left). Properly positioned study shows deformity (right).



1 Pathological genu valgum. Deformity due to physeal arrest from trauma (red arrow), and to osteochondromatosis (yellow arrows) are shown.

Feature	Physiologic	Pathologic
Frequency	Common	Rare
Family hx	Usually negative	May occur in family
Diet	Normal	May be abnormal
Health	Good	Other MS abnormalities
Onset	Second year for bowing Third year knock-knees	Out of normal sequence Often progressive
Sequence	Normal sequence	Variable
Height	Normal	Less than 5th percentile
Symmetry	Symmetrical	Symmetrical or asym
Severity	Mild to moderate	Often beyond ± 2 SD

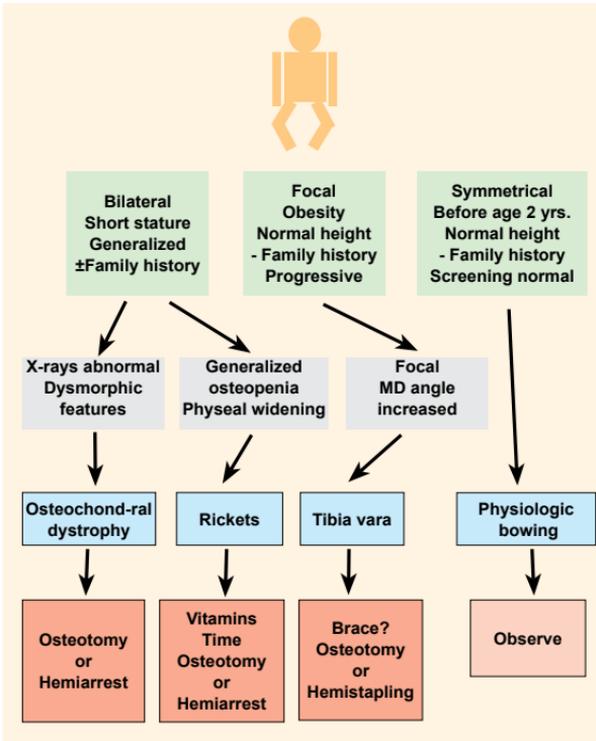
2 Differentiating physiologic and pathologic genu varum.

Cause	Genu Valgum	Genu Varum
Congenital	Fibular hemimelia	
Dysplasia	Osteochondrodysplasias	Osteochondrodysplasias
Developmental	Knock-knee > 2 SD	Bowing > 2 SD Tibia vara
Trauma	Overtgrowth Partial physeal arrest	Partial physeal arrest
Metabolic	Rickets	Rickets
Osteopenic	Osteogenesis imperfecta	
Infection	Growth plate injury	Growth plate injury
Arthritis	Rheumatoid arthritis knee	

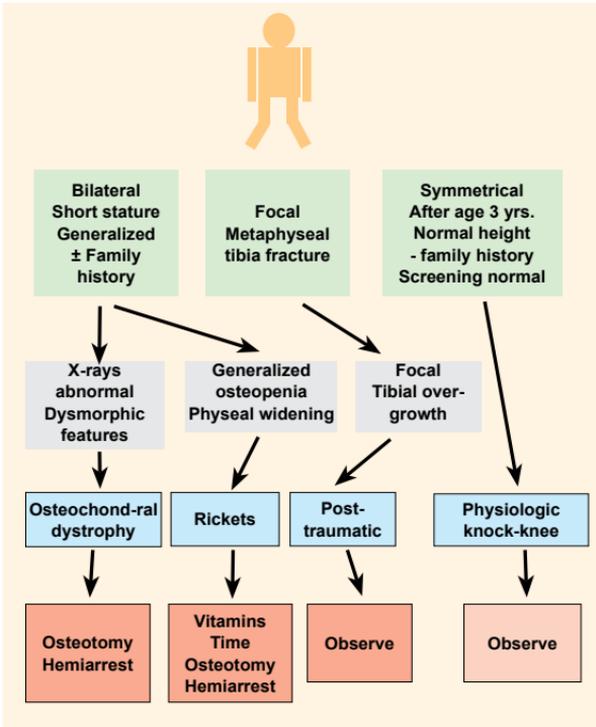
3 Classification of pathologic knee angle. Causes of genu varum and genu valgum are listed.



1 Evaluation of genu varum or bowlegs. This flowchart shows the differentiation of the common causes of change in knee angle.



2 Evaluation of genu valgum or knock-knees. This flowchart shows the differentiation of the common causes of change in knee angle.



Management

The vast majority of children have bowlegs or knock-knees that will resolve spontaneously. Document these physiological variations with a photograph and see the child again in 3–6 months for follow-up. No radiographs are necessary. If the problem is pathological, establish the cause. Treatment options are then considered.

Nonoperative treatment with shoe wedges is not effective and should be avoided. Long-leg bracing may be used for early tibia vara, but its effectiveness is uncertain. Avoid long-term bracing for conditions such as vitamin D-resistant rickets because the effectiveness of bracing is unclear and considerable disability results from brace treatment.

Operative correction options include osteotomy, or hemiarrest procedures either by hemiepiphysiodesis or unilateral physeal stapling. The objectives of operative treatment are to (1) correct knee angle, (2) place the articular surfaces of the knee and ankle in a horizontal position, (3) maintain limb length equality, and (4) correct any coexisting deformities. To achieve these directions, preoperative planning is required.

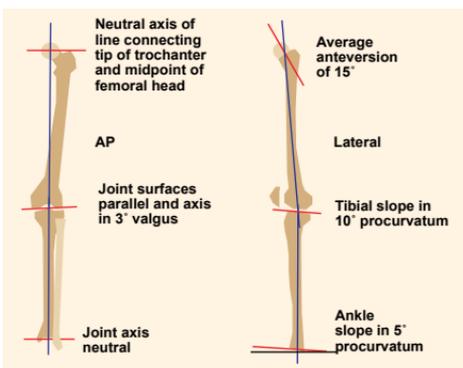
Mechanical axis Obtain a long, standing radiograph of the lower limbs. Be certain the child is positioned with the patella directly anterior when the exposure is made. Draw the axis of the femur and tibia connecting the center of the femoral head to the center of the distal femoral epiphysis [1]. Construct a second line between the midpoint of the upper and lower tibial epiphysis. Mark the articular surfaces. Measure the degree of valgus or varus.

Zone system On a full-length radiograph, draw a line between the femoral head and ankle. Note the position of the knee relative to this axis [2].

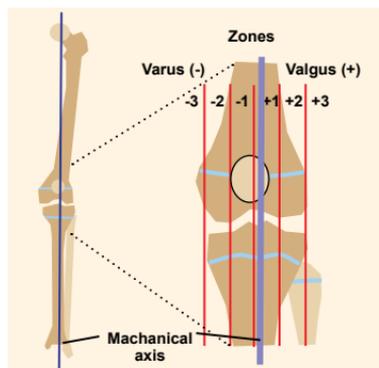
Make cutouts Before undertaking any osteotomy, make tracings of the bone and perform the intended osteotomy on the paper. This allows previsualizing the outcome and making necessary modifications.

Make corrective osteotomies as close to the site of deformity as practical.

Translation of the osteotomy may be necessary to position the joint within the mechanical axis.



1 Normal mechanical axis of lower limbs. These are average values. Based on Paley and Tetsworth (1992).



2 Zone system for assessing mechanical axis. The zone into which the mechanical axis falls is graded as a (-) for varus and a (+) for valgus, and into thirds with values ranging from 1 to 3. Based on Stevens et al (1999).

Multilevel osteotomies are often necessary in generalized deformities from metabolic conditions and osteochondrodystrophies. Balance the number of osteotomies with risks.

Recurrent deformity is likely, so delay each correction as long as possible to reduce the number of procedures required during childhood.

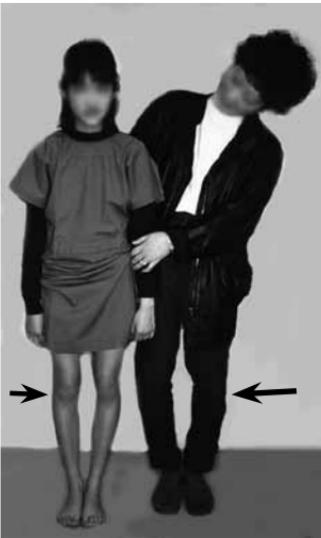
Idiopathic Genu Valgum and Valgum

Valgus deformity with an intramalleolar distance exceeding 8–10 cm is most common in obese girls. This deformity seldom causes function disability; the problem is primarily cosmetic. If severe, with an intramalleolar distance of >15 cm, consider operative correction by hemiepiphysiodesis or stapling. Make a standing radiograph and construct the mechanical axis. Determine the site(s) of deformity. In most cases the distal femur is most deformed at the appropriate site of correction.

Varus deformity is most common in Asians [1]. The varus deformity may be familial. Whether or not it increases the risk of degenerative arthritis of the knee is uncertain. This deformity seldom requires operative correction. Manage severe deformity by stapling or hemiepiphysiodesis.

Stapling

Stapling is a convenient method of correction [1 next page]. The disadvantages are the larger scar, the risk of staple extrusion, and a second operation for staple removal. The advantage is simplicity. The staples (usually 2) are placed, the patient carefully followed, and when the deformity is corrected the staples are removed. If the staples are placed extraperiosteal, growth can be expect to resume. The zone system [2 opposite page] is commonly used to determine the need for correction. A zone 3 deformity may be an indication for stapling. A rebound often occurs undoing some correction, so overcorrect slightly in anticipation of this common problem, especially in the children <12 years. See page xx.



1 Familial genu varum. Asians tend to have more varus than other groups.



2 Posttraumatic genu valgum. This deformity is due to overgrowth of the tibia following proximal tibial metaphyseal fracture.

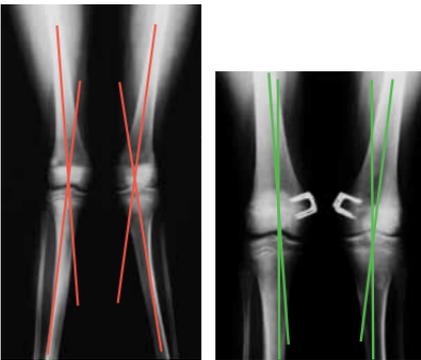
Hemiepiphysiodesis

With accurate timing, hemiepiphysiodesis has several advantages. The scar is short and the procedure simple and definitive. Bowen has developed a table to aid in timing [2]. Careful follow-up is essential because if the deformity appears to be destined for overcorrection, arresting the entire epiphysis becomes necessary.

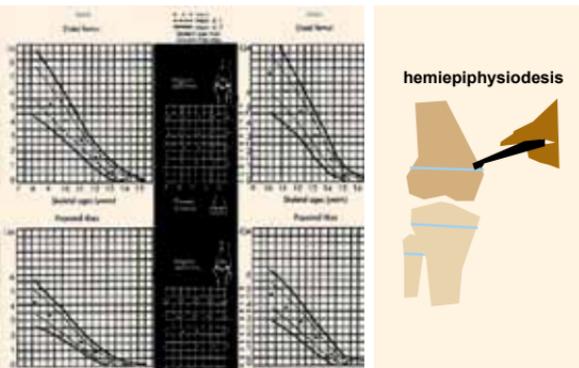
Pathologic Deformities

Posttraumatic genu valgum Posttraumatic genu valgum results from overgrowth following fracture of the proximal tibial metaphysis in early childhood [2 previous page]. Valgus may also be due to malunion or soft tissue interposition.

- **Natural history** The deformity develops during the first 12 to 18 months due to tibial overgrowth following the fracture. This is followed by a very gradual reduction of the valgus over a period of years. In the majority, this correction is adequate and no operative procedure is necessary.
- **Management** Manage proximal tibial fractures by correcting any malalignment and immobilize with a long-leg cast applied with gentle varus molding. Document reduction and position with a long film that includes the entire tibia. Advise the family of the potential of this fracture to cause a secondary deformity, which cannot be prevented. Avoid early osteotomy because recurrence is frequent and the deformity usually resolves spontaneously with time. Reassure the family that the knee will not be damaged by the deformity. Should the deformity persist, correct by osteotomy or by hemiepiphysiodesis or stapling near the end of growth.



1 Correction of excessive physiologic knock-knee by stapling. This 13-year-old girl had distal medial femoral stapling to correct this deformity. Note the knee angle before and after stapling.



2 Chart for timing hemiepiphysiodesis. This chart was created to time correction of angulatory deformities. From Bowen (1985).

Rickets

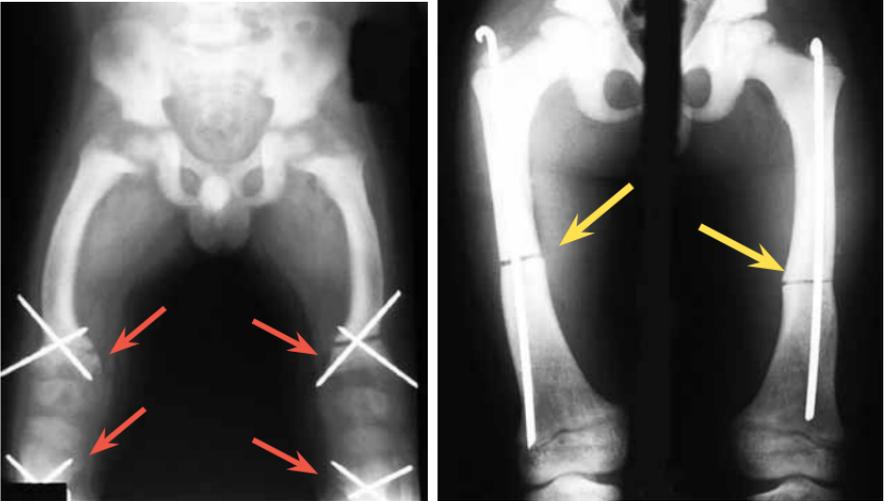
Suspect rickets in a child with increasing genu valgum, short stature, and a history of an atypical diet or similar deformities in other family members. Rickets produces a generalized genu valgum with bowing of the diaphysis and rarefaction of the epiphysis. Low calcium and phosphorus and a high alkaline phosphatase are confirming laboratory findings. Document severity with a 36-inch radiograph of the entire femur and tibia. Measure the hip-knee-ankle angle and mechanical axis zone.

Manage by first referring the child to an endocrinologist to optimize the medical management of the rickets. Despite optimal medical management, the deformities often persist in vitamin D-resistant forms of rickets.

Bracing The role of bracing is controversial as long-term bracing imposes a major added burden on the child and the value of bracing has not been shown.

Surgery If possible, delay correction until late childhood for stapling or adolescence for osteotomy. Correction at the end of growth reduces the risk of recurrence. If deformity is severe, correction may be necessary in childhood [1]. Plan osteotomy as discussed earlier with long films and cutouts. Drape the entire limb free to visualize adequacy of correction. Correct at one or more levels on each bony segment following the preoperative plan. Immobilize for about 10 weeks as healing may be slightly slower than normal.

If operative correction is performed before the end of growth, recurrence is common. Recurrence is most rapid in the younger child.



1 Operative correction of rickets deformity. Multilevel correction is often necessary (red arrows). In some, intramedullary fixation can be used (yellow arrows).

Tibia Vara

Tibia vara or Blount disease, is a growth disorder involving the medial portion of the proximal tibial growth plate that produces a localized varus deformity [1]. The incidence is greater if the child is black, obese, has an affected family, and resides in certain geographical locations such as the southeastern part of United States. The cause is unknown but it has been theorized that in susceptible individuals mechanical stress damages the proximal medial growth plate, thus converting physiologic bow legs into tibia vara.

Evaluation

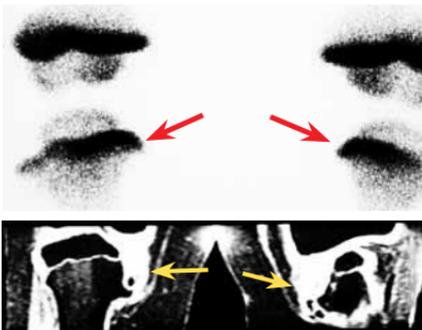
Two clinical patterns of tibia vara are seen. Radiographs in early infantile tibia vara may be difficult to differentiate from physiologic bowing. The metaphyseal–diaphyseal angle is often used. This angle [2] shows considerable overlap between physiologic and tibia vara cases. If the angle exceeds 15° , tibia vara is likely. Differentiation is made by following radiographs made every 3–6 months. Usually physiologic varus improves after the child's 2nd birthday. Tibia vara progresses and shows diagnostic metaphyseal changes.

Management

Treatment is based on the stage of the disease [2] and the age of the child.

Bracing As mild deformities may resolve without treatment, the beneficial effect of the brace is often uncertain. Often braces are used to treat stage 1 and 2 disease. If treatment is elected, order a long-leg brace with a fixed-knee that incorporates valgus loading. The brace should be worn during active play and at nighttime.

Osteotomy in the child If the disease progresses or is first seen in stages



1 Tibia vara. Bone scans early show increased uptake on the medial aspect of the proximal tibial epiphysis (red arrows). MR imaging of advanced disease shows cartilaginous replacement of the upper metaphysis (yellow arrows).



2 Common measures for tibia vara. The metaphyseal–diaphyseal angle (red arrow) and the Langenskiöld classification are commonly used.

3 and 4, osteotomy is indicated. Perform the osteotomy before age 4 years if possible [1]. Deformities of stages 5 and 6 are more complex and may require a double-level osteotomy to correct both the genu varum and the articular incongruity. Also assess the shape of the distal femur as varus or valgus deformity may contribute to the deformity. Medial tibial torsion is also a common associated deformity. Correct the varus and torsion by a simple closing wedge with rotation or by an oblique osteotomy. Correct the thigh-foot angle to about +10 degrees and overcorrect the varus to about 10 degrees of valgus. Use a sterile tourniquet so the entire limb can be seen to ensure appropriate correction. Release the anterior compartment fascia to reduce the risk of a compartment syndrome. Fix the osteotomy with crossed pins and supplement the fixation with a long-leg cast.

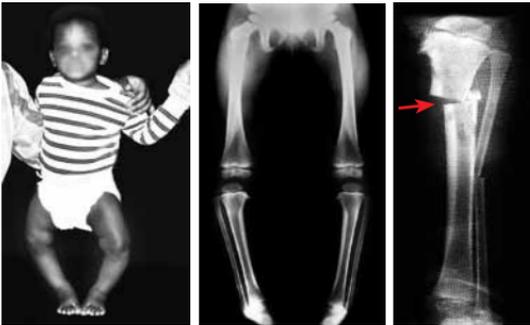
Hemistapling may be an alternative to osteotomy for stage 2 or 3 deformities.

Physal bridge resection Rarely a physal bridge is suspected in unilateral involvement in mid to late childhood. CT or MRI studies confirm the presence of the bridge. Resect the bridge, fill the defect with fat, and correct the tibial deformity by osteotomy.

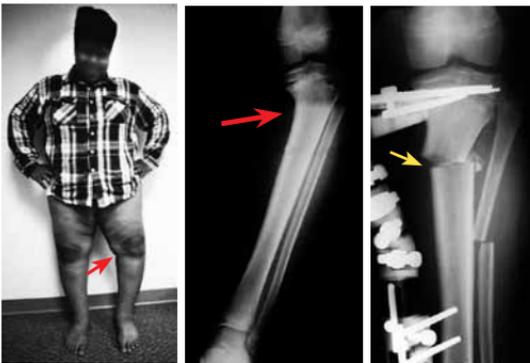
Surgery in the adolescent Operative correction in the older child or adolescent is usually complicated by obesity. Stabilize the osteotomy with an external fixator. External fixation provides adequate immobilization without need for a cast and allows the option to adjust alignment during the post-operative period [2].

Prognosis

The prognosis depends upon severity, stage, and treatment. Recurrence of varus and increasing shortening are common during childhood. Persisting articular deformity often leads to degenerative arthritis in adult life.



1 Infantile tibia vara. This disease is usually bilateral and may cause deformity that requires correction by osteotomy (red arrow).



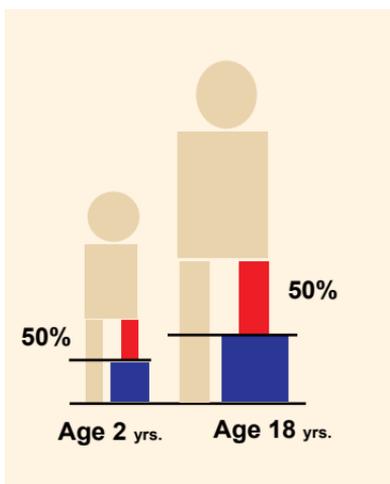
2 Adolescent tibia vara. This form is often unilateral (red arrows) and usually requires operative correction. External fixation is often an excellent choice in operative correction (yellow arrow).

Lower Limb Deficiencies

Lower limb deficiencies are rare deformities. The diagnosis and evaluation were covered in Chapter 2. Management of lower limb deficiencies is complex, requiring correcting length equalization, stabilizing unstable joints, and correcting angular and rotational deformity. Prudent management requires a balanced approach, balancing cosmetic and functional outcomes against risks and costs of surgery.

Principles of Management

1. Establish an accurate diagnosis. Refer to a children's limb deficiency clinic when available. Refer to a geneticist. Consider other problems.
2. Deal with the family's shock and guilt. Be positive. Most children can have a relatively normal childhood and can become independent and productive adults.
3. Plan a management strategy that is tailored to the unique deformities of the child and social and cultural values of the family.
4. Be prepared to deal with the family's preference to lengthening over amputation, even for deformities best managed by conversion and prosthetic fitting. Be prepared for the impact of the Internet, support groups and input from other parents on decision making.
5. Encourage the family to discuss management with others in support groups and medical centers.
6. Be prepared for the effect of growth.
7. Children rarely have phantom pain.
8. Amputations are well-tolerated in children, but peer and family issues often complicate management.
9. Children impose greater physical demands on prosthetics.



1 Growth in congenital deficiencies. Note that the percentage shortening of the limb remains constant throughout growth for congenital deficiencies.



2 Diaphyseal overgrowth. This causes "pencil" or penetration of the sharp end through the skin (arrow).



3 Syme amputation. This child had this conversion for fibular deficiency. This stump is endbearing.

Dealing with Deformity

1. Preserve length and growth plates.
2. Stabilize the proximal joints when possible.
3. Save the knee joint if possible.
4. Be prepared to deal with problems other than limb deficiency, as deformities are often complex.
5. Estimate roughly the anticipated magnitude of shortening at maturity to plan management [1 opposite page].
6. Limit lengthening to roughly 15% of the bone length.

Surgical Principles

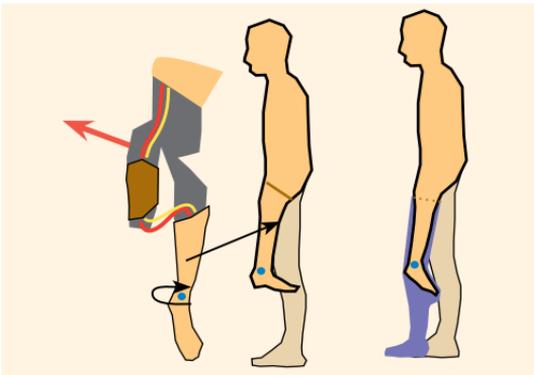
1. Perform disarticulations rather than transosseous amputations when possible to prevent diaphyseal overgrowth [2 opposite page].
2. The most commonly used procedures include: Syme amputation–disarticulation of ankle [3 opposite page]; Boyd amputation (midtarsal amputation); knee disarticulation; modified Van Nes rotationalplasty [1]; Brown procedure–centralization of the fibula and ankle fusion.
3. Coordinate operative and prosthetic management thoughtfully.

Tibial Deficiency

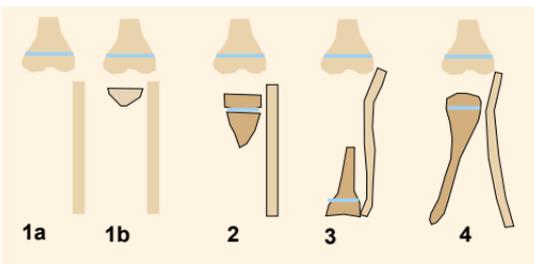
Tibial deficiency is a congenital hypoplasia or aplasia of the tibia. Classify the deformity based on the extent of loss [2]. This deficiency may be genetic. Refer to a geneticist for a consultation and counseling. Management is based on the adequacy of the upper tibial segment.

Adequate upper tibial segment Centralize the fibula under the tibia and disarticulate the ankle at about one year of age. Fit the child with a Syme prosthesis.

Inadequate upper tibial segment Usually management includes disarticulation of the knee and prosthetic fitting in late infancy or early childhood.



1 Van Nes rotationalplasty. Thigh resection was required for tumor eradication. Note that ankle (blue dot) is rotated 180° and becomes the knee within the prosthesis. Based on Krajbich (1998).



2 Tibial deficiency classification. Based on Jones, Barnes, and Lloyd-Roberts (1978).

Proximal Femoral Focal Deficiency

Proximal femoral focal deficiency or PFFD [1] includes a spectrum of deformities that may be associated with fibular deficiency. Differentiate PFFD from the congenitally short femur [2], which involves the shaft rather than the proximal part of the femur. Management of these conditions is different as lengthening is often appropriate for managing the congenitally short femur and rarely appropriate for PFFD.

Natural History

The normal and abnormal sides remain proportionately the same throughout growth. The limb length inequality is the most obvious source of problems. Less obvious but often significant is hip joint instability. Less significant is an external rotation deformity of the femur.

Evaluation

Study the shape of the acetabulum, the shape of the proximal ossification of the femur, and the length of the femur. Classify the deformity traditionally [1 opposite page] or as simply *short* or *too short*.



1 Proximal femoral focal deficiency. Note the shortening and lateral rotation.



2 Hamanishi's spectrum of the congenitally short femur. This figure shows the wide variation in deformities included in this classification. Based on Hamanishi (1980).

Deformity

As PFFD causes a combination of deformities, consider each as part of general management.

Length Length is a major problem. Calculate the estimated discrepancy at maturity to guide management. Base the management decision on severity.

Hip joint Predict hip status based on the volume of the acetabulum. A poor acetabulum suggests that the hip will be unstable. Only mild degrees of dysplasia are correctable. Be aware that an unstable hip joint jeopardizes the success of femoral lengthening.

Proximal femur A bulbous shape of the upper femur suggests that the proximal femur is complete, but with a varus deformity and slow ossification. In contrast a pointed and sclerotic upper femur suggests a more severe deformity. Perform arthrography early to determine the pathology [1 next page]. Correct the varus early to enhance ossification.

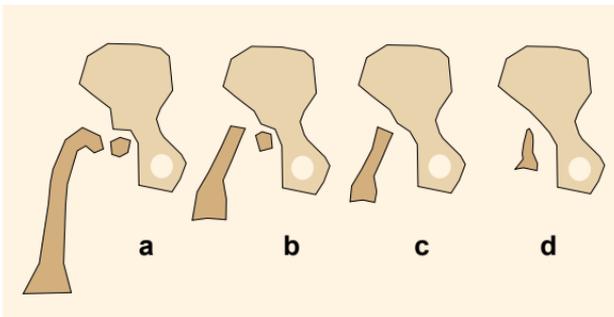
Timing of Correction

Correct proximal femoral deformity during the first year. Fit with temporary prosthesis by age 2. Staged lengthenings may be started as early as the second year. Rotationalplasties are best delayed until about age 4 years.

Procedures

Hip fusion is controversial. It provides stability for walking or lengthening procedures. It may make prosthetic fitting more difficult than the unfused mobile but unstable hip.

Subtrochanteric osteotomy is indicated for correction of varus. When associated with delayed ossification of the proximal femur, fusion and correction are more difficult.



1 Aiken's classification of proximal femoral focal deficiency.

Rotationalplasty includes excision of the knee joint and rotation the extremity 180° allowing the child's ankle to become the knee joint of the prosthesis. The procedure improves function but degrades appearance and is more suitable for males because clothing better hides the deformity.

Syme or Boyd amputation is indicated to allow prosthetic fitting with a knee disarticulation type of prosthesis. It is simple, cosmetic, easy prosthetic fit but at the sacrifice of efficiency.

Knee fusion is usually combined with Syme amputation to provide stability and to position the Syme stump just above the level of the opposite knee joint to facilitate prosthetic fitting.

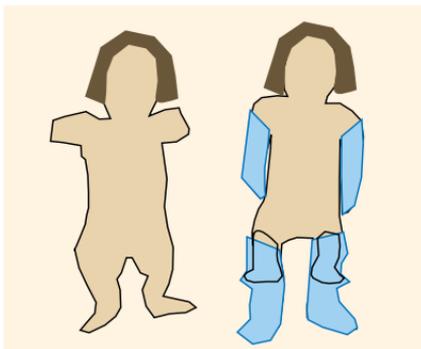
Femoral lengthening may be considered for a congenital short femur expected to be less than 10–20 cm at the end of growth. With increasing length gained, complications and risk of joint damage increase. Rapidly changing technology will undoubtedly increase feasibility of greater lengthening in the future.

Alternatives

Special situations such as bilateral deformity may make prosthetic management without conversion an option [2].



1 PFFD type b. Note the well-developed acetabulum (red arrow). The arthrogram shows the unossified femoral neck (yellow arrow) and severe varus.



2 Bilateral PFFD prosthetic fitting. Prosthetic fitting allows for increasing height and accommodates lateral limb positioning. Based on Krajbich (1998).

Fibular Deficiency

This deformity [1] is the most common of lower limb deficiency. It occurs sporadically and seldom has a genetic basis.

Pathology

There is partial or complete absence of the fibula [1 next page]. A fibrous analogue may replace osseous fibula. Fibular shortening causes lateral ankle instability. Tibial deformities may include shortening, anterior bowing, and valgus deformity. Foot deformities include absence of lateral portions of the foot [1 on page 162], talocalcaneal fusions, and ankle equinus.

Natural History

Shortening is progressive but remains proportionally shortened to the opposite normal side. Ankle instability results in deformity and pain in the second decade. Knee valgus may cause disability. Disability from shortening is proportional to severity.

Management

Classify type of deformity. Calculate roughly the expected shortening at skeletal maturation. Operative management is largely determined by the extent of foot deficiencies and ankle instability [1 and 2 on page 167]. Managing the deformity is often less difficult than effectively dealing with the family.

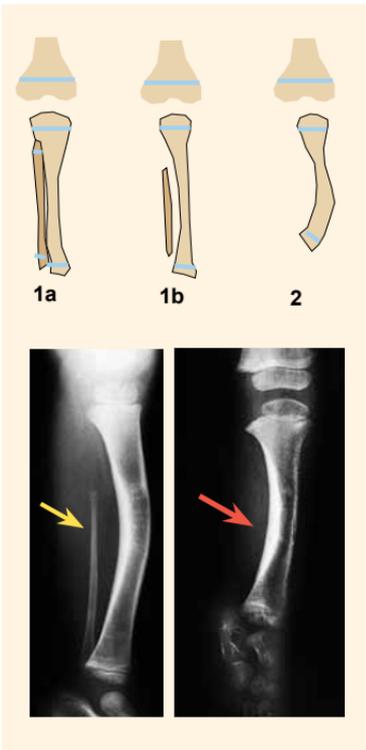


1 Foot deformity with fibular deficiency. The foot deformity cannot be corrected.

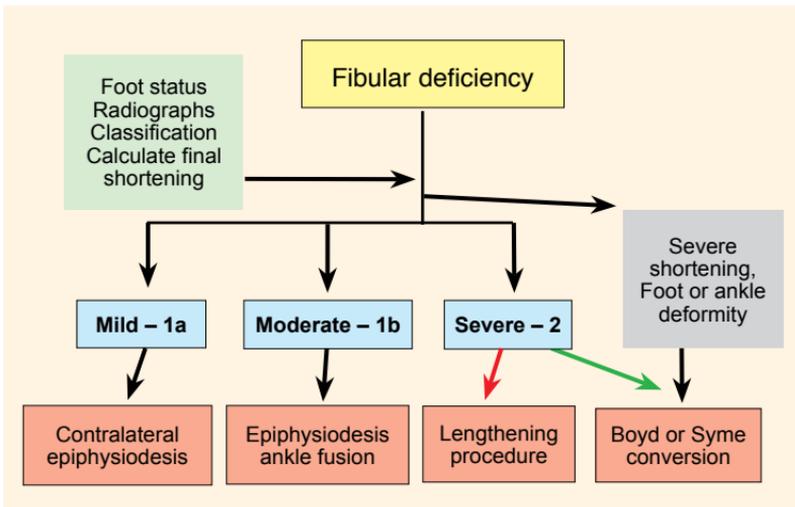
166 Lower Limb / Fibular Deficiency

Dealing with the family Families often have difficulty accepting Syme amputation and prosthetic management even for severe deformities. Families often wish to delay a decision in hope of new technology that will make amputation unnecessary or delay the decision until the child can participate in the decision. Families often use electronic communication with other families and may elect to visit centers where complex reconstructive procedures are offered. If the family cannot make a decision, provide a special prosthesis that can incorporate the foot while the decision is being made.

Operative management Plan amputation late in the first year just before the infant would normally stand and walk. Perform a Syme or Boyd procedure. The value of resection of the fibular analogue is controversial. Correct significant tibia vara to facilitate prosthetic fitting and walking. Lengthening is best delayed until midchildhood. A shoe lift may be necessary before lengthening. Make the lift light, least intrusive, and about one inch less in height than is necessary to level the pelvis.



1 Fibular deficiency classification. Clinical examples of type 1b (yellow arrow) and type 2 (red arrow). From Achterman and Kalamchi (1979).



1 Flowchart for managing fibular deficiencies.

	Amputation	Lengthening
Advantages	Definitive Few complications Resolves length and ankle problems	Attractive to family Restores body image
Disadvantages	Requires prosthesis Difficult to accept	Many complications Complex procedure Sometimes: retards tibial growth damages knee and ankle

2 Factors in making decision to lengthen or amputate. The choice between amputation for types 1b and 2 deformities and lengthening is often difficult. Consider these factors.

Chapter 5 – Foot

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Introduction

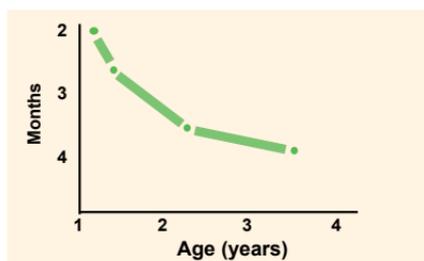
Developmental variations of the foot are common. Thus, they are a frequent source of concern to the family and a common reason for referral to an orthopedist.

Growth

The lower limb bud forms by about 4 weeks of gestation, and the foot develops over the next 4 weeks [1]. The foot achieves its adult length earlier than the rest of the body. Half of the adult length of the foot is achieved between 12 and 18 months of age. By comparison, half of adult height is achieved at 2 years and half of the lower limb length by 3 to 4 years of age. Rapid foot growth requires shoe changes frequently in infancy and childhood [2].



1 Fetal foot development. The limb bud appears by about 4 weeks of gestation and the foot is well formed by about 7 weeks.



2 Rapid foot growth. This graph shows the months required for a half size change in shoe size by year of age. From data by Gould et al. (1990).

Arch Development

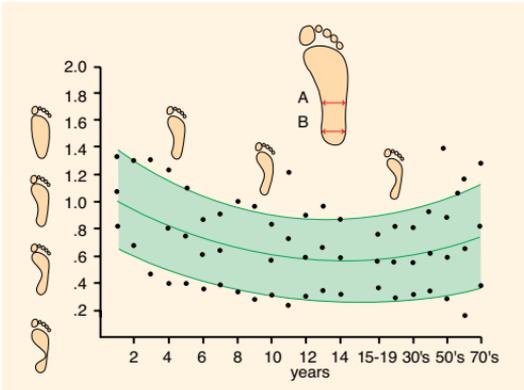
The longitudinal arch of the foot develops with advancing age [1]. The flatness of the infant's foot is due to a combination of abundant subcutaneous fat and joint laxity common in infants. This joint laxity allows flattening of the arch when the infant stands, and the fatty foot further obscures the longitudinal arch.

Normal Variability

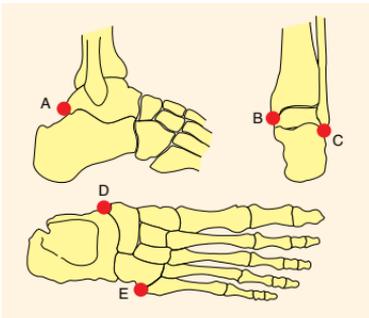
Accessory centers of ossification are common about the foot [2]. Most fuse with the primary center and become part of the parent ossicle. Others remain as separate ossicles, usually attached to the parent bone by cartilage or fibrous tissue. These ossicles are clinically important because they may be confused with a fracture, and they may become painful when the syndesmosis or synchondrosis is disrupted. Such disruptions commonly involve the accessory navicular and an ossicle inferior to the lateral malleolus.

Foot in Systemic Disorders

Evaluation of the foot is a useful aid in diagnosing constitutional disorders. For example, polydactylism is seen in chondroectodermal dysplasia. Dysplastic nails are found in the nail-patella syndrome.



1 Arch development. The longitudinal arch develops with growth in childhood. Note the wide range of normal. Flatfeet fall within the normal range. From Staheli et al. (1987).



2 Common accessory ossification centers about the foot. (A) Os trigonum (red arrow at top), (B) medial malleolar ossicle, (C) lateral malleolar ossicle, (D) accessory navicular (often painful), and (E) os vesalianum (yellow arrow at top).

Nomenclature

To clarify this discussion, terms describing joint motion versus those describing deformities are defined separately [1]. The anatomical position is considered neutral. Often deformity is designated simply by describing the motion and adding the term deformity behind it. Thus, the subtalar joint fixed in inversion is referred to as an inversion deformity. Note that the description of great toe position is inconsistent with standard terminology. The reference point is the center of the foot rather than the center of the body. Thus, the position of the great toe toward the midline of the body is referred to as abduction.

Both bones and joints may be deformed. For example, medial deviation of the neck of the talus occurs in clubfeet. This contributes to the adduction deformity. Joint deformity is usually due to stiffness with fixation in a nonfunctional position. Limit the use of the terms varus and valgus to describe deformities.

Site	Motion	Deformity
Ankle joint	Flexion	Equinus
	Extension	Calcaneus
Subtalar joint	Inversion	Heel varus
	Eversion	Heel valgus
Midtarsal joint	Adduction	Adductus
	Abduction	Abductus
	Flexion	Cavus deformity
	Extension	Rocker-bottom
	Pronation	Pronation deformity
	Supination	Supination deformity
Great toe	Abduction	Hallux varus
	Adduction	Hallux valgus
	Flexion	Flexion deformity
	Extension	Extension deformity
Toes	Flexion	Flexion deformity
	Extension	Extension deformity

1 Nomenclature for normal joint motion and deformity. Joint motion and deformity should be described independently.

Evaluation

Family History

Foot shape often runs in families [1]. If the deformity is present in an adult, inquiry about disability may help in managing the child's problem.

Screening Examination

Perform a screening examination. Look at the back for evidence of a spinal dysraphism that may account for a cavus foot. Assess joint laxity [2], as this may be a cause of a flexible flatfoot.

Foot Examination

The diagnosis of most foot disorders can be made by physical examination. Bones and joints of the foot have little overlying obscuring soft tissue, thus deformity and swelling are easily observed. Furthermore, localization of the point of maximum tenderness (PMT) is readily established.

Observation

Observe the skin on the sole of the feet for signs of excessive loading [3 this page and 1 opposite page]. Excessive loading that causes calluses is not normal in children. Common sites of excessive loading include the metatarsal heads, the base of the fifth metatarsal, and under the head of the talus. The deformities that cause the calluses are likely to cause pain in adolescence.

Observe the foot with the child standing. Note any change in the alignment of the heel. Heel valgus is common. Note the height of the longitudinal arch. Next ask the child to toe stand. A longitudinal arch is established in children with a flexible flatfoot [2]. With the child seated and the foot unweighted, a longitudinal arch appears in the child with a flexible flatfoot.



1 Familial hallux varus. Note the same deformity in the mother's and daughter's feet.



2 Generalized joint laxity. This child's thumb is easily opposed to the forearm. The child also had a flexible flatfoot.



3 Examine the sole for signs of excessive loading. Note the calluses under the metatarsal heads of both feet in this child with congenital toe deformities.

Range of Motion

Estimate the range of motion of the toes and the subtalar and ankle joints. Estimate subtalar joint mobility by the range of inversion and eversion motion. Assess ankle motion both with the knee flexed and extended and with the subtalar joint in neutral alignment [3]. Dorsiflexion to at least 20° with the knee flexed and to 10° with the knee extended should be possible.

Palpation

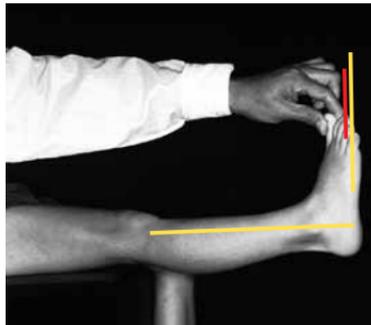
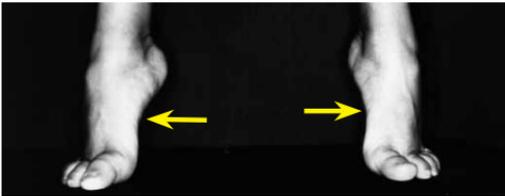
By palpation, determine if any tenderness is present. Determining the PMT is especially helpful in the foot because much of the foot is subcutaneous. The PMT is often diagnostic or at least helpful in making decisions regarding imaging.



1 Sole contact area. Note the broad even weight distribution on the soles of these normal feet. Child is standing on a mirrored glass surface.



2 Flexible flatfeet. The longitudinal arch absent on standing (red arrows) appears on toe standing (yellow arrows).



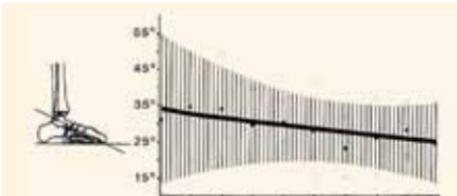
3 Assessing ankle dorsiflexion. Right angle (yellow) is neutral position. Assess dorsiflexion (red lines) with the knee flexed and extended to determine site and severity of triceps contractures.

Imaging

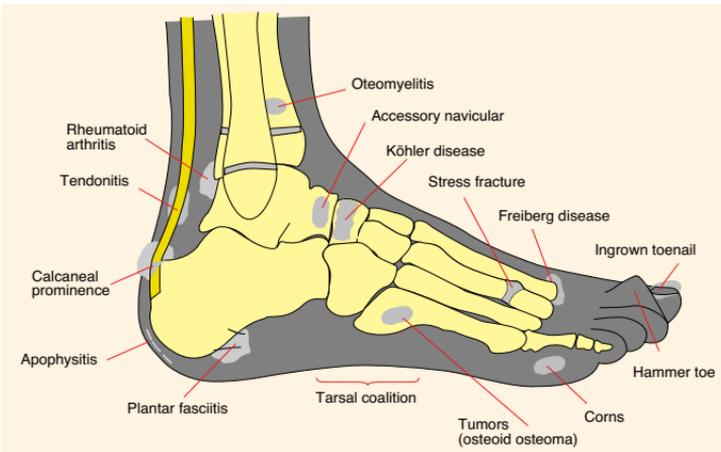
Whenever possible, radiographs of the feet should be taken with the child standing [1]. If radiographs are indicated, order AP and lateral projections. If subtalar motion is limited, an oblique view of the foot is added to rule out a calcaneonavicular bar. The ankle can be evaluated by AP and lateral radiographs. Order a "mortise" view if a problem such as an osteochondritis of the talus is suspected. Other special views such as flexion-extension studies may be helpful. Compare the radiographs to published standards for children. The normal range is broad and changes with age [2]. CT scans are useful in evaluating the subtalar joint for evidence of a talocalcaneal bar. Bone scans are useful in confirming the diagnosis of osteochondrosis such as Freiberg disease. The scan will be abnormal before radiographic changes are present. MRI is useful for evaluating tumors.



1 Standing radiographs. Standing radiographs allow the most consistent evaluation. In the adolescent with a skewfoot deformity, the talar inclination (yellow line), metatarsal axis (red line), and calcaneal pitch (orange line) are readily measured.



2 Inclination of the talus by age. The shaded area represents two standard deviations above and below the mean (heavy line). Note that the values change with age and that the normal range is very broad. From Vander Wilde, et al. (1988).



3 Foot pain localization. Because the foot is largely subcutaneous, localization of the tenderness will often aid in establishing the diagnosis.

Foot Pain

Foot pain in children is common and varied [3 opposite page and 1 this page]. During the first decade of life, foot pain is usually due to traumatic and inflammatory problems such as injuries and infections and is seldom due to deformity. During the second decade, foot pain is often secondary to deformity.

The cause of the foot pain can often be determined by the history and physical examination. Determining the PMT is especially useful about the foot because the structures are subcutaneous and easily examined [2]. This localization often allows a presumptive diagnosis.

Trauma

Stress–occult fractures Fractures without trauma history are not uncommon in infants and young children. They may be considered as part of the toddler fracture spectrum. Fractures of the cuboid, calcaneus, and metatarsal bones can be best identified by bone scans.

Tendonitis–fasciitis Repetitive microtrauma is a common source of heel pain in children. This is most common about the os calcis either from the attachment of the heel-cord or the plantar fascia.

Category	Disorder
Trauma	Fractures Sprains Soft tissue injuries Overuse syndromes
Infections	Osteomyelitis Septic arthritis Nail puncture wounds Ingrown toenail
Arthritis	Degenerative Juvenile rheumatoid Pauciarticular arthritis
Osteochondritis	Freiberg disease Köhler disease
Impingement pain	Os trigonum syndrome Anterior tarsal compression
Syndesmosis disruptions	Accessory navicular Lateral malleolar ossicle Medial malleolar ossicle
Idiopathic disorders	Osteochondritis dissecans Tarsal tunnel syndrome Reflex sympathetic dystrophy
Deformities	Bunions Bunionettes Tarsal coalitions Skewfoot Pathologic flatfeet

1 Classification of foot pain. The causes of foot pain can be placed in categories for classification and diagnosis.



2 Point of maximum tenderness. The ankle is swollen and tenderness is present just anterior to the distal fibula, typical of a sprain.

Infections

Infections of the foot are relatively common. Septic arthritis commonly affects the ankle and occasionally other joints of the foot. Osteomyelitis may occur in the calcaneus and other tarsal bones. Infection may be hematogenous or iatrogenic (heel sticks for blood sampling) or result from penetrating injuries.

Nail puncture wounds Nail puncture wounds are common injuries [1 and 2] that may be complicated by osteomyelitis [3]. About 5% of nail penetrations become infected, but less than 1% develop osteomyelitis. Puncture wounds over the metatarsal are more likely to be infected with pseudomonas septic arthritis. Infections in the heel are commonly from staphylococcus or streptococcus.

- Initial management* Examine the foot and remove any protruding foreign material. Probing the wound will be noisy and unrewarding. Update tetanus immunization. Inform the family about the risk of infection and the need to return if signs of infection occur. Usually infections will show signs several days after the injury and include increasing discomfort, swelling on the dorsum of the foot, and fever.

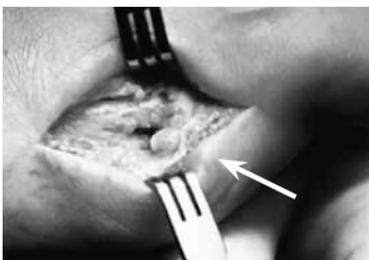
- Management of infection* Culture the wound and obtain an AP radiograph of the foot to serve as a baseline. The time of onset of signs of an infection suggests the infecting agent. If the interval between penetration and infection is 1 day, the organism is likely to be streptococcus. If the interval is 3–4 days, staphylococcus is most likely, and if a week, pseudomonas. Children with pseudomonas infections were usually wearing shoes at the time of the penetration. Operative debridement and drainage are indicated in all pseudomonas infections. Drainage is also indicated in all infections that fail to improve promptly with antibiotic treatment.



1 Nail puncture wound. Erythema and a puncture wound (arrow) show the site of nail entry. Swelling is most prominent on the dorsum of the foot.



2 Needle penetration into heel. Radiograph (left) shows broken needle. Photograph (right) shows site of entry of needle and surrounding inflammation.



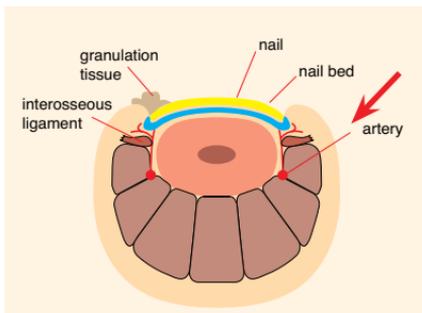
3 Chronic osteomyelitis. A nail puncture wound through a shoe was followed by this infection that involved the bone and joint (red arrow). The joint was destroyed. Chronic osteomyelitis of the first metatarsal (yellow arrow) is evident in the radiograph.

Ingrown Toenails Ingrown toenails [1] are common infections resulting from a combination of anatomical predisposition, improper nail trimming, and trauma. Injury or constricting shoes or stockings may initiate the infection. In children prone to developing this problem, the nail is abnormal, often showing a greater lateral curvature of the nail into the nailbed.

- *Management of early infections* Choose treatment based on the severity of the inflammation. Mild irritation requires only proper trimming of the nail and properly fitting shoes. Nails should be trimmed at right angles. Avoid trimming the nail to create a convex end. Instruct the family to trim the nail to create a concave end that leaves the nail edges extending beyond the skin to prevent recurrent ingrowth. Elevate the nail from the bed by packing cotton under the nail edge. Repeat this several times if necessary to lift the nail out of the inflamed nailbed. If inflammation is more severe, rest, elevation, protection from injury, soaking to clean and promote drainage, and antibiotics may be necessary.
- *Management of late infections* Persistent severe lesions require operative management. The hypertrophic chronic granulation tissue is excised, and removal of the lateral portion of the nail together with a portion of the nail matrix may be necessary to prevent recurrence.

Pauciarticular Arthritis

Pauciarticular arthritis may present with foot pain in the infant or young child. A limp, limited ankle or subtalar motion and swelling for more than 6 weeks duration suggests this diagnosis [2].



1 Ingrown toenails. Granulation tissue grows over the site of nail penetration (arrow). Photograph shows typical clinical appearance (yellow arrow).



2 Arthritis of the ankle. This child has pauciarticular arthritis of the ankle.

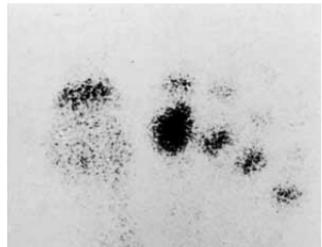
Osteochondritis

Köhler disease Tarsal navicular osteochondritis, also known as Köhler disease, is an avascular necrosis most common in boys between 3 and 5 years of age [1]. It also occurs uncommonly in girls 2 to 4 years of age. The disease produces inflammation, localized tenderness, and a limp. Radiographic changes depend on the stage of the disease. The navicular first shows collapse and increased density. Patchy deossification follows. Finally, the navicular is reconstituted. Because healing occurs spontaneously, only symptomatic treatment is necessary. If pain is a significant problem, immobilize the foot in a short-leg walking cast for 8 weeks to reduce inflammation and provide relief of pain. Long-term follow-up studies show no residual disability.

Freiberg disease Metatarsal head osteochondritis, also known as Freiberg disease or infraction, is an idiopathic segmental avascular necrosis of the head of a metatarsal. It most commonly occurs in adolescent girls and involves the second metatarsal. Pain and localized tenderness [2] are common. If the patient is seen early, a bone scan will show increased uptake and establish the diagnosis. Later, radiographs will show irregularity of the articular surface, sclerosis, fragmentation, and finally reconstitution. Residual overgrowth and articular irregularity may lead to degenerative changes and persistent pain. Treat with rest and immobilization to reduce inflammation. An orthosis to unweight the involved metatarsal head, sole stiffeners to reduce motion of the joint, and even a short-leg walking cast may be useful. For severe persisting pain, operative correction may be necessary. Options include joint debridement, excisional arthroplasty (proximal phalanx), interposition arthroplasty using the tendon of the extensor digitorum longus, and dorsiflexion osteotomy of the metatarsal (often the best choice).



1 Köhler disease in a 5-year-old boy. The tarsal navicular of the left foot is sclerotic (arrow) and the site of tenderness.



2 Freiberg disease. Tenderness over the head of the second metatarsal is usually present in Freiberg disease. Minimal changes are present in the radiograph (yellow arrow), and the bone scan shows increased uptake over the second metatarsal head (red arrow). The photograph shows the point of maximum tenderness (blue arrow).

Sever disease Calcaneal apophyseal osteochondrosis, also known as Sever disease, is commonly diagnosed by heel pain and radiographic features of fragmentation and sclerosis of the calcaneal apophysis. These radiographic changes occur commonly in asymptomatic children [1]. Most heel pain in children is due to inflammation of the attachment of the plantar fascia or heel-cord.

Impingement Pain

Os trigonum syndrome Compression of the ossicle in dancers often causes foot pain.

Anterior tarsal impingement Anterior foot pain is sometimes due to compression of articular margins often secondary to tarsal coalitions or heel-cord contracture [2].

Syndesmosis Disruption

Disruption of the syndesmosis between the primary ossicle and secondary centers of ossification (accessory ossicles) is a common cause of pain in the feet of children and teenagers. This disruption is the equivalent of a stress injury of the cartilaginous or fibrous attachment. This disruption becomes painful unless healing is complete. The condition commonly recurs.

Accessory navicular is an accessory ossification center on the medial side of the tarsal navicular that occurs in about 10% of the population and remains as a separate ossification center in about 2% [1 next page]. Disruptions are common during late childhood and adolescence and are probably due to repetitive trauma. This disruption causes pain and localized tenderness. This pain may cause inhibition of the function of the posterior tibialis muscle with secondary lowering of the longitudinal arch. Radiographs show an accessory ossicle and reduction of the calcaneal pitch in some cases.

Manage with a short-leg cast or splint. If the problem persists, excision of the accessory navicular may be necessary. Simple excision with or without plication of the posterior tibialis tendon is as effective as the more extensive



1 Calcaneal apophysis. This apophysis often shows a sclerotic pattern in the normal child.



2 Impingement. This is talar beaking associated with a tight heel-cord and impingement.

Kidner procedures, which require rerouting the tendon.

Malleolar ossicles Ossification centers occur below the medial and lateral malleoli. Persisting ossicles under the lateral malleolus are most likely to be painful [2]. Manage first by cast immobilization. Rarely, excision or stabilization by internal fixation is necessary.

Idiopathic Disorders

Osteochondritis dissecans talus Ankle pain, swelling, stiffness, and a trauma history suggest this diagnosis. Confirm with radiographs [1 and 2]. Manage with activity modification, immobilization, nonsteroidal anti-inflammatory drugs (NSAIDs), and time. Most will resolve. Lesions that are lateral with sclerotic margins and separated are more likely to require surgery. Remove loose bodies. The value of drilling is uncertain. Replace and fix large lesions.

Tarsal tunnel syndrome Foot pain, Tinel’s sign over tarsal tunnel, dysesthesias, and delayed nerve conduction suggest this diagnosis. This syndrome differs in children. Typically the child is female, walks with the foot in varus, may use crutches, and often requires operative release.

Reflex sympathetic dystrophy This syndrome usually affects the lower limb in girls [3]. Consider this diagnosis when the foot is swollen, stiff, cool, and generally painful. A history of injury is common. See Chapter 3 for management.

Foot Deformities

Foot deformities may cause pain due to pressure over bony prominences [4] or to altered mechanics of the foot. Pain from deformity is usually not difficult to recognize.



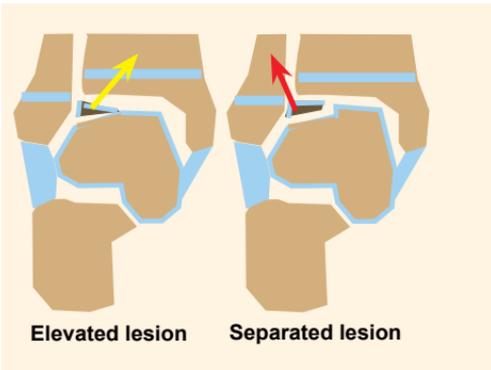
1 Accessory navicular. The accessory navicular is located on the medial aspect of the foot (arrows). It often produces a prominence and is sometimes painful.



2 Lateral malleolar ossicle. This ossicle was painful, not improved by casting, and eventually required fusion with screw fixation.



1 Medial osteochondritis dissecans lesion. Medial lesions are less common and often unassociated with trauma.



2 Lateral osteochondritis dissecans lesion types. A lesion that is elevated (yellow arrow) or separated (red arrow) may require surgery.



3 Reflex sympathetic dystrophy. The varus deformity of the left foot is due to reflex sympathetic dystrophy.



4 Foot deformities in a child with spina bifida. Prominence of the head of the talus caused skin breakdown in the foot with diminished sensation.

Toe Deformities

Generalized Disorders

If a toe deformity is found, carefully examine the hands and feet of the child and of the parents. These deformities are sometimes manifestations of a generalized disorder [1].

Cleft Foot Deformity

This rare deformity is transmitted as an autosomal dominant trait, is usually bilateral, and often involves the hands and feet [2]. A noninherited form is less common and is often unilateral. If it causes shoe-fitting problems, correct in late infancy or childhood by osteotomy and soft tissue approximation.

Microdactyly

Small toes are often found in Streeter's dysplasia and may be secondary to intrauterine hypotension, causing insufficient circulation to the toes [3]. No treatment is required.

Syndactyly

Fusion of the toes produces no functional disability and treatment is unnecessary. Look for some underlying problem.

Toe Deformities	Disorders
Polydactyly	Chondroectodermal dysplasia Carpenter syndrome Oto-Palato-Digital syndrome
Syndactyly	Apert syndrome
Metatarsal hypoplasia	Achondrogenesis Brachydactyly syndrome
Broad toe	Acromesomelic dysplasia Larsen syndrome Rubinstein-Taybi syndrome

1 Syndromes associated with toe deformities.



2 Cleft foot deformity. These deformities are present in both the father and the son. The major problems are difficulties with shoe fitting and the unusual appearance.



3 Microdactyly.

Polydactyly

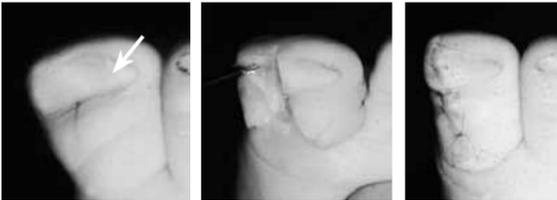
Polydactyly or supernumerary digits are common [1]. They are most common in girls and in blacks and are sometimes inherited as an autosomal dominant trait. Most involve the little toe and duplication of the proximal phalanx with a block metatarsal or wide metatarsal head. Excise the extra digit late in the first year when the foot is large enough to make excision simple and before the infant is aware of the problem. Plan the procedure to minimize the scar, establish a normal foot contour, and avoid disturbing growth. Central duplications often cause permanent widening of the foot. Poor results are more likely for great toe duplications with persistent hallux varus and complex deformities [2 and 3].

Curly Toes

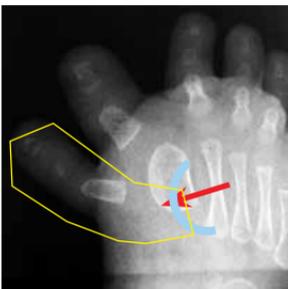
Curly toes [1 next page] are common in infancy and produce flexion and rotational deformities of the lesser toes. These deformities improve with time. Most will resolve spontaneously. Some advise tape splinting [2 next page], and, rarely, flexor tendinotomy is required for persisting deformity.



1 Polydactyly. Polydactyly causes a cosmetic and shoe fitting problem. Excision of the accessory toe is appropriate late in the first year.



2 Excision of bifid great toe. Half of the toe is removed. Excision of complex polydactyly is sometimes difficult.



3 Bracket epiphysis. Toe deformities may be complex. In this case, the metatarsal epiphysis is continuous for both toes. Excise the accessory digit and the adjacent portion of the growth plate.

Overriding Toes

Overriding toes are common. Overriding of the second, third, and fourth toes usually resolves with time. Overriding of the fifth toe is more likely to be permanent [1 opposite page] and cause a problem with shoe fitting. Overriding of the fifth toe is often bilateral and familial. If overriding becomes fixed, persists, and causes shoe-fitting problems, operative correction is appropriate. Correct with the Butler soft tissue alignment procedure.

Hammer Toes

Hammer toes are secondary to a fixed flexion deformity of the proximal interphalangeal (PIP) joint [2 opposite page]. The distal joint may be fixed or flexible. The condition is often bilateral, familial, and most commonly involves the second toes and less frequently the third and fourth. Operative correction is indicated in adolescence if the deformity produces pain or shoe-fitting problems. Correct by fusing the IP joint.

Claw Toes

Claw toes are usually associated with a cavus foot and are often secondary to a neurologic problem. Correction is usually part of the management of the cavus foot complex.

Hypertrophy

Hypertrophy [3 opposite page] is seen in children with Proteus syndrome, neurofibromatosis, or vascular malformation, or it can occur as an isolated deformity. Most show abnormal accumulation of adipose tissue, and some show endoneural and perineural fibrosis and focal neural and vascular proliferation. Management is difficult. Epiphysiodesis, debulking, ray resection, and through-joint amputations are often necessary. Recurrence is frequent, and several procedures are often required during childhood to facilitate shoe fitting.



1 Curly toes. This deformity may involve one or more toes and resolves spontaneously.



2 Taping of overlapping curly toes Paper tape is applied loosely to align toes (red arrow). Regardless of treatment, correction occurs over time (green arrow).



1 Overlapping toe. This overlapping fifth toe persisted and required operative correction.



2 Hammer toe. The fixed flexion deformity of the PIP joint of the second toe causes a callus (arrow) to form over the toe.



3 Hypertrophy. Overgrowth may cause severe shoe-fitting problems and require resection, epiphysio-desis, or amputation.

Forefoot Adductus

Metatarsus Adductus and Varus

Adductus of the forefoot is the most common foot deformity. It is characterized by a convexity to the lateral aspect of the foot [1] or a dynamic abduction of the great toe [2]. The deformities fall into four categories [3].

Metatarsus adductus is a common intrauterine positional deformity. Because it is associated with hip dysplasia in 2% of cases, a careful hip evaluation is essential. Metatarsus adductus is common, flexible, benign, and resolves spontaneously.

Metatarsus varus is an uncommon rigid deformity that often persists and requires cast correction. Metatarsus varus does not produce disability and does not cause bunions, but it does produce cosmetic and occasionally shoe-fitting problems.

Great toe abduction is a dynamic deformity due to overactivity of the great toe abductor. Sometimes called a "searching toe." The condition improves spontaneously. No treatment is required.



1 Metatarsus adductus. A convexity of the lateral border of the foot (red line) is the most consistent feature of this deformity.



2 Great toe abduction. This is a dynamic deformity that resolves with time.

Type	Etiology	Comment
Metatarsus adductus	Late intrauterine, positional deformity	Common form 90% resolve spontaneously
Metatarsus varus	Earlier onset, intrauterine position?	Often rigid Cast correction necessary
Skewfoot	Familial Generalized joint laxity	Hindfoot valgus Abduction midfoot Adduction forefoot Treatment difficult
Abducted great toe	Unknown	Dynamic deformity Resolves spontaneously

3 Types of forefoot adductus and varus deformities. The differential diagnosis of metatarsus adductus should include the rigid, nonresolving form, and the skewfoot.

Management

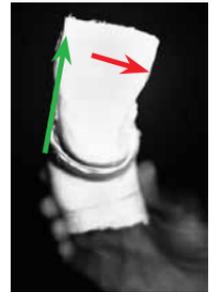
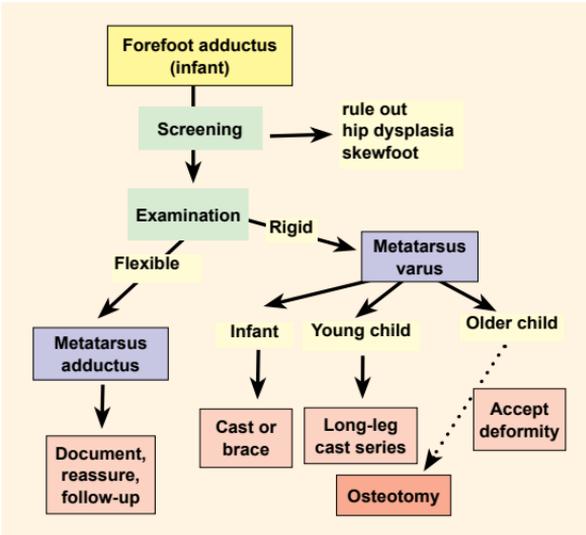
Evaluate by performing a screening examination, test for stiffness, and consider the child's age [1]. Manage metatarsus adductus by documentation and observation.

Manage metatarsus varus by serial casting [2 and 3] or bracing. Long-leg bracing is useful in the toddler. Serial casting is most effective. The deformity yields much more rapidly when the cast is extended above the flexed knee.

The following technique is useful to about age 5 years. Apply a short-leg cast first. As the cast sets, mold the forefoot into abduction. Finally, while holding the short-leg cast in external rotation and with the knee flexed about 30°, extend the cast to include the thigh. This long-leg cast allows both walking and effective correction.

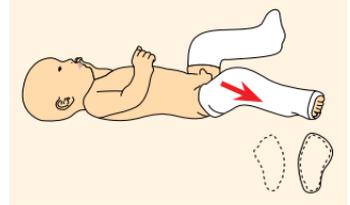
In the older child, it may be best to accept the deformity, as it does not cause disability. If operative correction is selected, correct by osteotomy rather than capsulotomy. Correction is not simple and complications are common.

1 Steps in managing forefoot adductus.



3 Cast treatment of metatarsus varus.

This child with a persisting stiff deformity was corrected using these long-leg casts. The knee is flexed to about 30° to control rotation. The foot is abducted in the casts. The casts are changed every 2–3 weeks until correction is completed.



2 Long-leg cast for metatarsus varus.

This treatment of metatarsus varus is most effective, as the flexed knee provides control of tibial rotation. Using the thigh portion of the cast as a point of fixation (yellow arrow), the foot is laterally rotated (green arrow) and abducted (red arrow) to achieve the most effective correction.

Skewfoot

Skewfoot, *Z-foot*, and *serpentine foot* are all terms given to a spectrum of complex deformities. This deformity includes hindfoot plantarflexion, midfoot abduction, and forefoot adduction [1]. A tight heel-cord is usually present in symptomatic cases. Skewfeet are seen in children with myelodysplasia; they are sometimes familial but are usually isolated deformities. There is a spectrum of severity. Some practitioners describe overcorrected clubfeet as skewfeet. Idiopathic skewfeet may persist and cause disability in adolescence and adult life.

Manage idiopathic skewfeet in young children by initial documentation with standing AP and lateral radiographs and observe to determine the effect of growth on the deformity. Most will persist. Plan correction in late childhood with heel-cord lengthening and osteotomies. Lengthen the calcaneus and first cuneiform [2].



1 Skewfoot deformity. Note the forefoot adduction in the photograph, the plantar flexed talus (red arrow), and the Z alignment (orange lines).



2 Sequence of correction of skewfoot. Calcaneal and cuneiform osteotomies and a heel-cord lengthening were performed. Note the changes in talar alignment between the preoperative (red arrows) and postoperative (orange arrows) radiographs.

Forefoot Deformities

Bunion

A bunion is a prominence of the head of the first metatarsal [1]. In children they are usually due to metatarsus primus varus, a developmental deformity characterized by an increased intrametatarsal angle [2] that exceeds about 9° between the first two rays. Hallux varus is a secondary deformity probably due to the effect of wearing shoes, as the great toe must be positioned in valgus to fit within a shoe. The normal hallux valgus angle is $<15^\circ$. The combination of primary and secondary deformities cause the typical adolescent bunion.

Bunions are often familial [1 next page] and may occur in children with neuromuscular disorders [2 next page]. Other factors may include pronation of the forefoot, joint laxity, and pointed shoes. Bunions are rare in barefoot populations.

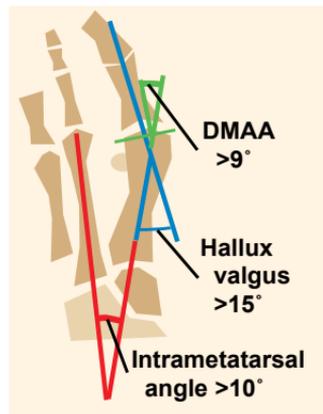
Evaluation Look for evidence of joint laxity, heel-cord contracture, pes planus, or other skeletal defects. Is the toe rotated? Is there a family history of bunions? Order AP and lateral standing radiographs. Measure the intermetatarsal angle. Measure the distal metatarsal articular angle (DMAA). This is normally less than 8° . Is the metatarsal-phalangeal joint subluxated? Is the cuneiform-metatarsal articulation oblique? Note the relative lengths of the first and second rays.

Management Attempt to delay operative correction until the end of growth to reduce the risk of recurrence.

- *Shoes* Encourage girls to avoid shoes with pointed toes and high heels, as they aggravate the deformity and increase discomfort.
- *Splints* For nighttime use, splints may be effective but are difficult to use because of the required duration of management [3 next page].
- *Operative correction* Tailor the choice of operation based on the pathology. There are many possible procedures.



1 Mild bunion deformities. The bunion is a prominence over the head of the first metatarsal. The right bunion (arrow) is most prominent here, and both are relatively mild. Usually no active treatment is required for bunions of this severity.



2 Measurements for bunion assessment.

Complications Ray shortening, elevation or depression of metatarsal heads, subluxation of the metatarsal phalangeal joint, and overcorrection or undercorrection are all possible complications of operative treatment.

Dorsal Bunion

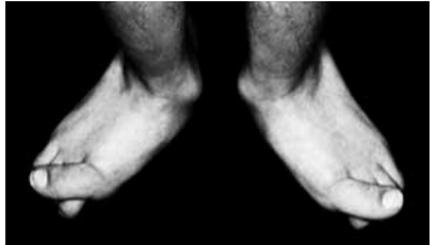
The uncommon dorsal bunion [1 opposite page] is due to an elevation of the first metatarsal. This elevation is caused by an imbalance between the stronger tibialis anterior and peroneus longus muscles. It is most common in clubfeet. This can be corrected by a plantar flexion osteotomy of the cuneiform or metatarsal and a muscle-balancing procedure.

Bunionette

Bunionette (tailor's bunion) is a painful bony prominence on the lateral side of the fifth metatarsal head, often associated with an inflamed thickened bursa and calluses. These deformities are developmental and involve an increased metatarsalphalangeal angle of the fifth toe, an increased intermetatarsal angle between fourth and fifth toes, and an increased intermetatarsal angle between the great and second toes. Management often requires an osteotomy for correction.



1 Familial bunions. Both mother and daughter have bunions.



2 Severe hallux valgus in cerebral palsy. Note the absence of metatarsus primus varus and any prominence of the metatarsal head.



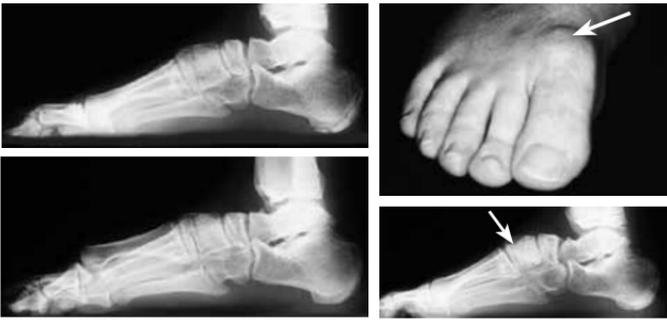
3 Treatment of juvenile bunions. Night splinting or operations are options. This child had metatarsus primus varus corrected by osteotomy and fixed with crossed pins (arrow).

Hallux Rigidus

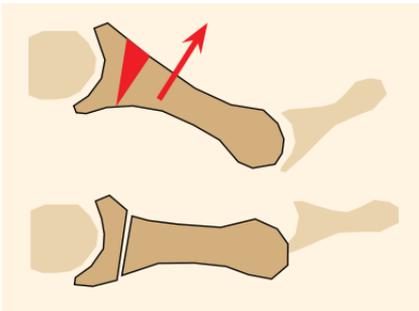
This is a degenerative arthritis of the first metatarsal phalangeal joint due to repetitive trauma, which causes stiffness, limited dorsiflexion, and pain. Manage by protecting the joint with a shoe stiffener. If severe and persistent, correct by a dorsal dorsiflexion osteotomy [2] to move the arc of motion into more extension.

Short Metatarsal

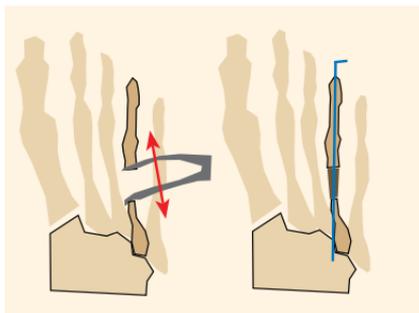
Shortening of one or more metatarsals may be due to a developmental abnormality as part of a generalized disorder or from trauma, infection, or tumors. Severe shortening may cause metatarsalgia and a cosmetic disability. Rarely the deformity is severe enough to justify operative correction. This can be done by a single-stage lengthening technique [3] or by gradual distraction histiogenesis.



1 Dorsal bunion. This child's bunion developed between the ages of 14 (yellow arrow) and 18 years (red arrows). The deformity was corrected by a dorsal opening wedge osteotomy (orange arrow) and lateral transfer of the anterior tibialis.



2 Dorsal osteotomy for hallux rigidus. A dorsal closing wedge osteotomy moved the arc of motion into more plantarflexion at the metatarsal phalangeal joint.



3 Technique of metatarsal lengthening. This one-stage lengthening is described by Baek and Chung (1998).

Clubfoot

Clubfoot (CF) is a complex congenital deformity that includes components of equinus, varus, adductus, and medial rotation [1]. Clubfoot is also referred to as *talipes equinovarus* (TEV). Clubfoot occurs in about 1 in 1000 births, is bilateral in half the cases, and affects males more frequently.

Etiology

The cause of clubfeet is multifactorial. In affected families, clubfeet are about 30 times more frequent in offspring. Fetal ultrasound screening shows the deformity in the first trimester [2]. It is frequently associated with other congenital abnormalities such as neural tube defects, anomalies of the urinary or digestive system, and other musculoskeletal abnormalities. Most show polyhydramnios, and amniocentesis often shows abnormal karyotype. The clubfoot deformity can have many etiologies, as evidenced by the variability of expression and response to management.

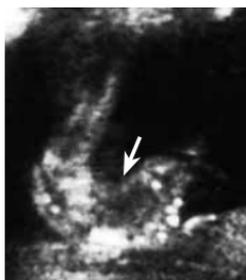
Mild clubfoot is a late intrauterine deformity (see Chapter 1) and corrects rapidly with cast treatment. At the other end of the severity spectrum, severe clubfoot behaves like a disruption, having an origin earlier in fetal life and requiring operative correction. Severe clubfoot is seen in conditions such as arthrogryposis [3]. Classic or idiopathic clubfoot is a polygenic disorder, is relatively common, and occupies the middle range of the severity spectrum.

Pathology

The pathology of clubfoot is typical of a dysplasia. The tarsals are hypoplastic. The talus is most deformed; the size is reduced and the talar neck is shortened and deviated in a medial and plantar direction. The navicular articulates with the medial aspect of the neck of the talus due to the abnormal shape of the talus. The relationship of the tarsals is abnormal. The talus and calcaneus become more parallel in all three planes. The midfoot becomes more medially displaced and the metatarsals are adducted. In addition to the deformities of cartilage and bone, the ligaments are thickened and the muscles hypoplastic. This results in a generalized hypoplasia of the limb with shortening of the foot and smallness of the calf [1 opposite page]. Because the hypoplasia primarily involves the foot, limb length discrepancy is usually less than 1 cm. The foot is small, and split size shoes are often required. The amount of foot shortening is proportional to the severity of the clubfoot.



1 Typical appearance of bilateral clubfeet. The deformity includes equinus, adductus, varus, and medial rotation.



2 Clubfoot on ultrasound at 16 weeks.



3 Severe clubfoot. Note the prominent medial crease (arrow).

Natural History

Untreated, clubfoot produces considerable disability [2]. The dorsolateral skin becomes the weight-bearing area. Calluses form and walking becomes limited. Treated clubfoot often causes minor disability from excessive loading on the lateral aspect of the foot due to residual varus and stiffness.

Clinical Features

The diagnosis of clubfoot is not difficult and is seldom confused with other foot deformities. Sometimes severe metatarsus varus is confused with clubfoot, but the equinus component of clubfoot makes the differentiation clear. The presence of a clubfoot should prompt a careful search for other musculoskeletal problems.

Examine the back for evidence of dysraphism, the hips for dysplasia, and the knees for deformity. Perform a screening neurologic examination. Note the size, shape, and flexibility of the feet. Take radiographs of the spine or pelvis if abnormalities are found on physical examination. If the infant is 6 months of age or older, initial AP and lateral radiographs are useful to supplement the physical examination. Measure the AP and lateral talocalcaneal angles. Compare these with the normal values (see Chapter 17). MRI studies will show the cartilaginous elements but are not practical for routine use.

Note the degree of stiffness of the foot [1 next page] and compare the size of the foot with the uninvolved foot. Marked differences in foot length suggest that the deformity is severe and foretell the need for operative correction. Document the components of the clubfoot deformity, the equinus, heel varus, forefoot adductus, and medial rotation. In the child, cavus is common.



1 Reduction in leg size. (Left) This girl with bilateral clubfeet has bilateral calf hypoplasia. (Above) The left clubfoot is corrected, but the foot is significantly shorter than the normal one. The degree of hypoplasia parallels the severity of the clubfoot deformity.

2 Untreated clubfeet. These are the feet of a 13-year-old Cambodian boy with untreated clubfeet. A large callus and bursa have formed over the site of weight bearing on the dorsum of each foot.

Equinus is due to a combination of a plantar flexed talus, posterior ankle capsular contracture, and shortening of the triceps.

Varus results from frontal plane parallelism of the talus and calcaneus, contracture of the medial subtalar joint capsules, and contracture of the posterior tibialis muscle.

Adductus and medial rotation are due to medial deviation of the neck of the talus, medial displacement of the talonavicular joint, and metatarsus adductus. Tibial rotation is normal. Tibial or femoral torsion are not primary features of clubfeet.

Classification

A number of classifications have been proposed.

Etiologic classification This is based on the possible causes of clubfoot and include several types.

- *Idiopathic clubfeet* include the classic forms with an intermediate degree of stiffness. This category is further divided into categories:
- *Postional clubfoot* are very flexible and correct with a few casts.
- *Idopathic clubfoot* are the classic clubfeet. If treatment is delayed they require more casts. These feet may recur. If treated by non-Ponseti cast techniques or by surgery the outcome is compromised.
- *Non-idiopathic clubfoot* include several categories
- *Syndromatic clubfeet* associated with condition such as arthrogryposis.
- *Neurogenic clubfoot* are associated with conditions such as myelodysplasia.
- *Other* such as rigid, teratologic (tarsal coalitions), associated with Streeter dysplasia, etc.



1 Testing flexibility. Assessing flexibility is a good way to determine the severity of the clubfoot.

Imaging Studies

Radiographs, ultrasound, and MRI imaging are used for assessment. Because active treatment usually occurs during early infancy when ossification is incomplete, the value of radiographic studies is limited. Because MRI studies are expensive and require deep sedation, they are not practical. Ultrasound studies are promising and likely to become more widely used with time. Currently, radiographs are still the most practical. Radiographs become increasingly valuable with increasing age [1]. The common measures are as follows:

Tibial calcaneal angle in maximal dorsiflexion is a measure of equinus. To fall into the normal range, the angle should be $>10^\circ$ beyond a right angle.

Lateral talocalcaneal angle is a measure of varus. Parallelism is a sign of residual heel varus.

AP calcaneocuboid alignment provides an assessment of the severity of the midfoot adduction and varus.

Navicular position Dorsal displacement of the navicular is a sign of malalignment of the midtarsal joints.

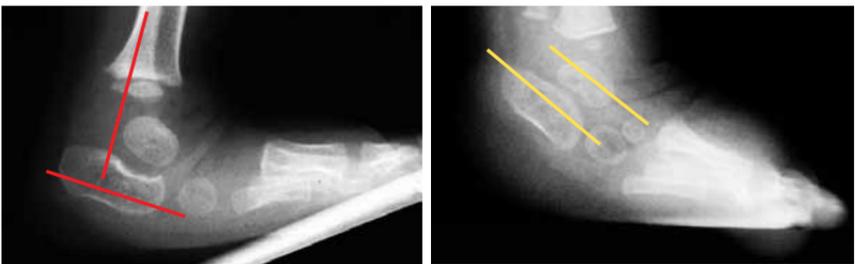
The value of radiographs is uncertain because long-term studies suggest that triceps strength, foot mobility, and plantar loading as measured clinically may be more significant than static radiographic measures in assessing results.

Pirani Score

The Pirani Clubfoot Score document the severity of the deformity and sequential scores are an excellent way of monitoring progress.

Method Six clinical signs seen in all clubfeet that change in severity as the foot deformity changes are each scored 0 (normal), 0.5 (mildly abnormal) or 1 (severely abnormal). They create a simple scoring system [1]. Score each clubfoot every visit and record the finding on a form. Scoring the foot at every visit helps tell the treating practitioner if deformity is correcting satisfactorily and when heel-cord tenotomy is indicated.

Progress assessment when used during Ponseti treatment, the record shows whether the deformity is correcting normally [2] or whether there is a problem and the degree of correction of each component of the clubfoot.



1 Radiographic evaluation of clubfoot. On a maximum dorsiflexion radiograph, measure the tibial calcaneal angle (red lines). On resting or standing radiographs, note the parallelism between the axes of the talus and calcaneus (yellow lines).

Ponseti Clubfoot Management

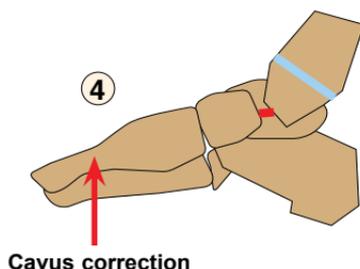
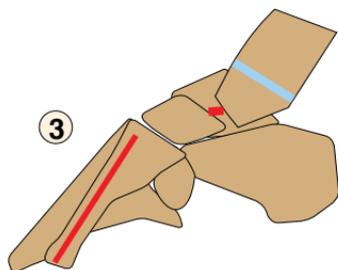
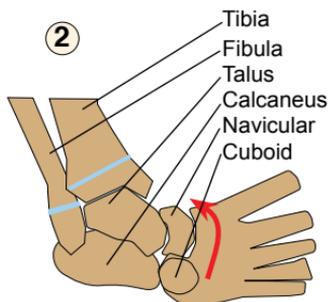
The Ponseti approach to management of clubfeet has been refined over a period of 50 years, has been shown to produce excellent long-term results, and is becoming the management standard throughout the world.

Management

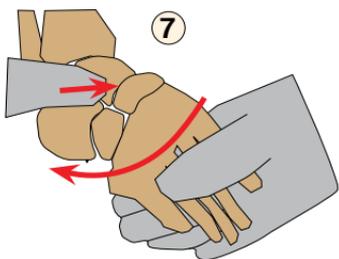
This management shows 90+% success in idiopathic clubfoot, and is also appropriate for other forms of clubfoot. Correction is achieved by manipulation and cast correction in a definite sequence. A percutaneous heel-cord tenotomy corrects equinus, and prolonged night-time bracing is essential to prevent recurrence. The technique for correction involves specific steps.

Understand pathology The idiopathic clubfoot [1] is due to a medial displacement of the mid and forefoot around the head of the talus [2]. In addition, a cavus deformity is present [3] with plantarflexion of the first ray. The initial step is correction of the cavus.

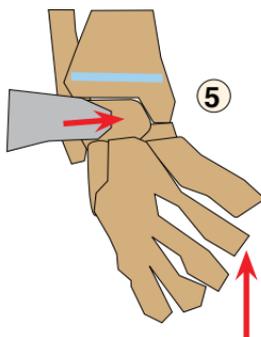
Cavus correction This correction is achieved by dorsiflexion of the first ray. This a brief manipulation [4 and 5 opposite page] followed by casting the forefoot in supination [6 opposite page].



Cavus correction



Adduction and varus correction



6

Adduction and varus correction This correction requires several casts applied 3–7 days apart. With the finger over the head of the talus, the mid and forefoot are abducted and externally rotated [7 opposite page]. The foot is then immobilized in a long-leg cast with maximum correction [1]. Note that the foot requires an extreme position to achieve correction.

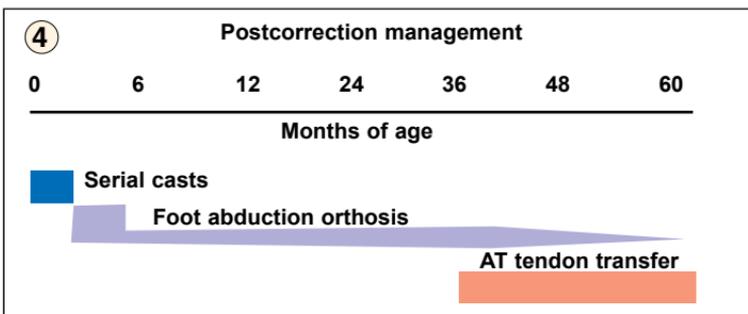
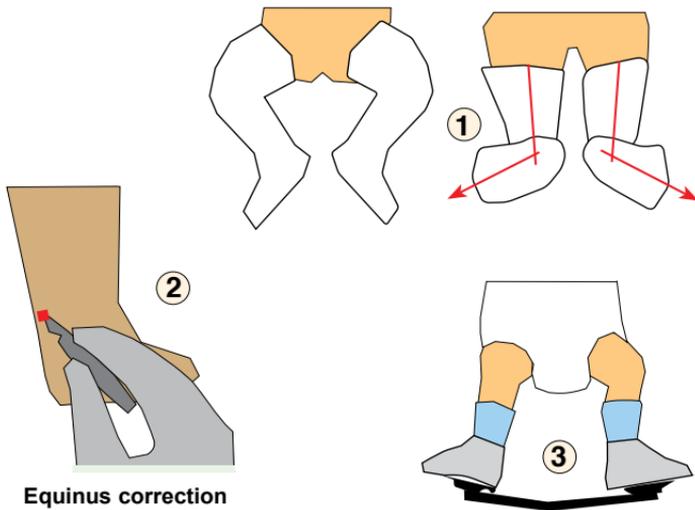
Equinus correction A percutaneous tenotomy [2 is nearly always performed to fully correct the equinus component. A holding cast is applied for about a month following the procedure.

Brace treatment This step is often the most difficult as night bracing [3] is required until the child is age 3–5 years. Without bracing, expect recurrence. This requires skill in dealing with the family.

Anterior tibial transfer Some children require this procedure [4] to correct the muscle balance about the foot. This procedure is performed in early childhood

Bracing

At the end of casting, the foot is abducted to an exaggerated amount, which should measure 60 to 70 degrees (thigh-foot axis). After the tenotomy, the final cast is left in place for 3 weeks. Ponseti’s protocol then calls for a brace to maintain the foot in abduction and dorsiflexion. This is a bar attached to straight last open toe shoes. This degree of foot abduction is required to



maintain the abduction of the calcaneus and forefoot and prevent relapse. The foot will gradually turn back inward, to a point typically of 10 degrees of external rotation. The medial soft tissues remain stretched out only if the brace is used after the casting. In the brace, the knees are left free, so the child can kick them “straight” to stretch the gastrosoleus tendon. The abduction of the feet in the brace, combined with the slight bend (convexity away from the child), causes the feet to dorsiflex. This helps maintain the stretch on the gastrocnemius muscle and Heel-cord tendon

Results

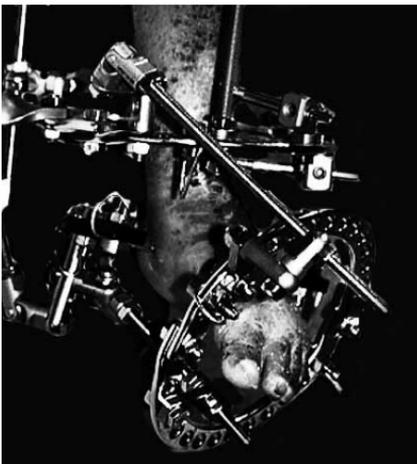
More than 90% of children can expect to have an excellent result. The foot is plantigrade, mobile, and strong. The results are far superior to those achieved by surgery with the traditional posteromedial release procedure. The surgically corrected feet usually become painful during adolescence or early adult life.

Severe clubfeet as seen in conditions such as arthrogyriposis are best managed by initial casting, multiple tendon–ligament releases, and a resumption of casting. Often an open postero–medial–plantar release is required at about 1 year of age. Night splinting in the position of maximum correction is essential to reduce the risk of recurrence. Correct recurrent deformities with casts. Avoid repeated major operative procedures. Plan a final bony correction at the end of growth.

Complications of treatment are common.

- *Recurrence* is the most frequent problem. About one-third of clubfeet require more than one procedure.
- *Stiffness* may result from excessive articular pressure during treatment, compartment syndromes complicating surgery, internal fixation, avascular necrosis of the talus, and operative scarring.
- *Weakness* of the triceps jeopardizes function. Overlengthening and repeated lengthening procedures increase this risk.
- *Varus deformity* commonly causes excessive plantar pressure over the base of the fifth metatarsal.

Severe deformity in older children are sometimes best managed with the Ilizarov frame [1]. To release the deformity, use the frame to stretch soft tissues to achieve gradual correction.



1 Ilizarov frame. This is an effective method of correcting severe deformity in the older child.

Flatfeet

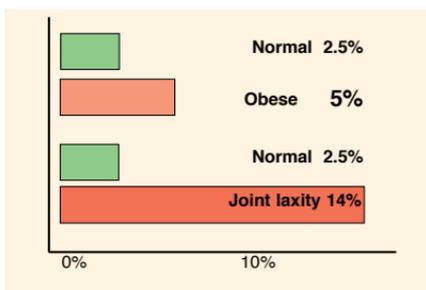
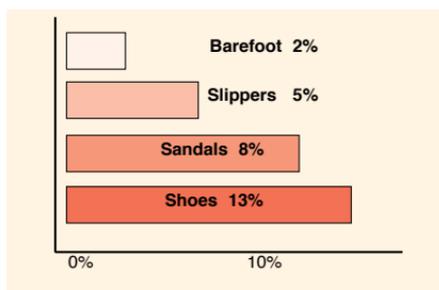
The flatfoot (FF), or pes planus, is a foot with a large plantar contact area. The flatfoot is often associated with a valgus heel and a reduction in height of the longitudinal arch. Flatfeet are classified as physiologic or pathologic. Physiologic flatfeet are flexible, common, benign, and a variation of normal. Pathologic flatfeet show some degree of stiffness, often cause disability, and usually require treatment. Ankle valgus, as seen in myelodysplasia and poliomyelitis, may be confused with a flatfoot deformity. Valgus is in the subtalar joint. Make the differentiation radiographically.

Flexible Flatfoot

The flexible flatfoot or physiologic flatfoot is present in nearly all infants, many children, and about 15% of adults. Flatfeet often run in families [1]. Flatfeet are most common in those who wear shoes, are obese, or have generalized joint laxity [2]. There are two basic forms. Developmental flatfeet occur in infants and children as a normal stage of development [3]. The hypermobile flatfoot persists as a normal variant. Two studies of military populations have shown that the flexible flatfoot does not cause disability and, in fact, is associated with a reduction in stress fractures.



1 Familial flatfeet. Each member of this family had flexible flatfeet. None were symptomatic. Demonstrating the flatfoot in the asymptomatic adult provides reassurance for the parents.



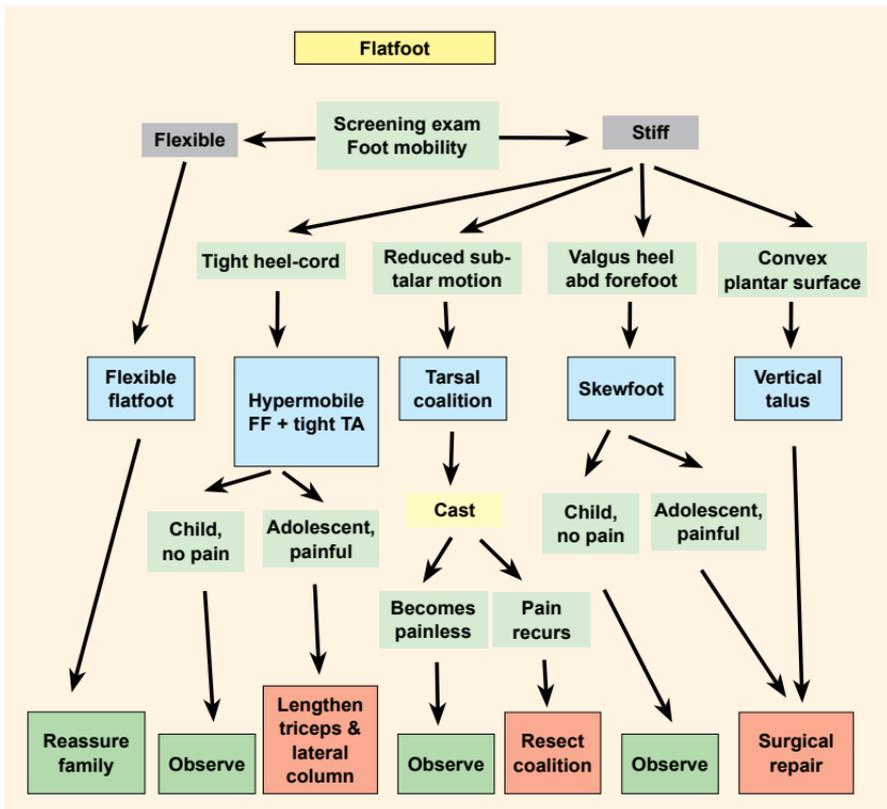
2 Associations of flexible flatfeet. These studies from India demonstrate that flatfeet are more common in adults who wore shoes as children, the obese, and those with joint laxity. From Roe and Joseph (1993).



3 Developmental flatfeet. Most infants and many children have flatfeet. The infant's flatfeet are often due to their thick subcutaneous plantar fatpad and joint laxity.

Evaluation Evaluate to establish a diagnosis [1]. The screening examination may show generalized joint laxity. On standing, the foot appears flat and the heel may show mild valgus. The arch reappears when the child toe stands or the foot is unweighted. Subtalar and ankle motions are full. Radiographs are unnecessary. Flatfeet can be classified into physiologic (flexible) types and pathologic types [1 opposite page].

Management The flexible flatfoot [2 opposite page] requires no treatment, as it has been shown that the condition is not a source of disability. Shoe modifications or inserts [3 and 4 opposite page] are ineffective, expensive, result in a bad experience for the child, and may adversely affect the individual's self-image (see page 208). Operative intervention to create an arch by blocking subtalar motion may establish an arch but may expose the child to risks of an operation [1 on page 202], months of postoperative discomfort, and, possibly because of damage to the subtalar joint, cause degenerative arthritis of the subtalar joint in adult life.



1 Flatfoot management. This algorithm outlines the evaluation and management of flatfeet.

Interventions should not be imposed on the child to "satisfy" the parent. Provide reassurance and make copies of the parent education material to show the grandparents and other family members. If the family insists that something be done, encourage the use of flexible shoes, limitation of excess weight, and a healthy lifestyle for the child.

Calcaneovalgus Deformity

This congenital deformity is due to intrauterine crowding, producing both calcaneus and valgus [2 next page]. The condition may be confused with a vertical talus. Differentiation is made by determining the degree of stiffness. The calcaneovalgus foot is very flexible and the calcaneus lies in dorsiflexion. This condition is associated with developmental hip dysplasia, which should be ruled out by a careful examination of the hips. Because the calcaneovalgus flatfoot is a positional deformity, it resolves spontaneously. Treatment is not required.

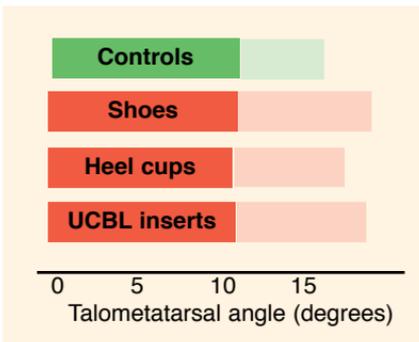
Category	Disorder
Flexible flatfoot	Developmental flatfoot Hypermobile flatfoot Calcaneovalgus foot
Pathologic flatfoot	Hypermobile flatfoot and tight tendo-achilles Lateral tibial torsion, obesity Tarsal coalitions Talocalcaneal Calcaneonavicular Neurogenic flatfoot Severed posterior tib tendon Vertical talus



1 Classification of flatfoot.

Flatfeet are categorized into physiologic and pathologic types.

2 Arch develops on toe standing. This is a characteristic finding in flexible flatfoot.



3 Effect of shoe modifications in flatfeet. This prospective, controlled study compared arch development with various treatments. No difference was found. Talar-metatarsal angles before (light shade) and after (dark shade) treatment. From Wenger et al. (1989).

4 Orthotics can be discarded. This girl was pleased when she was told she could discard the orthotics. Plastic inserts are uncomfortable and make the child feel abnormal.

Hypermobile Flatfoot and Heel-Cord Contracture

Heel-cord contractures cause an obligatory heel valgus, altered tarsal motion, lateral column shortening, and a painful pathologic flatfoot.

Evaluation The patient is usually in the second decade and has vague activity-related foot pain. The foot is flat on standing and the heel-cord contracted. The foot cannot be dorsiflexed beyond neutral with the knee extended [3]. Radiographs often show excessive plantarflexion of the talus. This condition is often confused with simple hypermobile flatfoot and inappropriately called a *symptomatic flexible flatfoot*.

Management Lengthen the contracture of the triceps. If the soleus is contracted, lengthen the heel-cord. If only the gastrocnemius is contracted, perform a recession. Most cases have secondary shortening of the lateral column and require a calcaneal lengthening.



1 Extrusion of silastic implant. This implant was placed in a child with flexible flatfeet.



2 Calcaneovalgus deformity. This is a positional deformity that requires no treatment.



3 Heel-cord contracture with flexible flatfoot. This child with a flexible flatfoot also has a heel-cord contracture. Note that the foot cannot be dorsiflexed above a neutral position with the knee extended (yellow arrow) and that the foot shows plantarflexion of the talus (red arrow).

Tarsal Coalitions

Coalitions are fusions between tarsal bones that cause a loss of inversion and eversion motion. They are often familial, may be unilateral or bilateral, and occur in both sexes equally. The fusion imposes increased stress on adjacent joints and sometimes causes degenerative arthritis, pain, and peroneal spasm. These symptoms usually develop during early adolescence. Sometimes coalitions remain silent. Two common forms are present: Be aware that coalitions may involve more than one joint.

Calcaneonavicular coalitions are most common and sometimes identified on a lateral radiograph [1] but are readily shown by an oblique radiograph of the foot [2]. The coalition may be composed of bone, cartilage, or fibrous tissue. Incomplete coalitions may show only narrowing or irregularity of the calcaneonavicular articulation.

Manage symptomatic coalitions with a trial of immobilization. Apply a short-leg walking cast for 4 weeks. The pain should disappear. If pain recurs soon after removal, operative correction is usually necessary. Resect the coalition (see Chapter 16) and interpose extensor hallucis brevis muscle to prevent recurrence.

Talocalcaneal coalitions (T-C) usually involve the middle facet of the subtalar joint. Conventional radiographs are often normal, but a special calcaneal or "Harris" view may show the fusion. However, the coalition is best demonstrated by CT scans of the foot [3].

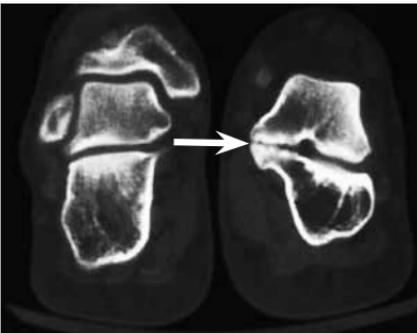


1 "Anteater sign." The calcaneonavicular coalition is seen in lateral radiographs with this characteristic feature (arrows).



2 Calcaneonavicular coalition.

This T-C coalition is readily seen on oblique radiographs of the foot before resection (red arrows). Surgical resection (orange arrow) reduced discomfort and restored motion.



3 Subtalar coalition. The middle facet subtalar coalition (arrow) is readily identified by CT imaging.



4 Failed resection. This resection failed because the coalition was not fully resected.

Manage symptomatic coalitions with a trial using a short-leg cast. If pain recurs, consider operative resection. Assess the size of the coalition by CT imaging. Resection is likely to fail if coalitions exceed 50% of the joint. Technical problems are common [4 previous page]. Heel valgus may be increased by resection. Sometimes a calcaneal lengthening will be needed to correct this component. Outcomes for resection of subtalar coalitions are much less predictable than for the more common calcaneonavicular fusions. Advise the family of the potential for an unsatisfactory result and the possibility that additional procedures may be necessary.

Other coalitions may occur at the talonavicular and naviculocuneiform joint. More extensive coalitions may be present in children with clubfeet, fibular hemimelia, and proximal focal femoral deficiencies. Be aware that pain and stiffness of the subtalar joint may occur with arthritis, tumors, and articular fractures. Consider these uncommon causes of pain if calcaneonavicular and talocalcaneal fusions are ruled out by radiography.

Vertical Talus

The vertical talus is the most severe and serious pathologic flatfoot [1]. It is a congenital deformity that produces not only flattening but an actual convexity of the sole of the foot [2].

Evaluation Vertical talus is usually associated with other conditions such as myelodysplasia and arthrogryposis. The foot is stiff, with contractures of both the dorsiflexors and plantarflexors. The head of the talus projects into the plantar aspect of the foot, producing the convexity of the sole. The diagnosis is suggested by a lateral radiograph of the foot showing the vertical orientation of the talus [3]. Vertical talus may be confused with flexible oblique talus, a different condition. Make the differentiation by studying lateral flexion and extension radio-graphs of the foot. The vertical talus will show stiffness and fixation in contrast to the flexible oblique talus, which shows a freely mobile mid- and hindfoot [1 opposite page]. Note especially the mobility of the calcaneus. If the calcaneus is fixed in plantarflexion in both views of the foot, the diagnosis is vertical talus.



1 Vertical talus in an adult. Note the prominences on the sole of the foot causing calluses and pain with walking.



2 Vertical talus in arthrogryposis. Note the convexity to the sole of the foot.



3 Vertical talus. The talus is positioned in a vertical position (red arrow) as part of the complex deformity of hindfoot equinus and forefoot dorsiflexion.

Management Correct by a single-stage procedure late in the first year of growth. Lengthen the heel-cord and anterior structures and perform a posterolateral release. Elevate the plantarflexed talar head to reestablish the talonavicular joint. Fix with a single smooth longitudinal transcutaneous K wire. Consider transfer of the tibialis anterior to the talus. In the older child, a naviculoectomy or subtalar fusion may be required.

Neuromuscular Flatfeet

Flatfeet are common in cerebral palsy [2] because of spastic contractures of the heel-cord and muscle imbalance. These flatfeet may require operative stabilization because of skin breakdown on the medial aspect of the foot and to provide more stability in walking.

Other Causes

Laceration of the posterior tibialis tendon causes flatfeet in children as it does in adults.

Overcorrected clubfeet is a common cause. This complication may occur in children who have clubfeet and ligamentous laxity.

External tibial torsion and obesity are associated with painful flatfeet in adolescence.

Skewfoot produces heel valgus and a flat-appearing foot, as discussed earlier in this chapter.

Ankle valgus occurs in children with myelodysplasia and clubfeet. Correct by tethering distal tibial growth with a screw [3]. Remove it when deformity is slightly overcorrected.



1 Dorsiflexion-plantarflexion study. This study in an infant shows a dorsiflexion of the calcaneus (red lines) and alignment of the talus and metatarsals (orange line) consistent with a hypermobile foot and inconsistent with a vertical talus.



2 Foot in cerebral palsy. Note the skin breakdown on the medial side of the foot (red arrow). Note the eversion of the foot on the footrests of the wheelchair (yellow arrows).



3 Heel valgus secondary to ankle deformity. Note the valgus ankle (red line) in this 10-year-old child with myelodysplasia. This was managed by placing a tethering medial malleolar screw. Note the improvement early (yellow line) and at 2 years postoperative (orange line). The screw was then removed.

Cavus Feet

A cavus foot is characterized by increased height of the longitudinal arch and is often associated with clawing of the toes and heel varus [1]. Cavus is most often physiological. It is simply the extreme end of the spectrum of normal variability of the shape of the longitudinal arch. This physiologic form is often familial. Pathologic forms of cavus deformity are usually neurogenic.

Physiologic Cavus

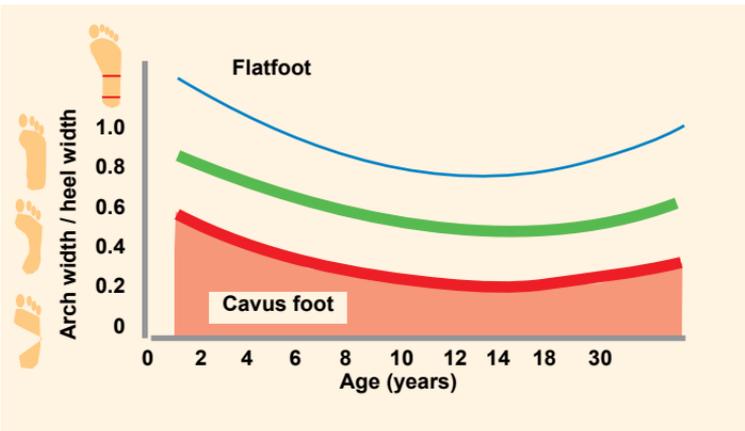
This deformity falls outside the normal range (beyond ± 2 SD from the mean) of variability in arch height [2]. Often a parent’s feet have a high arch. The parent often volunteers that they have a “good” (high) arch. In fact, these parents are more likely to have pain than those with normal or low arches. The cavus is usually bilateral, with an onset in infancy. They may also have calluses under the metatarsal heads. The child’s musculoskeletal and neurologic screening examinations are normal, and clawing of the toes is absent. This is a diagnosis of exclusion. Occasionally the teenager will complain of metatarsal pain. This is best managed by shock-absorbing shoe-ware and, if necessary, a soft shoe insert to unload the metatarsal heads.

Pathologic Cavus

Pathologic cavus is usually secondary to a neuromuscular disorder causing muscle imbalance. A major objective of management is to determine the underlying cause of the deformity.



1 Typical cavus foot. Note the high arch and clawing of the great toe.



2 Normal values for foot contact area. Contact area is described by the ratio of the arch width to the heel width. The mean value (green line), and two standard deviation levels for a low arch (blue line) and a high arch (red line) are shown. A foot is considered to have a cavus deformity if the arch contact area falls outside the ± 2 SD level (red area).

Evaluation The neuromuscular disorders causing cavus deformities are often familial, so the family history is important. Look at the parent's feet. Sometimes they may claim their feet are normal when they are clearly deformed. Perform a careful screening examination of the child. Examine the musculoskeletal system for other problems. Look for midline skin lesions over the spine. A careful neurologic examination is essential. Check muscle strength. Examine the foot, noting the severity of the cavus, degree of rigidity, and presence of clawing of the toes and skin changes over the metatarsal heads. Standing radiographs of the feet are useful in documenting the type and severity of the deformity [2]. Special studies such as spine radiographs for spinal dysraphism, electromyography (EMG), DNA blood tests for Charcot–Marie–Tooth (CMT) disease, nerve conduction velocity measurements, and CPK determination for muscular dystrophy assessment may be necessary. Consultations with a neurologist may be appropriate. Establish the etiology of the cavus deformity [1].

Natural history Because of the reduced area of plantar contact, deformity, and rigidity, cavus feet often cause considerable disability [3].

Category	Type	Etiology
Physiologic Pathologic	Cavovarus	Familial Clubfoot residual cavus Idiopathic Neuromuscular disease Friedreich's ataxia Charcot–Marie–Tooth
	Cavovarus deformity	Spinal dysraphism Spina bifida Poliomyelitis
	Calcaneocavus	Spina bifida Poliomyelitis Overlengthened heel-cord

1 Classification of cavus deformity. This classification includes the majority of causes of cavus feet. Pathologic cavus is often associated with neurologic disorders.



2 Radiographic features of cavovarus deformity. Note the calcaneal pitch (red–white angle) and the first (yellow) and fifth (blue) metatarsal alignment.



3 Skin irritation with cavus deformity. The cavus deformity increases the load on the metatarsal heads. If sensation is poor, as in the child with spina bifida, skin breakdown is common (arrow).

Types of Cavus Deformities

Congenital cavus is a rare deformity that may be due to intrauterine constraint or fixed deformity [1]. Assess the effect of growth.

Calcaneocavus results from weakness of the triceps, an increase in the calcaneal pitch, and a cavus deformity. Correct muscle imbalance if possible. This deformity is seen in poliomyelitis [2], in spina bifida, and following overlengthening of the triceps.

Cavovarus is the most common form. Muscle imbalance results in a mild increased calcaneal pitch and plantarflexion of the forefoot. This deformity is seen in CMT disease. Clawing of the toes is often seen.



1 Congenital cavus. This deformity gradually resolved during the first 2 years of growth. Note the calcaneal pitch (red line) and first metatarsal (yellow line) alignments.



2 Hindfoot cavus deformity. This severe deformity was secondary to weakness of the triceps from poliomyelitis. Note the calcaneal pitch (red line) and first metatarsal (yellow) alignments.



3 Calcaneo-varus deformity. This patient underwent a calcaneal lengthening to reduce deformity of the calcaneus.

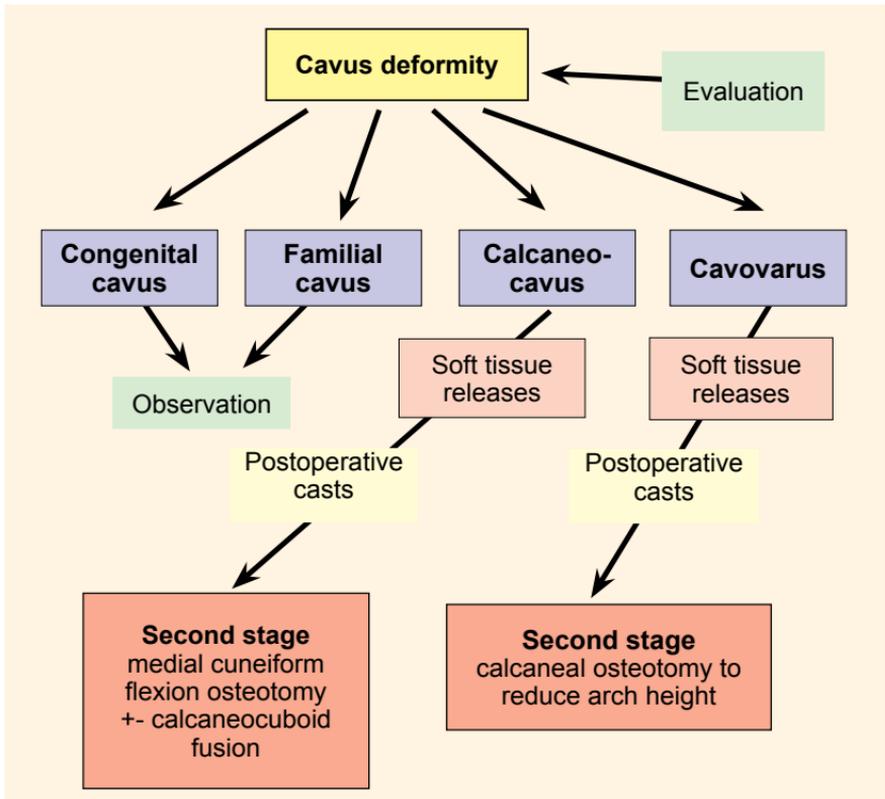
Management

Follow a flowchart to manage [1]. The teenager will often complain of difficulty in fitting shoes, calluses over the claw toes and under the metatarsal heads, and pain.

Mild deformity Order shock-absorbing footwear and soft molded shoe inserts to broaden the load-bearing area of the foot.

Moderate or severe deformity This requires operative correction. Operations improve muscle balance, flatten the arch to broaden the weight-bearing surface, and correct toe deformity.

- *Flexible deformities* or those in young children are best managed by a plantar medial release and appropriate tendon transfers. Plan serial postoperative casting starting at 2 weeks following surgery. Continue until the deformity is resolved. If performed during childhood, be prepared for recurrence.
- *Fixed deformities* require correction in two stages. First, perform a soft tissue release as described above. Follow this by osteotomies to correct bony deformity and tendon transfers to balance the foot. In most cases, perform a calcaneal osteotomy [3 opposite page] for calcaneocavus deformity and a plantar flexion medial cuneiform osteotomy for cavovarus correction. Avoid arthrodesis whenever possible to maintain mobility and reduce the risk of degenerative arthritis of adjacent joints.



1 Cavus management flowchart. From Mosca (2000).

Other Foot Conditions

Toe Walking

Toe walking, or equinus gait, may occur in otherwise normal children [2] or in children with neuromuscular disorders [1].

Neurogenic equinus This deformity is common in children with cerebral palsy, muscular dystrophy, and incompletely treated clubfeet. Correction usually requires lengthening of the heel-cord and sometimes opening of the ankle joint capsule.

1 Toe walking classification. This classification includes the common causes of an equinus gait.

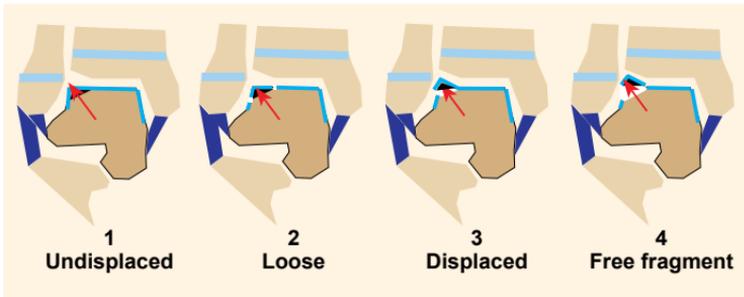
Category	Diagnosis
Congenital	Clubfoot
Idiopathic	Gastroncnemius contracture Accessory soleus muscle Generalized triceps contracture
Neurologic	Cerebral palsy Poliomyelitis
Myogenic	Muscular dystrophy
Functional	Hysterical toe walking



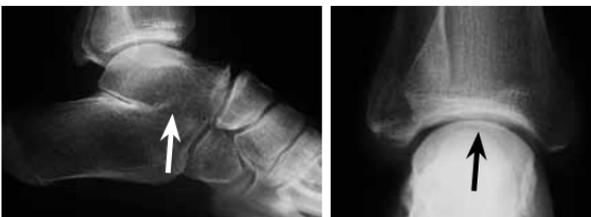
2 Idiopathic toe walking. (Left) This girl has a contracture of the gastrocnemius causing an equinus gait. (Middle and right) This girl's shoes show excessive wear on the toes (arrows).

Idiopathic toe walking Persistent toe walking in infants and young children is uncommon and usually due to shortening of the triceps muscle. Toe walking can be classified into three distinct clinical categories.

- *Gastrocnemius contracture* is the most common form. This isolated deformity is often familial and varies in severity. Ankle dorsiflexion is limited with the knee extended. The sole shows wear under the toes. Physical therapy and stretching exercises are ineffective, and although casting will correct the contracture, recurrence is common. Operative lengthening of the gastrocnemius aponeurosis is effective.
- *Accessory soleus* is a rare congenital deformity in which the body of the soleus muscle extends to the ankle. This produces equinus and a fullness on the medial side of the ankle. Operative lengthening may be necessary.
- *General triceps contracture* is a rare condition that may result in fibrosis and shortening of the triceps muscle group. Ankle dorsiflexion is limited regardless of knee position. Operative correction by heel-cord lengthening may be necessary.



1 Osteochondritis dissecans of the talus. This Berndt and Hartly classification is based on Giguera et al. (1998). Most can be diagnosed with conventional radiographs (red arrows). The degree of displacement can be demonstrated by CT studies.



2 Ball and socket ankle. Note the subtalar fusion (white arrow) and the spherical shape of the ankle (black arrow).

Osteochondritis Dissecans of the Talus

These lesions are regions of avascular bone that occur most commonly on the anterolateral and posteromedial aspects of the talar dome. The lateral lesions are more likely to be associated with an ankle sprain and to be traumatic in origin.

The causes of these lesions may vary. Some follow trauma, while most occur spontaneously and may represent variations in ossification or idiopathic avascular necrosis. They are most common during adolescence in both genders. Most lesions are medial in location.

Evaluation Order AP, lateral, and mortise radiographs. CT and MRI may be helpful to assess the extent of the lesions and cartilage status if operative management is being considered.

Classification As with other osteochondritis lesions, these are classified into four categories [1 previous page].

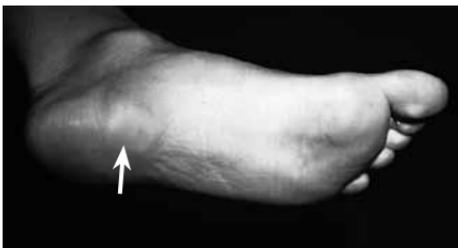
Management This is based on the type of lesion.

- *Type 1 and 2 lesions* Manage in a short-leg cast for 4–6 weeks.
- *Type 3 lesions* Manage by reduction and immobilization of the fragment with bioabsorbable pegs. Approach by arthroscopy or with the aid of a transmalleolar osteotomy.
- *Type 4 lesions* Excise if small or old. Replace and fix if feasible.

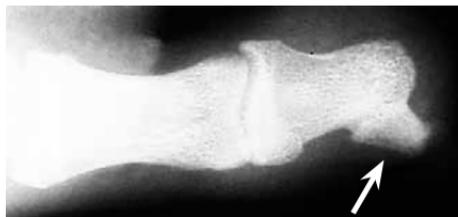
Prognosis Good to excellent in the 90% range.



1 Calcaneal prominences. This teenage girl has bilateral prominences with some discomfort.



2 Plantar fibromatosis.



3 Subungual exostosis

Ball-and-Socket Ankle

Ball-and-socket ankle is a rare deformity associated with conditions such as extensive tarsal coalitions [2 on page 211], congenital shortening of the lower limb, absent digital rays, and aplasia or hypoplasia of the fibula. The deformity causes little or no disability, and no treatment is required.

Calcaneal Prominence

This deformity is seen in adolescence, is often bilateral, and is thought to be related to irritation from footwear [1 opposite page]. Manage most cases with thoughtful shoe selection. Rarely, exostectomy is required. Operative results are often poor.

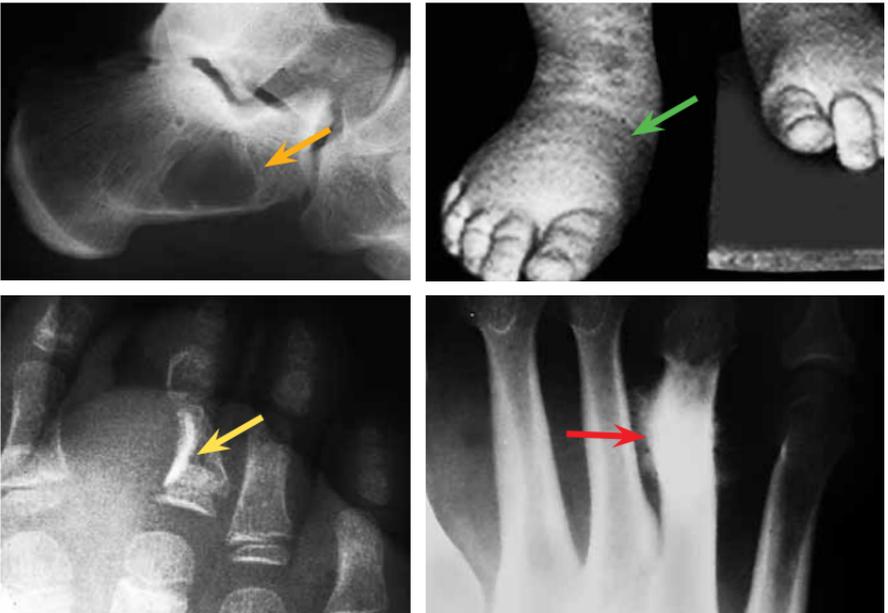
Tumors of the Foot

Accessory soleus is a rare variation in the soleus causing a swelling just medial to the achilles tendon. The mass is smooth, round, and nontender and grows proportionately with the foot. No treatment is required.

Plantar fibromatosis is a rare tumor with a characteristic location on the anteromedial portion of the heel pad [2 opposite page]. As recurrence following resection is common and many resolve spontaneously, observational management is usually indicated.

Subungual exostosis is a benign bone tumor of the distal phalanx occurring beneath or adjacent to the nail [3 opposite page]. This rare tumor occurs in late childhood or adolescence and most commonly affects the great toe. Its characteristic location and radiographic appearance establish the diagnosis. Manage by careful and complete excision to avoid recurrence.

Other tumors Many other benign and malignant tumors occur in the foot and usually have no unusual features that are unique to the foot [1].



1 Tumors of the foot. Numerous tumors occur about the foot. Some are benign, such as cysts of the calcaneus (orange arrow) and vascular tumors (green arrow). Less common are malignant tumors, such as desmoid tumors (yellow arrow) and osteogenic sarcoma (red arrow).

Chapter 6 – Knee and Tibia

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In this chapter, disorders of the knee and tibia are addressed. In all age groups, knee problems account for over one-fourth of musculoskeletal complaints. In children, knee complaints are substantially less common but increase in frequency during the teen years. Osteomyelitis and osteogenic sarcoma develop more often about the knee than at any other site due to the rapid growth rate of the distal femoral and upper tibial physes.

Introduction

Nomenclature

The fully extended knee is the neutral or zero position. The normal range of motion extends from neutral to about 140° , with most activities performed in the 0° – 65° segment of the flexion arc. In the child, hyper-extension of up to 10° – 15° is normal [1]. The difference between active and passive motion is termed *lag*.

Hyperextension, if associated with stiffness, is called a *recurvatum deformity*. Restricted motion is described by specifying the arc of motion. For example, a stiff knee may be described as having an "arc of motion from



1 Hyperextension deformities of the knee. The boy's knees hyperextend due to generalized joint laxity. The girl's have a hyperextension deformity due to a knee injury with loss of knee flexion.

20°–55°." The arc of motion in hyperextension is preceded by a minus sign. A child with a hyperextension deformity may have a range from –20° to 30°, giving a 50° arc of motion.

The **knee angle** is the thigh–leg angle or femoral–tibial angle (see Chapter 4). Changes in the knee angle that represent normal variations are physiological and cause bowlegs or knock-knees. Deformities, those falling outside the normal range (± 2 SD) and those due to pathological processes, are termed *genu varum* or *genu valgum*.

Normal Development

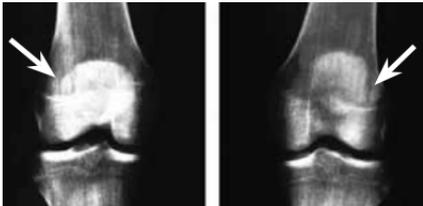
The knee develops as a typical synovial joint during the third to fourth fetal months. The secondary centers of ossification for the distal femur form between the sixth and ninth fetal months and for the upper tibia between the eighth fetal and first postnatal months. The patella ossification center appears between the second and fourth years in girls and the third and fifth years in boys.

Developmental Variations

Variations of ossification or development may cause confusion in assessing radiographs.

Bipartite patella is due to an accessory ossification center of the patella that usually occurs in the superior–lateral corner [1].

Fibrocartilagenous defects are usually insignificant developmental variations that are most common about the knee. They are eccentric and show sclerotic margins and radiolucent centers. They resolve spontaneously [2].



1 Bilateral bipartite patellae. Secondary centers may appear on both knees. They are usually asymptomatic.



2 Fibrocartilagenous defect. Typical large lesion in distal femur (red arrow). Two years later, spontaneous healing has occurred (yellow arrow).

Clinical Feature	Disorder
Patellar hypoplasia	Nail-patella syndrome Beals syndrome Diastrophic dysplasia Neurofibromatosis
Genu varum	Rickets Achondroplasia TAR syndrome Metaphyseal dysplasia
Genu valgum	Rickets Morquio syndrome Poliomyelitis Ellis-Van Creveld syn.
Genu recurvatum	Myelodysplasia Arthrogryposis Larsen syndrome
Flexion contracture	Arthrogryposis Pterygium syndrome Myelodysplasia
Patellofemoral disorders	Nail-patella syndrome Rubinstein-Taybi syn.

3 Syndromes associated with knee deformity. These are examples that illustrate the relationship of knee deformities in various generalized disorders.

Evaluation

Evaluating the child's knee is different from that of the adult because disorders are more likely to be due to some underlying generalized dysplasia or to focal congenital or developmental deformity.

Screening Examination

Screen for some underlying abnormality [3, opposite page], such as nail-patella syndrome [1]. Dislocation of the patella is common in Down syndrome. Dimpling over the knee is common in arthrogryposis. Recurvatum occurs in spina bifida and in arthrogryposis. Genu varum and valgum are common in ricketic disorders, and genu valgum is common in Morquio's and Ellis-Van Creveld syndromes.

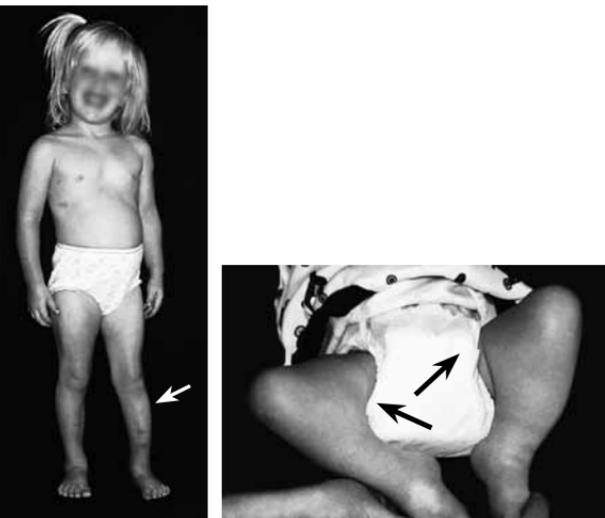
Physical Examination

The physical examination usually provides the diagnosis or at least the basis for ordering further studies.

General inspection Look for obvious deformity, check the knee angle, and perform a rotational profile [2].



1 Nail-patella syndrome. Note the nail dysplasia and absence of the patella.



2 Inspection. The screening examination of this standing child shows the shortening and bowing of the left tibia (white arrow) and the café-au-lait spots of neurofibromatosis. Severe flexion contractures are present in the popliteal pterygium syndrome (red arrows).

Knee Observe the child standing and note symmetry, knee angle, position of patella, masses, joint effusion, muscle definition and atrophy [1], and signs of inflammation. Is there full extension or hyper-extension?

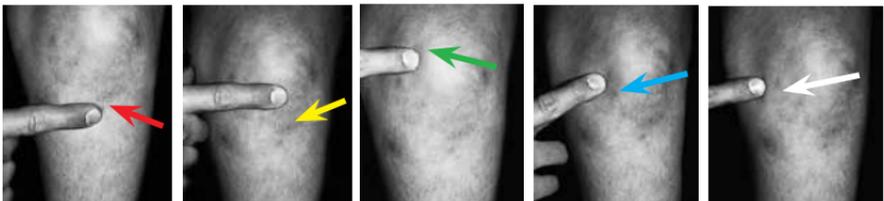
- **Patellar tracking** Ask the child to sit and slowly flex and extend the knee. Observe the tracking of the patella. Does it move in a linear fashion or displace laterally as the knee extends [1, opposite page]? Does the knee fully flex and extend?
- **Q angle** is the angle formed by a line connecting the anterior superior iliac spine with the midportion of the patella and a second line from the patellar mid-point to the tibial tubercle. Normally the enclosed angle is less than about 15°. A larger angle may or may not be associated with patellofemoral instability.
- **Point of maximum tenderness** Locate the PMT by systematically examining the entire knee and tibia. The PMT often establishes a working diagnosis [2].



1 Hypoplasia of the quadriceps. Hypoplasia is a common dysplastic feature in patellofemoral disorders in children and adolescents. Note the lack of definition of the VMO (arrows).



3 Knee tests. Compressing the suprapatellar bursa (red arrows) displaces any joint fluid into the joint to demonstrate an effusion. Displacing the patella laterally (yellow arrows) may elicit the patellar apprehension sign.



Arrow	PMT	Condition
Red	Tibial tubercle	Osgood–Schlatter disease
Yellow	Distal pole patella	Sinding–Larsen–Johansson syn.
Green	Medial patellar margin	Patellar instability
Blue	Medial joint line	Meniscal lesion
White	Medial collateral ligament	Ligament injury

2 Point of maximum tenderness. Evaluate the PMT for common painful conditions about the knee.

Palpate to assess temperature, swelling, and tenderness. Is the affected knee warmer than the other knee? Is a joint effusion present [3, opposite page]? Parapatellar fullness suggests a joint effusion. Evaluate any fullness by extending the knee, compressing the suprapatellar region, and checking for a fluid wave in the knee. A posttraumatic effusion is a sign of a significant intraarticular injury such as a torn peripheral meniscus, anterior cruciate ligament (ACL) injury, or osteochondral fracture.

Manipulate to determine if the patella is displaceable. In loose-jointed children, the patella is very mobile and more likely to dislocate.

- **Patellar apprehension** is elicited by extending the knee and attempting to displace the patella laterally. Patients with recurrent dislocations who sense that this may cause the patella to dislocate may become apprehensive and may reach out to stop the examination.
- **Knee motion** Is the arch of motion free and unguarded? Is crepitation or snapping present?
- **Knee tests** Anteroposterior laxity can be assessed with the Lachman test. Flex the knee about 15° – 20° and attempt to displace the tibia anterior in its relationship to the femur. Normally a firm endpoint will be felt. Check for instability with varus and valgus stress [2]. With the knee flexed to a right angle, evaluate for anterior or posterior drawer signs.



1 Patellar tracking. As the child slowly extends the knee, the patella normally tracks vertically. Lateral patellar displacement as the knee becomes fully extended (arrow) is described as “J” tracking.



2 Knee stability. Test for medial lateral instability with the knee flexed to 30° . Test the opposite or normal knee to determine what is normal for the child.

Imaging Studies

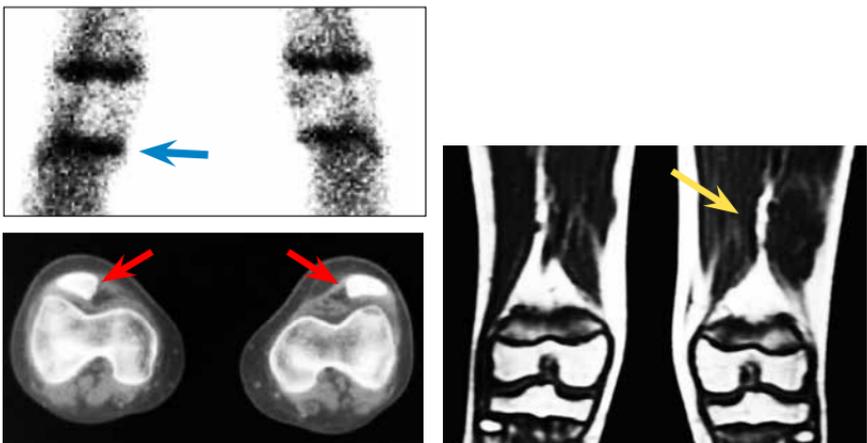
Special radiographic projects such as sunrise and notch views [1] may be useful. If conventional radiographs are not adequate, order special imaging studies [2]. Bone scans may be helpful in determining the location or activity of lesions, and MRI studies are most useful for tumors or meniscal abnormalities. Be aware that because of the sensitivity of these tests, over-reading is common.

Arthroscopy

Arthroscopy is essential for assessing meniscal injuries and for other ligamentous and osteochondral problems in children. It is less valuable for assessing pain.



1 Notch view. The condylar fracture is seen only in the notch view (red arrow).



2 Special imaging studies. These special studies demonstrate lateral patellar positioning (red arrows), a hemangioma by MRI (yellow arrow), and increased uptake of the proximal medial tibial physis in tibia vara (blue arrow).

Knee Pain

Knee pain is a common presenting complaint [1].

Referred Pain

First consider the possibility of referred pain from slipped capital femoral epiphysis [2] or pain from a tumor. These conditions require urgent treatment.

Osgood–Schlatter Disease

Osgood–Schlatter disease (OSD) is a traction apophysitis of the tibial tubercle due to repetitive tensile microtrauma. It occurs between ages 10 and 15 years, with the onset in girls about 2 years before that in boys. OSD is usually unilateral and occurs in 10–20% of children participating in sports. OSD is associated with patella alta. Whether this association is a cause or an effect of OSD is not known. The tibial tubercle maybe enlarged on the asymptomatic side.

Physical examination will demonstrate swelling and localized tenderness over the tibial tubercle [3] and no other abnormalities. Order a radiograph if the condition is unilateral or atypical. Radio-graphs usually show soft tissue swelling and sometimes a separate ossicle over the tubercle.

Natural history OSD resolves with time in most children [1, next page]. In about 10% of knees, some residual prominence of the tibial tubercle or persisting pain from an ossicle may cause problems.

Category	Disorder
Referred	SCFE, other hip problems Tumor
Stress	Osgood–Schlatter disease Sinding–Larsen–Johansson Stress fractures Proximal tibia Patella Distal femur Medial collateral ligament
Bursitis	Prepatellar Pes anserina
Intraarticular	Meniscus Ligaments Osteochondritis dissecans
Tumors	Popliteal cyst Miscellaneous
Arthritis	Septic Pauciarticular Juvenile rheumatoid arthritis Rheumatoid spondylitis

1 Classification of knee pain. Knee pain has many causes. Some examples are listed.



2 Pitfalls in evaluating knee pain. Referred pain can occur from this slipped capital femoral epiphysis (arrow).

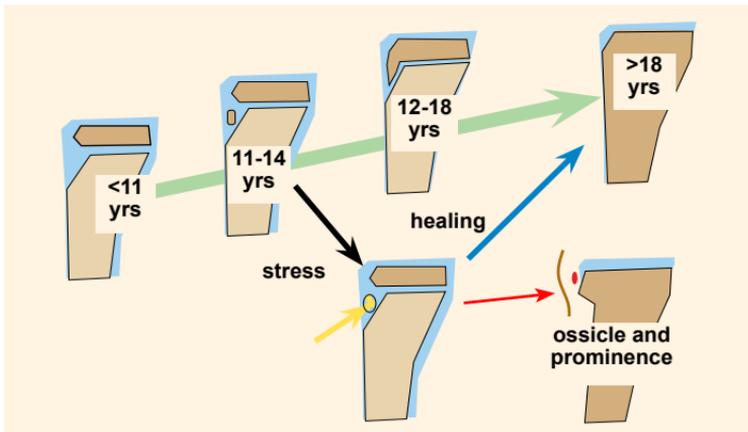


3 Osgood-Schlatter disease. Note the prominence and the ossification over the tibial tubercle (red arrows). Persisting tenderness over an ossicle in the mature knee (yellow arrows) is an indication for excision.

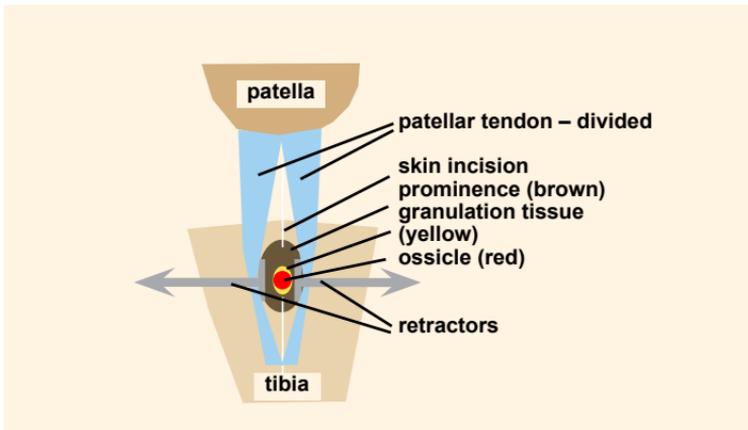
222 Knee and Tibia / Knee Pain

Manage by ordering modifications of activities, use of NSAIDs, and a knee pad to control discomfort. If OSD is severe or persists, apply a knee immobilizer for a week or two to relieve inflammation. Injection of steroids is not recommended. Often quadriceps and hamstring flexibility exercises are prescribed. To reduce apprehension, consider referring to OSD as a *disorder* or *condition* rather than a *disease* when discussing the problems with the patient and family. Make certain that they are aware that resolution is usually slow, often requiring 12–18 months. Persisting disability from tenderness and tubercle prominence may be sufficient to require excision of the ossicle and prominence [2].

Complications are rare and include growth arrest with recurvatum deformity and rupture of patellar tendon or avulsion of the tibial tubercle.



1 Natural history of OSD. The normal development of the apo-physis is shown by the green arrow. Excessive traction (yellow) of the patellar tendon causes inflammation. Usually this process heals. In some cases, inflammation and a separate ossicle persist (red). Based on Flowers and Bhadrshwar (1995).



2 Excision of the ossicle and prominence. The procedure is performed through a midline incision to the ossicle and prominence. Sometimes inflammatory granulation tissue is present in active lesions.

Sinding–Larsen–Johansson Syndrome

Sinding–Larsen–Johansson syndrome is a traction apophysitis of the distal pole of the patella [1]. The condition is most common in males at or before puberty. Resolution occurs in 6–12 months. Rest the knee to resolve the pain and tenderness. Quadriceps flexibility exercises are commonly prescribed. No residual disability has been reported.

Pes Anserina Bursitis

Inflammation of the pes anserina bursa causes pain and tenderness over the hamstring tendon insertions on the posterior medial aspect of the upper tibial metaphysis. This uncommon condition occurs during the teen years. Manage the bursitis with rest and nonsteroidal antiinflammatory medications.

Medial Collateral Ligament Pain

Medial collateral ligament pain is an overuse condition causing pain and tenderness over the medial collateral ligament. This ligament lies on the posteromedial aspect of the knee at or above the joint line.

Bipartite Patella

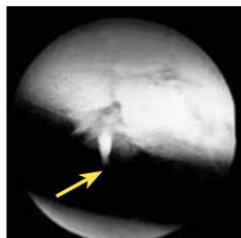
Accessory centers of ossification of the patella [2] may produce a bipartite patella. The separate ossicle is attached to the body of the patella by fibrous or cartilaginous tissue. Trauma may disrupt this attachment, and the ossicle then becomes painful. The disruption may heal with rest. In others, healing fails to occur and the ossicle remains chronically painful. Small painful ossicles may be removed. Larger ossicles should be fixed with a screw to the patella and grafted to promote union.



1 Sinding–Larsen–Johansson syndrome. Note the separate lesion of the distal pole of the patella. This should be differentiated from the uncommon type of bipartite patella involving the inferior pole of the patella.



2 Bipartite patella. Note the separate ossicle on the superolateral aspect of the patella. The lesion was painful.



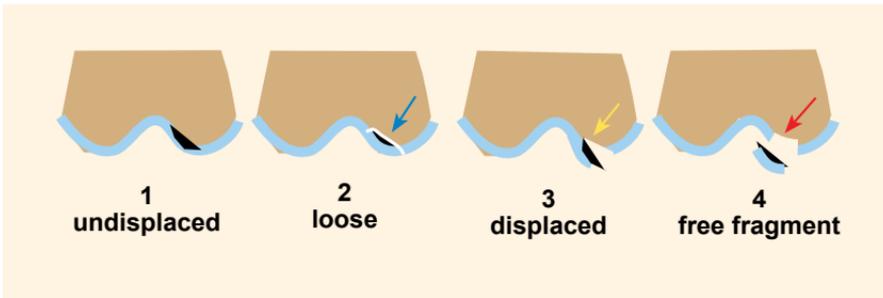
3 Osteochondritis dissecans. MRI demonstrates a large defect (red arrow) with intact cartilage. Arthroscopy shows a lesion with overlying irregular cartilage (yellow arrow).

Osteochondritis Dissecans

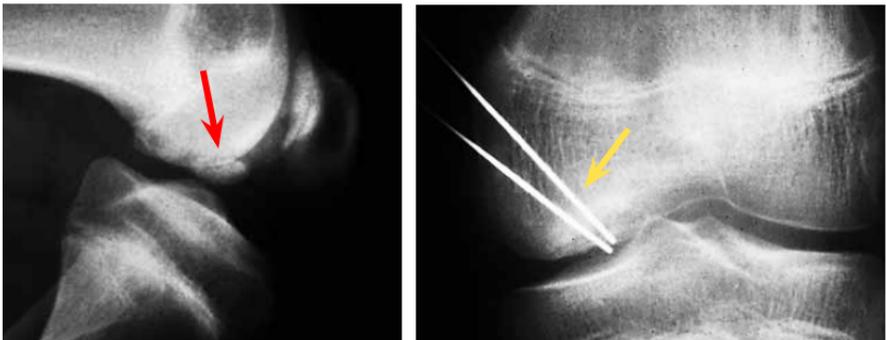
Osteochondritis dissecans may involve the medial or lateral condyle or patella [3, previous page]. The lateral side of the medial condyle is involved in 75% of cases. Symptoms include pain, a mild effusion, or later mechanical symptoms. Image by a notch view. Classify based on degree of displacement [1]. A good prognosis is related to a younger age and to a small lesion in a nonweight-bearing location without displacement.

Manage type 1 and 2 lesions with activity modification, isometric exercises, and a knee immobilizer. Manage based on symptoms rather than on radiographic appearance. Radiographic healing takes many months. Manage type 3 lesions by drilling and stabilizing with K wires [2] or absorbable pins. Manage type 4 lesions if small by excision. Replace large lesions or those involving the weight-bearing areas and fix internally if adequate subchondral bone exists on the fragment.

Prognosis For large lesions involving the weight-bearing surfaces, osteoarthritis is common in adult life.



1 Classification of osteochondritis dissecans. The lesion (black) may be undisplaced or loosely in place (blue arrow). Displacement may be partial (yellow arrow) or complete (red arrow), allowing the fragment to be free.



2 Treatment of osteochondritis dissecans. The lesion (red arrow) was treated with drilling and fixation by diverging pins (yellow arrow). Cannulated screw fixation can also be used.

Intraarticular Disorders

Meniscal and ligamentous lesions are relatively rare in children but become more common during adolescence.

Discoid Meniscus

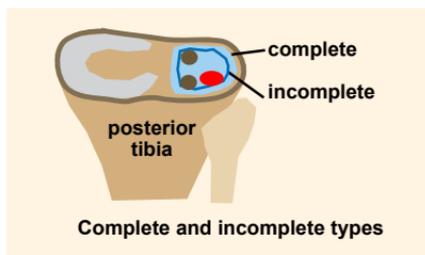
Three types of discoid meniscus occur.

Complete and incomplete types are thicker than normal and cover all or part of the tibial surface [1].

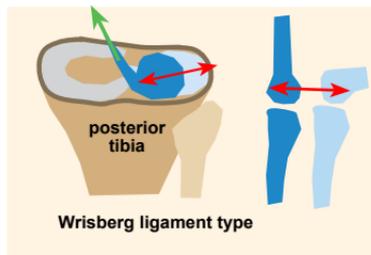
Wrisberg ligament type is attached to the meniscomfemoral ligament posteriorly [2]. This meniscus has no other fixation and is mobile. It is most likely to cause snapping and symptoms in the younger child. Because of its mobility, it may be caught between the femoral condyles and become torn or eroded.

Diagnosis Symptoms include pain, snapping or locking, loss of knee extension, and giving way. Tenderness and fullness may be present over the lateral joint line, and crepitation may be present with motion. In the young child, snapping may be the only complaint. Radiographs may show widening of the joint space [3]. MRI studies are usually diagnostic and usually allow differentiation of the type of lesion. Arthroscopic confirmation should be delayed until operative treatment is considered necessary.

Management depends on the type, symptoms, and activity level of the child. Be conservative. Menisectomy is indicated for the meniscus with posterior attachments. Attempt to preserve the meniscus whenever possible. Total menisectomy is the last resort. Long-term outcomes are poor due to premature osteoarthritis.



1 Complete and incomplete types of discoid meniscus. These menisci are fixed to the tibia both in front and back of the knee (brown spots). The most vulnerable site for injury is posterior (red spot). Based on Dickhaut and DeLee (1982).



2 Wrisberg ligament type of discoid meniscus. The ligament is attached to the Wrisberg ligament (green arrow). This ligament (meniscomfemoral ligament) attaches to the femur. With knee flexion and extension, the meniscus moves (red arrow) because it has no fixation to the tibia.



3 Discoid lateral meniscus. Note the widening of the lateral joint space (red arrows) and the bow-tie-shaped discoid meniscus on MRI (yellow arrows).

Tears of Medial Meniscus

These lesions become more common during the teen years. The findings are similar to tears seen in adults [1]. Tears are usually longitudinal in type. Preserve the meniscus by repairing reduceable outer third lesions. Perform a partial meniscectomy for inner third or comminuted lesions. Avoid total meniscectomy to preserve the stress shielding function of the meniscus. Meniscectomy in childhood leads to premature osteoarthritis [2].

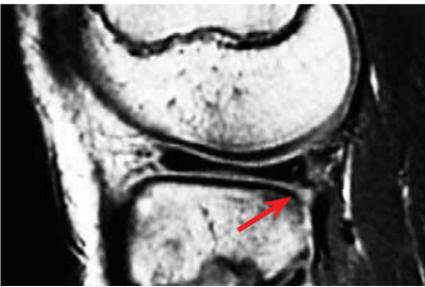
Cruciate Ligament Deficiencies

Cruciate ligament insufficiency is seen in a variety of conditions in children.

Congenital deficiencies are common in fibular hemimelia and proximal focal femoral deficiencies. These deficiencies complicate limb lengthening procedures in these children. Isolated absence of both anterior and posterior cruciate ligaments have been reported.

Acquired deficiencies occur from traumatic rupture of the ligament [3], attenuation associated with tibial spine fractures, and sometimes in association with diaphyseal femoral shaft fractures without known knee injury.

Management ACL tears are repaired in traumatic ruptures that cause disability, especially when associated with meniscal injuries.



1 Meniscal tear. Note the meniscal tear on this MRI (arrow).



2 Long-term effect of meniscectomy. At age 13 years, this 29-year-old man had a total meniscectomy of the right knee. Note the degenerative changes compared to the unoperated knee.



3 Traumatic ACL tear in adolescent. This 15-year-old boy has a tear in his ACL as shown on the MRI.

Tumors of the Knee

Meniscal Cysts

Meniscal cysts are uncommon lesions that usually occur over the lateral aspect of the knee and may be associated with a meniscal tear. Image with ultrasound or by MRI [1]. Manage by arthroscopic excision or repair.

Popliteal Cysts

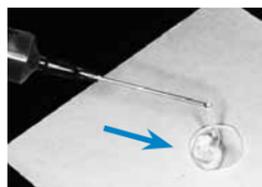
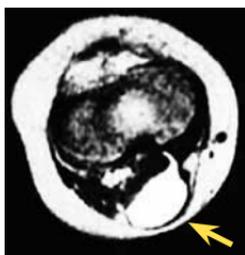
These cysts are different in children than in adults. The cysts seldom communicate with the joint and are not related to intraarticular defects. The natural history is of spontaneous resolution.

Diagnosis is usually not difficult [2]. Most cysts are found by the parents through observation. The cysts are usually nontender, smooth, cystic to the touch, and located between the medial head of the gastrocnemius and the semitendinosus. Transillumination demonstrates that the mass is a cyst. Ultrasound shows the lesion well, so that MRI is seldom necessary.

Management Reassure the family that the condition is benign and will



1 Lateral meniscal cyst. Note the cyst upon physical examination of the knee and on the MRI.



2 Popliteal cyst. Note the classic location (red arrow) and MRI appearance (yellow arrows) of the cyst. Aspiration treatment (green arrow) removes viscous fluid (blue arrow) from within the cyst.

resolve with time. If the family is still nervous, consider confirming the diagnosis by aspirating the cyst. Advise the family that the aspiration is only to confirm the diagnosis and not for treatment because the cyst will recur. Aspiration reassures the family that it is not cancer. Cysts resolve spontaneously over a period of several years. Resection is indicated only with large painful cysts. Recurrence following resection is common.

Synovial Disorders

Intraarticular hemangiomata These lesions infiltrate and thicken the synovium, making it subject to injury and bleeding [1]. The diagnosis can be made by aspirating blood from the joint and confirmed by biopsy done concurrently with a synovectomy. Warn the family that recurrence during growth is likely.

Juvenile rheumatoid arthritis causes chronic synovitis with stiffness, overgrowth of the tibia and femur, contractures, and eventually joint destruction. Steroid injections, arthroscopic synovectomies, and contracture releases supplement medical management.



1 Intraarticular hemangioma. Note the swelling of the knee. This boy had repeated synovectomies for recurrent hemangiomata of the knee.

Condition	Effect
Femoral and tibial torsion	Increases Q angle Overall limb malalignment
Genu valgum	Increases Q angle
Condylar hypoplasia	Promotes lateral subluxation or dislocation
Patella alta	Results in less lateral stability
Quad insufficiency	Produces imbalance in quadriceps
Med. cap. attenuation	Inadequate medial check-rein
VL contracture	Tethers patella laterally

2 Factors contributing to patellofemoral instability. These factors combine to increase the risk of patellar subluxation or dislocation.

Patellofemoral Disorders

Many factors may contribute to patellofemoral instability [2, opposite page].

Systemic Disorders

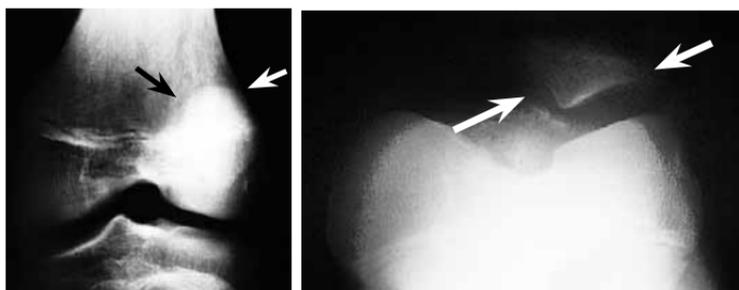
Patellofemoral instability is more common in children with (1) knee dysplasias, such as occurs in nail–patella syndrome [1], Rubinstein–Taybi syndrome, and Turner syndrome; and (2) conditions with increased joint laxity such as Down syndrome. These underlying conditions complicate management.

Congenital Dislocation

Congenital patellar dislocation is a rare condition that causes a progressive flexion, valgus, and tibial external rotational deformities of the knee. Reduce the dislocation and realign the quadriceps mechanism late in the first year. An extensive lateral release is often required.

Patellar Subluxation or Dislocation in Childhood

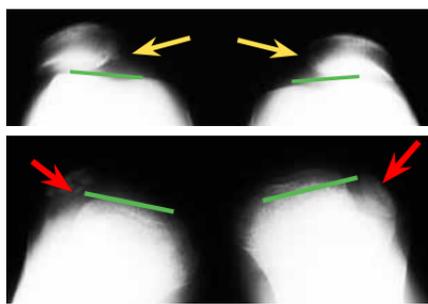
So-called *habitual* subluxation or dislocation is usually due to a dysplastic knee with contracture of the lateral portion of the quadriceps mechanism. This causes the patella to displace laterally whenever the knee is flexed. Early operative realignment is appropriate, but because the dysplastic features are severe, recurrence is common.



1 Patellar hypoplasia. This deformity was part of the nail–patella syndrome.



2 Quadriceps hypoplasia. Note the loss of delineation of the VMO. The hypoplasia and weakness contribute to patellar instability.



3 Sunrise views. These projections show subluxation (yellow arrows) and dislocations (red arrows) of the patella. Note the position of the anterior femoral articular surface (green lines).

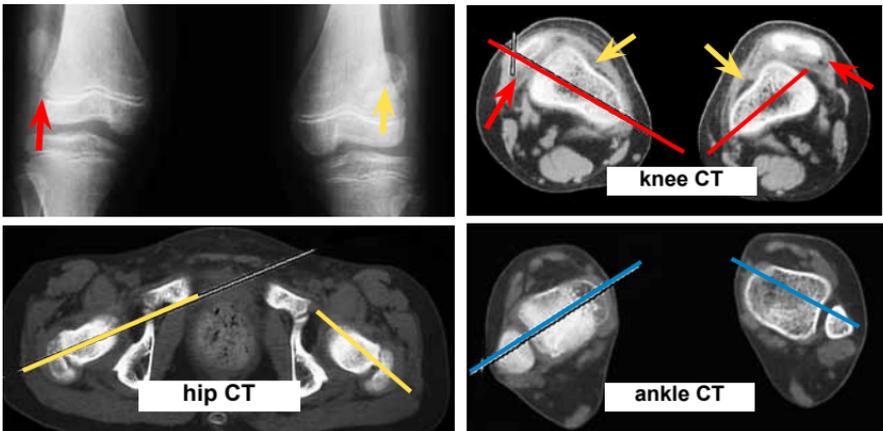
Traumatic Patellar Subluxation or Dislocation

Traumatic patellar dislocations cause an articular fracture. If the injury is severe, producing a tense hemarthrosis, arthroscopic evaluation may be appropriate.

Adolescent Recurrent Dislocation

Most recurrent dislocations occur in individuals with dysplastic knees. They may show generalized joint laxity, lateral tibial torsion, genu valgum, hypoplasia of the quadriceps [2, previous page], attenuation of the medial joint capsule, limited medial mobility of the patella, and abnormal patellar tracking. Observe the tracking of the patella as the patient slowly extends the knee. Lateral displacement of the patella as the knee nears full extension is described as J tracking. J tracking is a common finding. Sometimes the patella becomes subluxated with a sudden lateral shift. The patellar apprehension sign may also be positive. The patient becomes fearful that the patella will dislocate when the examiner applies lateral pressure to the patella. Radiographs may show lateral displacement of the patella [3, previous page].

Management Manage first by ordering isometric quadriceps exercises. If the instability persists, operative correction is often necessary. Identify and correct each dysplastic component. In the growing child, often a lateral release, a medial plication, and a transfer of the semitendinosus to the patella are required. After growth is completed, reposition the tibial tubercle more medial and anterior to align and to optimize quadriceps alignment.



1 Rotational malalignment syndrome. This child had habitual dislocations at age 5 years. Realignment was performed on the worse left side. She was not seen again until age 10 years. At this time, she is asymptomatic. Her left patella is subluxated and right patella dislocated. The child has severe rotational malalignment as demonstrated by CT scans. Note that the bicondylar axis is medially rotated 30° (yellow lines). This results in 60° of anteversion (red lines) and about 75° of lateral tibial torsion (blue lines). Note the displaced patella (red arrows) and the shallow condylar grooves (yellow arrows). It was elected not to attempt operative repair because rotational osteotomies of both femora and tibiae and realignment would be necessary. The chance of success was considered too poor to justify the magnitude of the procedures. This case demonstrates the complex pathology of some types of congenital development patellofemoral disorders.

Anterior Knee Pain

Anterior knee pain is common during the teen years. It may be associated with some underlying patellofemoral malalignment or may be idiopathic, occurring in individuals without evidence of any underlying abnormality.

Idiopathic anterior knee pain is common in adolescent girls. The pain is often activity related, is poorly localized, and may cause disability. It has been described as the *headache of the knee*. About one-third of these patients have features of the MMPI found in individuals with nonorganic back pain. Its natural history is one of spontaneous improvement over a period of years.

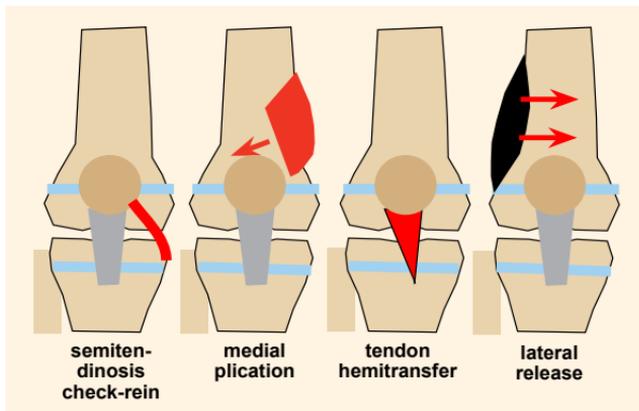
Manage by NSAIDS, isometric exercises, activity modification, and reassurance. Avoid arthroscopy and lateral release procedures.

Structural anterior knee pain is more serious and often requires operative correction.

Identify the underlying dysplastic features such as lateral tibial torsion, genu valgum, patella alta, vastus medialis hypoplasia, lateral tether, shallow sulcus, or excessive joint laxity. Consider imaging the patellofemoral joint with a CT scan to rule out maltracking [1, opposite page].

Manage first with NSAIDS and isometric exercises. During the first visit, introduce the possible need for a realignment procedure [1]. Identify and, if possible, quantitate the severity of each dysplastic feature. Thoughtfully place the operative incision [2].

Correct obvious manageable deformities early. In other cases, the decision is difficult. For example, bilateral double-level osteotomies are necessary to correct severe rotational malalignment.



1 Components of operative repair.

These components are usually combined to correct all dysplastic features. The lateral release alone is usually inadequate.



2 Operative knee scars. Knee scars cause considerable disability (red arrow). A mid-line vertical incision is optimal for extensive realignment procedures (yellow arrows).

Knee Flexion and Extension Deformities

Knee flexion and extension deformities are common and disabling. They have many causes, including congenital contractures, deformities from neuromuscular disorders, trauma, and infection.

Congenital Hyperextension

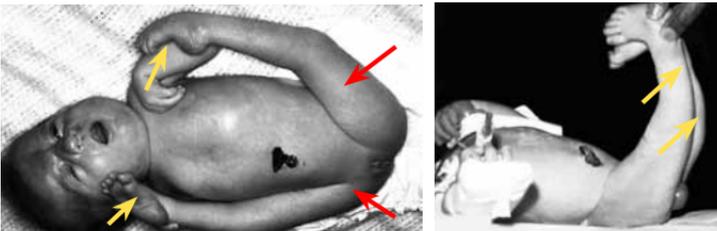
This deformity is often associated with other conditions [1] such as arthrogryposis, spina bifida, developmental hip dysplasia, and clubfeet. In many cases, the child was born breech.

Pathology depends upon severity. In dislocated knees, fibrosis of the quadriceps muscle, absence of the suprapatellar pouch, and valgus deformity of the knee are often present.

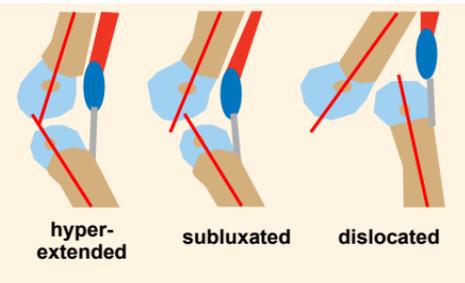
Evaluation Look for other abnormalities. Make a radiograph of the pelvis to make certain the hips are not dysplastic or dislocated. Ultrasound or MRI imaging may be necessary to assess the knee. Grade the severity of the deformity [2].

Management Manage by gentle stretching and casting [3] or by using the Pavlik harness if the knee can be flexed to 60°. For knee dislocations, perform a quadriceps lengthening at about 1–3 months of age [1, opposite page]. Immobilize in 90° of flexion for about one month. Consider correction of other deformities such as hip dislocations and clubfeet concurrently.

If treatment is delayed [2, opposite page], management is more difficult. Limited quadriceps lengthening may move the arc of motion into a more functional plane. In older children or adolescents, bony deformity may require flexion osteotomy to improve alignment.



1 Typical hyperextension deformity in the newborn. This deformity is common in breech presentations and is often associated with other deformities such as dislocated hips (red arrows) and clubfeet (yellow arrows).



2 Classification of extension deformities. These deformities can be classified by severity. Based on Curtis and Fisher (1969).



3 Nonoperative management. Serial cast correction is usually effective in hyperextended or subluxated hips.

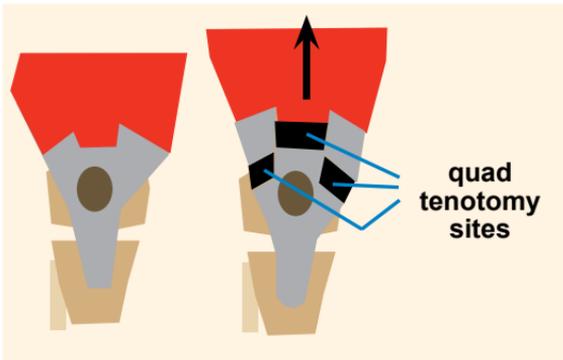
Prognosis is determined by severity [3]. It is generally better for unilateral cases with early operative correction.

Acquired Recurvatum Deformity

Bony deformity of the upper tibia usually results from trauma to the anterior proximal tibial physis [1, next page]. This portion of the physis is vulnerable to arrest. Recurvatum has been reported following traction, spica cast immobilization, proximal tibial traction pin placement, femoral shaft fractures, and meningococcal infections.

Evaluation Make radiographs of the upper tibia. Note that the tibia is usually inclined posteriorly by about 9°. Assess the status of the growth plate with MRI or CT scans.

Management Consider resection of physeal bars of the anterior tibia if 2 years of growth remain. At the end of growth, correct the deformity with an opening wedge osteotomy just proximal to the insertion of the patellar tendon [2, next page].



1 Percutaneous quadriceps recession. This procedure is performed in early infancy through three percutaneous incisions. The infant is immobilized in a spica cast for 4–6 weeks. From Roy and Crawford (1989).



2 Recurvatum during childhood. This deformity has been present since birth. It causes the child considerable disability.

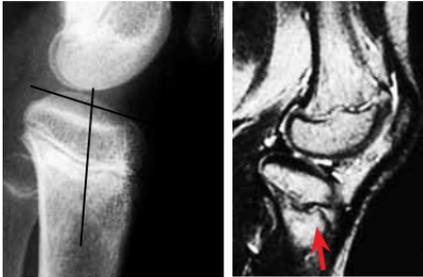


3 Quadriceps lengthening. This child had an open quadriceps lengthening of the right knee through a vertical mid-line approach.

Flexion Deformity in Neuromuscular Disorders

Congenital and acquired flexion contracture deformities are common in children with neuromuscular problems. Acquired deformities result from imbalance between the quadriceps and hamstrings. Flexion deformity is common in arthrogryposis [3], cerebral palsy (Fig. 6.49), and myelodysplasia.

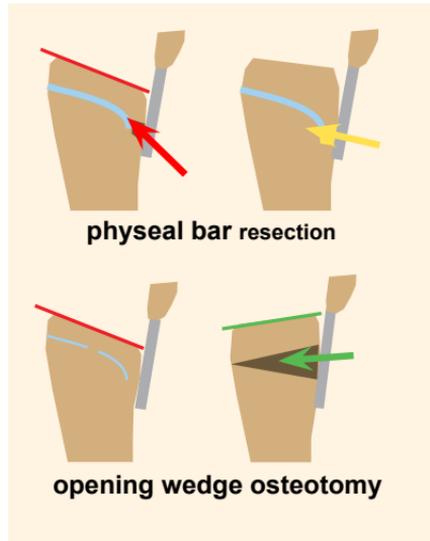
Manage by restoring muscle balance and correcting secondary bone deformity (see Chapter 14).



1 Recurvatum from physeal injury. Note the anterior inclination of the proximal tibia (red line) and irregular physis on the MRI (yellow arrow). Normally the tibial articular surface inclines posteriorly by about 9° (green lines).



3 Knee flexion in arthrogryposis. This child was unable to walk because of the knee flexion contracture. Following correction by soft tissue releases and femoral shortening, the child became ambulatory.



2 Correction of physeal bar. This child was treated with a proximal tibial pin (blue arrows) causing a physeal bar (red arrow) and a secondary recurvatum deformity (red lines). This bar was resected and the defect filled with fat (yellow arrows). The established deformity at the end of growth can be corrected by an opening wedge osteotomy (green arrow) just proximal to the tibial tubercle.

Direction	Comment	Natural history
Lateral	Physiological in infancy	Resolves
Anterior	With other deformities	Persists
Posteromedial	Classic pattern	Resolves incompletely
Anterolateral	Pre-pseudoarthrosis	Progressive deformity

4 Patterns of tibial bowing. The direction of the apex of the tibial bow determines the prognosis and management. Simple lateral bowing is benign in contrast with anterolateral bowing, which often leads to pseudoarthrosis of the tibia.

Tibial Bowing

Tibial bowing is common and varied. The prognosis varies depending upon the direction of the apex or convexity of the bowing [4 opposite page].

Lateral Tibial Bowing

Lateral tibial bowing is common in infants and is simply a variation of normal [1]. The condition is usually mild, symmetrical, and unassociated with other problems. Reassure the family and provide a follow-up if necessary. Radiographs are usually unnecessary.

Anterior Tibial Bowing

Anterior bowing is often associated with fibular hemimelia [2]. Sometimes a dimple is present over the apex. Limb shortening is the major problem.

Focal Fibrocartilagenous Dysplasia

This rare deformity has a characteristic radiographic appearance [3]. The lesions tend to heal with growth.

Posteromedial Tibial Bowing

Posteromedial bowing [1 next page] is a rare condition associated with a calcaneal deformity of the foot and mild limb shortening. The condition may be due to abnormal intrauterine position. The calcaneal foot deformity resolves with time. The bowing improves with growth. Shortening tends to increase with time and correction by epiphysiodesis or lengthening is often necessary.

Anterolateral Tibial Bowing

Anterolateral bowing is a serious form of tibial bowing [2 next page]. The bowing may increase spontaneously and fracture at its apex. This leads to a pseudoarthrosis of the tibia that is difficult to manage. Anterolateral bowing is managed by protection with a cast or brace to prevent fracture or by operatively augmenting the bone strength to reduce the risk of fracture.

Pseudoarthrosis of the Tibia

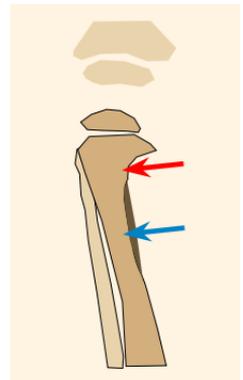
Pseudoarthrosis of the tibia results from a pathological fracture [3 next page] that may occur before or after birth. It may be preceded by an anterolateral bowing of the tibia and is sometimes associated with neurofibromatosis. The pseudoarthrosis occurs in the distal tibial diaphysis and can be graded by severity [1 on page 237].



1 Physiologic bowing. This physiologic lateral tibial bowing resolves spontaneously in late infancy. Radiographs are usually not necessary.



2 Anterior bow in fibular deficiency.



3 Focal fibrocartilagenous dysplasia. Note the lucency (red arrow) and the sclerosis (blue arrow).

Management is extremely difficult, and some patients eventually require amputation because of a failure to achieve union. Operative correction is necessary, with several options available [2, next page]. First, stabilize with an intramedullary (IM) rod and a graft to promote union [3, next page]. If this fails, place a vascularized graft from the other fibula or immobilize with an Ilizarov device. The Ilizarov fixator allows a segment of diaphysis to be transported. The pseudoarthrosis is compressed while the proximal metaphysis is lengthened. If successful, union without sacrificing length is achieved. Place an IM rod to prevent recurrent deformity. Trials using electrical stimulation to promote union have had limited success.

Union may not be achieved even after several procedures. In other cases, only a tenuous union is obtained. The tibia is dysplastic and may refracture, and the leg is short and weak. The outcome is unsatisfactory and amputation is required.

Isolated Fibular Pseudoarthrosis

Rarely, only the fibula is affected by the pseudoarthrosis [3]. Manage by plate fixation, autogenous grafting, and correcting ankle valgus. If the pseudoarthrosis persists, create a synostosis between the distal fragment and the tibia to prevent further shortening.



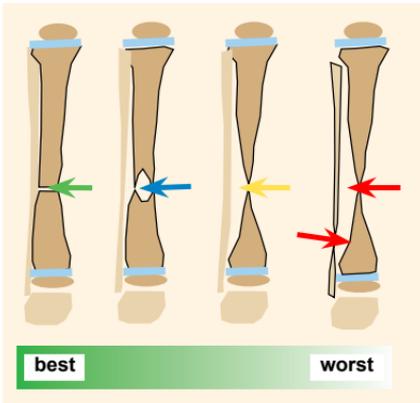
1 Posteromedial tibial bowing. This bow was present at birth (red arrows). The bowing improved gradually throughout childhood, as shown at age 3 years (yellow arrows) and 10 years (blue arrow). This is an unusually severe form with persistent bowing and shortening of 4.5 cm. The deformity was corrected by an osteotomy and lengthening (white arrow) with a circular frame at age 12 years.



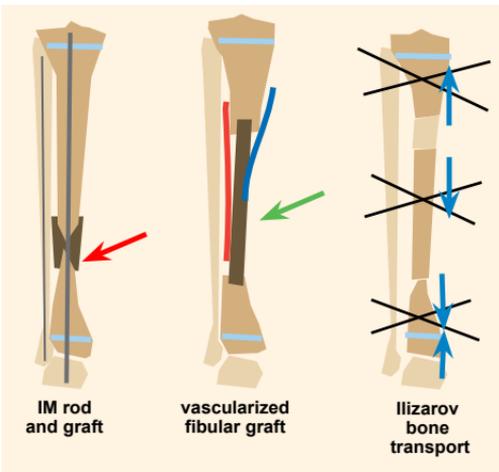
2 Anterolateral bowing and pseudoarthrosis of the tibia. This girl has a pseudoarthrosis of the right tibia (arrows).



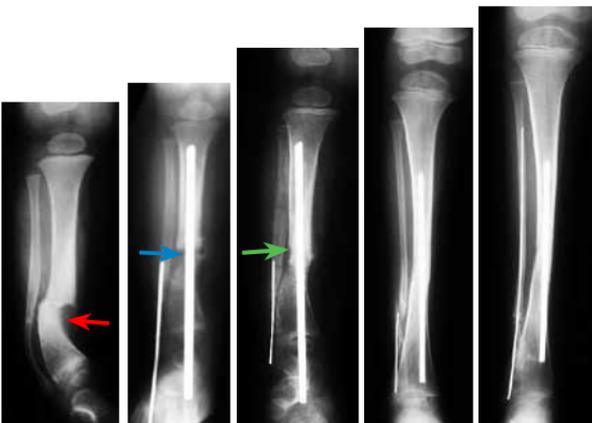
3 Fibular pseudoarthrosis.



1 Prognosis of tibial pseudoarthrosis. outlook is best for simple pseudoarthrosis (green arrow), cystic (blue arrow), sclerotic (yellow arrow) and worst for sclerotic type with pseudoarthrosis of the fibula (red arrows)



2 Pseudoarthrosis tibia treatment methods. The most successful methods for treating this defect are the intermedullary rod and autogenous bone graft (red arrow), the vascularized fibular graft (green arrow), and the ilizarov method with compression of the lesion and lengthening of the proximal tibia.



3 Management by intramedullary fixation and grafting. This infant was born with an anterolateral bow. He was managed in a clam-shell brace. While bathing without the brace, the infant fell and fractured the dysplastic tibia (red arrows). This was treated with IM rod fixation and grafting (blue arrow). Tenuous union was achieved (green arrow). The fibula remained unhealed at age 4 years, and ankle valgus developed. A distal tibial–fibular fusion was created to avoid further valgus from developing.

Chapter 7 – Hip

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General

Problems of the hip account for about 15% of the practice of orthopedists. Many hip problems in adults have their origin during growth.

Development

Ossification of the ischium, ilium, pubis, femoral shaft, and distal femoral epiphysis occur before birth. The femoral head ossifies between the second and eighth postnatal months. [1] and fuses with the neck between 15 and 21 years in boys and one year earlier in girls.

Growth of the upper femur occurs not only in the capital epiphysis and trochanteric epiphysis but also along the neck of the femur [1 next page]. Trauma to specific sites causes specific types of deformities [2 next page].

Most growth of the acetabulum occurs from the triradiate cartilage. Closure will cause severe progressive dysplasia. Additional growth of the acetabulum occurs from the acetabular epiphysis. This growth is especially important late in childhood and during adolescence.

Damage to these growth centers, either from trauma or as a complication of treatment, is a common source of deformity and disability. The upper femur is very susceptible to vascular or epiphyseal injury.



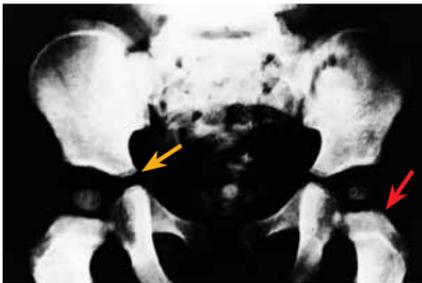
1 Ossification of proximal femur. This sequence shows ossification in the normal child. Redrawn from Tönnis (1984).

Vascularity

Disturbances in blood supply to the upper femur are a common cause of many serious deformities and subsequent disability.

The femoral head may receive blood through the ligamentum teres, epiphyseal vessels, or metaphysis. The femoral head in the infant is supplied by epiphyseal vessels and vessels that traverse the epiphyseal plate [3]. These transepiphyseal vessels disappear as ossification develops in the femoral head. Circulation in the child is primarily through the metaphyseal vessels. Only in late childhood and adolescence do the ligamentum teres vessels make a significant contribution. After closure of the capital epiphyseal plate, the metaphyseal vessels contribute to the circulation.

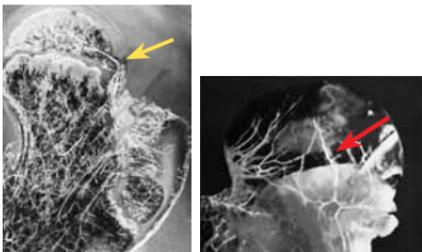
During most of childhood, two anastomotic rings formed by the medial and lateral circumflex vessels provide blood [4]. These signs are variable and deficiencies may contribute to the development of avascular necrosis.



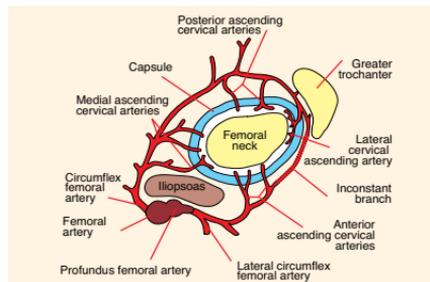
1 Pelvic growth. This child had a phosphorized oil dietary supplement as a child. Growth patterns are shown. Note the growth that occurs in the triradiate cartilage (orange arrow) and upper femur (red arrow). Courtesy I. Ponseti.



2 Proximal femoral growth. Note that growth (red arrows) occurs at many sites about the upper femur, including appositional growth of the femoral neck. Damage to the greater trochanteric apophysis from curettage for a bone cyst (yellow arrow) or from reaming to place an IM nail (orange arrow) causes deformity.



3 Vascularity of the femoral head. In infancy, transepiphyseal vessels are often present (red arrow). In childhood, the femoral head is supplied by the lateral retinacular vessels that must traverse the joint (yellow arrow). From Chung (1976).



4 Vascularity of the femoral head. Note that the proximal femur is supplied by an arcade of vessels that arise from the profundus femoral artery.

Biomechanics

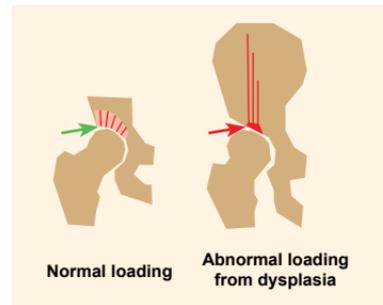
Loading within the joint is affected by the load-bearing area [2]. Increased loading is prominent when the hip is subluxated or shallow. Increased loading leads to osteoarthritis in adult life.

Operative procedures, especially osteotomies of the pelvis and femur, dramatically affect the biomechanics of the hip. The hip joint normally carries about four times its body weight. Hip joint loading is reduced by varus femoral osteotomy or by medializing the joint, as done in the Chiari osteotomy. When reconstructing a hip, try to achieve as normal anatomy as possible.

Nomenclature

Hip terminology is reasonably straightforward [1]. The most significant recent change was the replacement of the term *congenital* with *developmental* in hip dysplasia. Congenital hip disease (CDH) thus becomes developmental hip dysplasia (DDH). Hip disorders caused by muscle disorders secondary to neurologic disorders such as cerebral palsy are called *neurogenic* dysplasia of the hip (NDH). The term *dysplasia* is a broad term covering disorders that may involve the acetabulum, upper femur, or both elements.

Term	Definition
Coxa	Refers to Joint
Vara	Reduction neck–shaft angle
Valga	Increased neck–shaft angle
Plana	Flattening femoral head
Magna	Enlarged femoral head
Breva	Shortened femoral neck
Hip dysplasia	Abnormal features of hip joint
Acetabular	Dysplastic acetabulum
Femoral	Dysplastic femur
Joint status	Acetabulofemoral relationship
Congruous	Concentric reduction
Subluxated	Loss of concentricity
Dislocated	No joint elements in contact
Joint fit	Joint surface relationship
Spherical	Round femoral head
Congruous	Congruous fit
Incongruous	Incongruous
Aspherical	Femoral head nonspherical
Congruous	Congruous fit
Incongruous	Incongruous



1 Nomenclature for deformity. These terms are commonly used to describe various patterns of hip deformity.

2 Biomechanics of the hip. In the normal hip, loading (green arrow) is low and well distributed. In dysplasia, loading is concentrated (red arrow), resulting in eventual cartilage degeneration.

Evaluation

A thorough evaluation of the hip is important due to the vulnerability of the hip joint to damage, especially from impaired blood supply. Delays in diagnosis of DDH, septic arthritis, and slipped epiphysis are relatively common and sometimes result in joint destruction. The deep position of the hip joint makes evaluation more difficult than most extremity joints such as the knee or ankle. This, together with its tenuous vascularity, places the hip at special risk.

History

Is there a family history of hip problems? DDH occurs in families. Has the child complained of pain? Night pain suggests a neoplastic origin. Remember that hip pain may be referred to the knee [1]. Has the child limped? Were there systemic signs? Has the problem been getting worse or plateaued? Be certain to rule out septic arthritis and slipped epiphysis as acute disorders and DDH as a long-term problem.

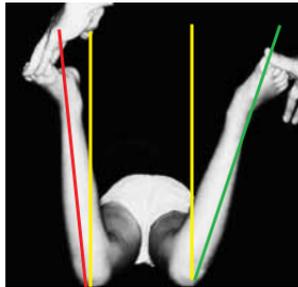
Physical Examination

Observation Does the child appear ill? Is there spontaneous movement of the limb? Pseudoparalysis is common in trauma and infections. Does the child limp? Limping from hip problems is usually antalgic or due to an abductor lurch.

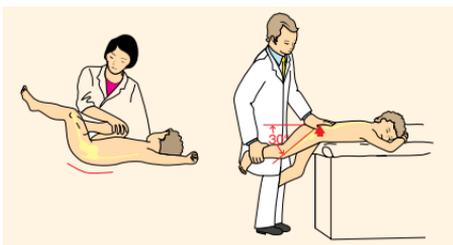
Palpation Palpate for tenderness over the bony prominences. Tenderness is often found in the adolescent with bursitis, tendonitis, or overuse syndromes. By determining the point of maximum tenderness exactly, a presumptive diagnosis can often be made.



1 Hip pain referred to knee. The obturator nerve supplies articular branches to the hip and cutaneous coverage about the knee. Hip disorders may present with knee pain.



2 Hip rotation test. Position the child prone with the knees flexed to 90°. Rotate the hips internally and note any guarding and the extent of rotation. Asymmetry of rotation is usually abnormal. In this child with Legg–Calvé–Perthes disease on the left hip, rotation was limited as compared to the right hip.



3 Hip flexion contracture assessment. The Thomas test (left) is performed with the contralateral hip flexed. Extend to measure the degree of contracture. The prone extension test (right) is performed with the child prone. Gradually extend the hip until the hand on the pelvis begins to rise. The horizontal–thigh angle indicates the degree of contracture.

Range of Motion Hip disorders often result in loss of motion. Inflammatory disorders usually cause a reduction in internal hip rotation early on and eventually flexion and adduction contracture of the hip.

- *Hip rotation* Assess with the child prone. Assessing the range of medial rotation is a valuable screening test [2 opposite page]. The finding of asymmetric hip rotation is abnormal and indicates the need for a radiograph of the pelvis.
- *Flexion* Detect the presence of a contracture using the Thomas or prone extension test [3 opposite page]. The prone extension test is most accurate, especially in children with neuromuscular disorders.
- *Abduction–Adduction* Assess while stabilizing the pelvis with one hand.

Trendelenburg Test

Assess an abductor lurch using the Trendelenburg test [1]. Ask the child to lift one leg at a time. The pelvis should rise on the elevated side. A drop of that side is a positive sign and suggests that the abductor mechanism is weak on the opposite side. This lurch may be due to weakness of the muscles, a change in shape of the upper femur, or inflammation of the joint.

Laboratory Studies

A CBC and ESR are often helpful in evaluating hip disorders. The ESR and CRP are useful in differentiating septic arthritis from toxic synovitis. Infections usually elevate the ESR above 25–30 mm/hr. Toxic synovitis causes only a slight elevation in the ESR and CRP. Hematologic disorders such as leukemia and sickle cell disease may cause pelvic pain.

Hip Joint Aspiration

The aspiration of the hip is the most certain method of establishing the diagnosis of septic arthritis. Aspirate the joint promptly if the diagnosis of septic arthritis is seriously included in the differential. Although a negative aspirate (even when documented by an arthrogram) is not absolutely definitive, it is highly suggestive that the problem is not within the joint.

Delays in diagnosing septic arthritis may be catastrophic because it jeopardizes the vascularity to the femoral head and articular cartilage. Joint aspiration does not affect bone scans and should not be delayed by plans to perform imaging procedures.



1 Trendelenburg test
This girl has DDH with weakness of the left hip abductors. When standing on her right leg, right hip abductors contract to elevate the left pelvis to maintain the head centered over the body (green lines). When standing on the weaker left leg, abductor weakness allows the right pelvis to fall (blue arrow). She must then shift her weight over the left leg (red lines).

Imaging

Imaging is required to evaluate hip disorders in children. Imaging is the only way to establish a prognosis. The vast majority of hip problems in children can still be managed adequately by careful examination and conventional radiographs.

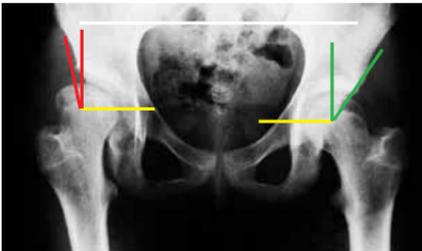
Conventional radiography Evaluate most hip problems with conventional radiography. Except for the initial study, use a gonad shield. Obtain a single AP study [1]. Several useful measures may be made from this simple study [2]. Note any asymmetry in ossification of the pelvis. A painful condition such as an osteoid osteoma results in hemideossification [3]. Be aware of the situations in which false negative studies are commonly misleading. A negative study does not rule out DDH in the neonate or an early septic arthritis. An AP radiograph may not show a mild slipped capital femoral epiphysis (SCFE).

Add other views as necessary. The frog-leg lateral allows comparison of both upper femora. The true lateral is useful in assessing the degree of slip in SCFE or the degree of involvement in Legg–Calvé–Perthes disease [1 opposite page].

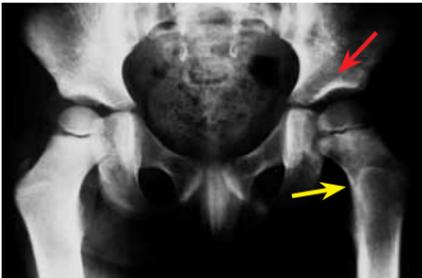
Useful special views include the abduction–internal rotation study for hip dysplasia [2 opposite page], maximum abduction and adduction views for assessing hinge abduction problems, and anteversion studies. Femoral anteversion measurement is seldom necessary.



1 AP X-ray of pelvis. Much can be learned from this simple study. The right hip is normal. Acetabular dysplasia is present on the left. Note the triangular shape of the tear drop (red arrows). Note that the joint space (orange line) is widened. Shenton's line (green lines) is disrupted. The sercil (yellow arrows) is sclerotic. The left hip joint is slightly higher and more laterally positioned than the normal side.



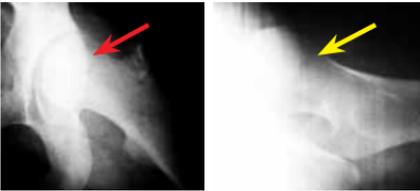
2 Center–edge (CE) angle. This child has a normal left hip with a CE angle of 30°. The right hip is aspherical and subluxated and the CE angle is 10°. Note that measures are made with the pelvis level (white line).



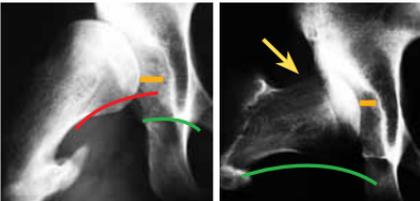
3 Hemideossification. Note the bone loss of the left hemipelvis (red arrow) due to an osteoid osteoma (yellow arrow) of the proximal femur.

The load-bearing area of the hip significantly affects its longevity. A reduction in this area may be due to one or more of the following factors:

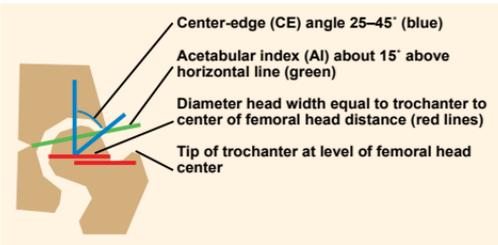
- **Simple hip dysplasia.** The hip joint is either maldirected or shallow. Both reduce contact area. The depth of the acetabulum is often assessed by the CE angle. This angle increases during childhood as the joint ossifies. At the end of growth, values are like those of adults with a normal range of 25°– 45°. Features of the normal hip are used as a basis for assessing deformity [3] and planning reconstruction.
- **2. Incongruity reduces contact area.** The femoral head is normally round and matches the shape of the acetabulum ([4]. An aspherical femoral head is usually due to vascular problems. In the young child, the acetabulum usually remodels to become congruous and the hip become aspherical and congruous [1 next page]. If acetabular remodeling fails to occur, the hip may be aspherical and incongruous—a bad combination.
- **3. Displacement of the femoral head.** The relationship between the femoral head and acetabulum is normally congruous. If the head is displaced, it becomes subluxated. If all cartilage contact is lost, the joint is dislocated.



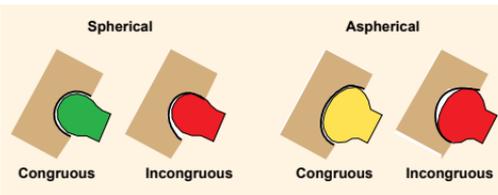
1 Lateral X-rays of the proximal femur. Frog-leg lateral (red arrow) is only an oblique view. A true lateral (yellow arrow) requires special positioning but provides more information as it is made at right angles to the AP view.



2 Abduction internal rotation (AIR) view. The resting position (red arrow) shows the hip in a 14-year-old child with cerebral palsy. The hip is subluxated (orange lines), and Shenton's line (green arc) is disrupted (red arc). The AIR view (yellow arrow) shows improved congruity and less subluxation and restoration of Shenton's line.



3 Normal measurements. These are measurements of the normal adolescent hip.



4 Congruity. Congruity of the hip may be either spherical or aspherical and congruous or incongruous. Incongruity (red) results in areas of excessive load, causing excessive cartilage wear and eventually osteoarthritis.

Ultrasonography (US) Ultrasound studies are of greatest value when readily available and performed by an orthopedist in conjunction with the overall evaluation. Cost, restricted access, and operator inexperience may limit value. Ultrasound’s greatest potential usefulness is in assessing DDH in early infancy. Assessing joint effusion, localizing abscesses, and assessing the severity of SCFE, head size in LCP disease, and neck continuity in coxa vara are other applications. This imaging technique is underutilized in North America.

Scintigraphy Bone scans (BS) are useful in localizing inflammatory processes about the pelvis [2] and in assessing the circulation of the femoral head. Order high-resolution or pinhole-collimated AP and lateral scans of both proximal femora when assessing avascular necrosis (AVN). The bone scan is useful in confirming a preslip and assessing bone tumors.

Arthrography The usefulness of this procedure is limited, as it is invasive and requires sedation or anesthesia. Arthrography is appropriate to confirm joint penetration in negative taps for suspected joint sepsis and for special situations in managing DDH. The role in LCP disease is more controversial.

Magnetic resonance imaging (MRI) These studies are the most expensive and require sedation for infants and young children. MRI studies are most useful in assessing intrarticular disorders of the hip. Cartilagenous loose bodies or fracture fragments, deformity of the cartilagenous femoral head, status of the growth plate, and avascular necrosis are usually definable.

Computerized tomography (CT) Order CT studies to evaluate inflammatory conditions such as an iliopsoas abscess or the configuration of the upper femur and acetabulum. CT scans have replaced tomography in assessing AVN and physeal bridges.

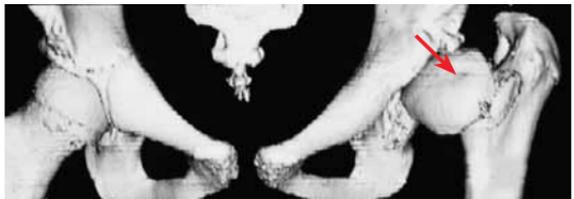
Three-dimensional CT reconstructions are often helpful in visualizing complex deformities of the hip necessary when planning reconstructive surgery [3].



1 Aspherical congruity. This deformity resulted from Legg–Calvé–Perthes disease during middle childhood. The head became flattened and the acetabulum remodeled to become congruous.



2 Imaging options. This bone scan shows inflammation of the sacroiliac joint (red arrow), and the MRI shows a slipped epiphysis (yellow arrow).



3 Three-dimensional CT reconstruction. Reconstructions are useful in assessing complex hip deformity prior to reconstruction. This deformity is secondary to avascular necrosis associated with DDH management in infancy.

Hip and Pelvic Pain

The causes of hip and pelvic pain are numerous [1], sometimes making the diagnosis difficult.

Diagnosis

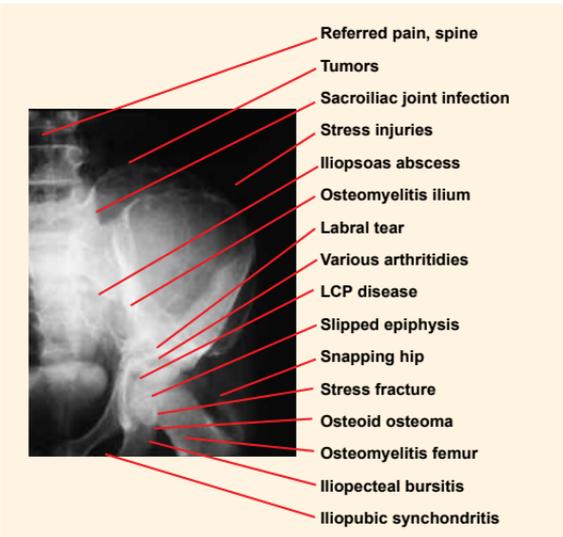
Detailing these features may help establish the diagnosis.

Age LCP disease is most common in boys in middle childhood [2]. SCFE must be considered in the older child or adolescent. Overuse syndromes are most common in the adolescent.

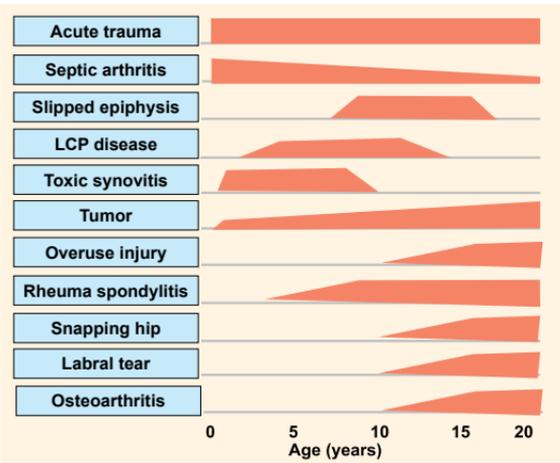
Onset Acute onset is suggestive of injury or a rapid onset of infection. SCFE may be chronic or sudden. Acute slips are characterized by a mild injury and inability to walk. LCP disease onset is usually insidious. Overuse syndromes are most painful when active.

Spontaneous movement The most consistent physical finding for septic arthritis of the hip is a loss of spontaneous movement of the affected limb.

Systemic illness The child is ill with septic arthritis, and less sick with toxic synovitis, rheumatoid spondylitis, and tumors.



1 Causes of hip pain in children. The differential diagnosis is extensive.



2 Hip pain causes by usual age of onset. These are ages when conditions are most common.

Resting position of the limb Intraarticular hip disorders usually result in the spontaneous positioning in slight flexion and lateral rotation [1]. This position reduces the intraarticular pressure.

Tenderness Palpate to determine the site of tenderness [2].

Hip rotation test Guarding and a loss of medial rotation suggest the problem is within the joint [3].

Night pain Nocturnal pain suggests the possibility of a malignant tumor.

Back stiffness Limitation of forward bending suggests that the disorder may be referred from the spine.

Causes

Establish the diagnosis by considering the features and the common causes of hip pain [1 opposite page].

Infection is a common cause of pelvic pain. The early diagnosis of septic arthritis is critical because it may severely damage or destroy the hip joint [4]. Because of the tenuous vascularity of the hip, joint infections must be diagnosed and drained promptly. Soft tissue abscess, such as the psoas abscess, may be suspected by the finding of tenderness on rectal examination and soft tissue swelling on the AP radiograph of the pelvis. Confirm the diagnosis by CT or MRI studies. Sacroiliac infections are identified by bone scans.



1 Observation. Observation revealed pseudoparalysis of the left leg, and the left hip is positioned in slight flexion and external rotation. These findings are typical for septic arthritis of the left hip.



2 Thigh tenderness. Palpate for localized tenderness.



3 Hip rotation test. This child has reduced medial rotation of the right hip due to toxic synovitis.



4 Joint damage from septic arthritis. Note the severe joint damage from septic arthritis treated 2 weeks after onset.

Stress injuries or repetitive microtrauma may cause hip pain. Such pain is most common during the second decade and often follows vigorous activity. It may involve the upper femur but more commonly involves the origin of muscles such as the greater trochanter and iliac spines. The diagnosis is usually suggested by the history, physical findings of well-localized tenderness, and negative radiographs but a positive bone scan.

Tumors A variety of tumors occur about the hip and pelvis. Osteoid osteoma is common in the proximal femur and produces pain in a pattern that is nearly diagnostic. The pain is nocturnal and relieved by aspirin. The tumor produces reactive bone with a radiolucent nidus on conventional radiographs [2].

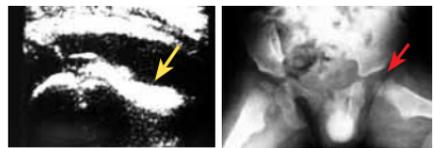
Toxic synovitis (or transient synovitis) is an idiopathic benign inflammation of the hip joint [3] that occurs in children. This condition is important, as it may be confused with septic arthritis and less commonly with LCP disease. The condition causes pain and irritability of the hip. It subsides over several days spontaneously.

Idiopathic chondrolysis This uncommon condition is seen in late childhood or adolescence. The hip becomes painful, and stiff and joint space narrowing is present [4].

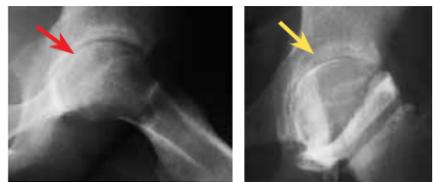
Rheumatoid spondylitis Unlike juvenile rheumatoid arthritis, hip involvement may be the first sign of rheumatoid spondylitis. Establish the diagnosis with serologic tests.



2 Osteoid osteoma. These lesions are common in the proximal femur and cause night pain.



3 Toxic synovitis. The hip is often positioned in slight flexion and external rotation (red arrow). Ultrasound studies often show an effusion (yellow arrow).



4 Chondrolysis. This hip shows narrowing of the joint space on conventional radiographs (red arrow), and the arthrogram (yellow arrow) shows thinning of the cartilage on the femoral head.

	Night pain	History trauma	Sick child	Pseudoparalysis	Hip rotation test	Localized tenderness	Snapping hip	Radiographs	Bone scan	MRI	Ultrasound
Acute trauma		+						+			
Septic arthritis			+	+	+	+					+
Slipped epiphysis					+				+		
LCP disease					+				+		
Toxic synovitis					+						
Tumor	+								+		
Overuse injury		+				+				+	
Rheum. spondylitis					+						
Snapping hip							+				+
Labral tear								+			+
Chondrolysis					+			+			

1 Hip pain and major diagnostic features. These are some of the major clinical features that differentiate each cause of pain.

Developmental Hip Dysplasia

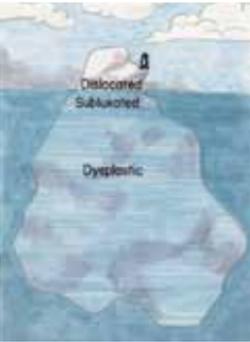
Developmental hip dysplasia (DDH) is a generic term describing a spectrum of anatomic abnormalities of the hip that may be congenital or develop during infancy or childhood. The spectrum covers mild defects such as a shallow acetabulum to severe defects such as *teratologic* dislocations. Teratologic dislocations occur before birth and include severe deformity of both the acetabulum and proximal femur.

Incidence

DDH incidence depends on how much of the spectrum is included [1]. At birth, hip instability is noted in 0.5–1% of joints, but classic DDH occurs in about 0.1% of infants. The incidence of mild dysplasia contributing to adult degenerative arthritis is substantial. It is thought that half of the women who develop degenerative arthritis have pre-existing acetabular dysplasia.

Etiology

DDH is considered to be inherited by a polygenic mode. DDH is more common in breech deliveries [2], in children with joint laxity [3], and in girls.



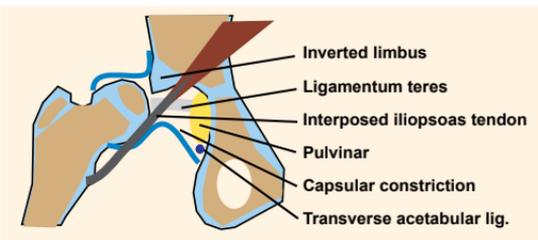
1 Spectrum of hip dysplasia. Dislocated hips are usually diagnosed during infancy but hip dysplasia may not become evident until adult life and then present as degenerative arthritis.



2 Breech association. DDH is often associated with breech presentation.



3 DDH and joint laxity. Children with DDH often show excessive joint laxity.



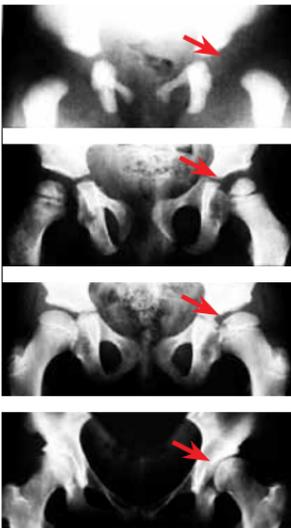
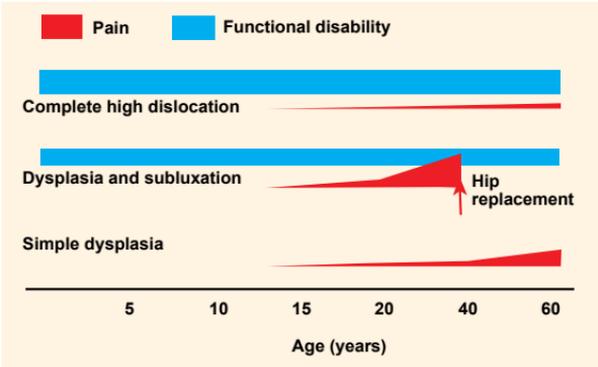
4 Structures blocking reduction in DDH. These interpositions may block reduction of the hip.

Pathology

The acetabulum is often shallow and maldirected. The proximal femur shows antetorsion and coxa valga. Structural interpositions between the displaced femoral head and acetabulum are common [4 opposite page]. The iliopsoas tendon is insinuated between the femoral head and acetabulum, causing a depression in the joint capsule. This gives the capsule an hour-glass configuration. The acetabular labrum is inverted into the joint, the ligamentum teres is enlarged, and the acetabulum may contain fat (pulvinar).

Natural History

Residual acetabular dysplasia is common in DDH. This may occur even following an apparently good early reduction [2]. The disability from dysplasia is related to the degree of displacement [1]. Greater displacement causes more function disability. Pain is most common with severe subluxation or articulation in a false acetabulum [3].



2 DDH with residual acetabular dysplasia. Radiographs at birth, 3, 10, and 19 years (top to bottom) show persisting dysplasia.



3 Adult degenerative arthritis. Note that arthritis is most severe in the subluxated (red arrow) hip as compared with the totally dislocated hips (yellow arrows).

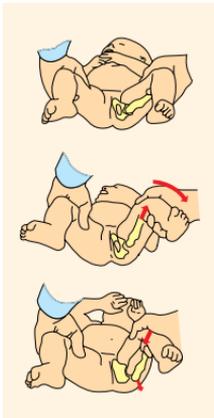
Diagnosis

The early diagnosis of DDH is critical to a successful outcome. Acetabular development is abnormal if a hip is subluxated or dislocated. Delays in management result in residual abnormalities and eventual degenerative arthritis.

Neonatal Examination Every newborn should be screened for signs of hip instability. The hip should be examined using both the Barlow and Ortoloni techniques [1 and 2]. Examine one hip at a time. The infant should be quiet and comfortable so the muscles about the hip are relaxed. Use no force. Test for instability in several positions.

Changing manifestations of DDH The signs of DDH change with the infant’s age [3]. For example, the incidence of hip instability declines rapidly, 50% within the first week. The classic findings of stiffness and shortening increase over the first few weeks of life. These signs become well established in the older infant [4].

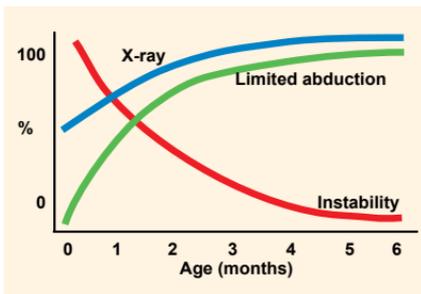
Repeated examinations The hip should be examined during each “well baby” examination. In the neonatal period, DDH is detected by different signs based on the infant’s age. In early infancy, instability is the most reliable sign. Later, limitation of abduction and shortening are common. Beware of the bilateral dislocations, as they are more difficult to identify [1 opposite page]. If hip abduction is less than about 60° on both sides, order an imaging study.



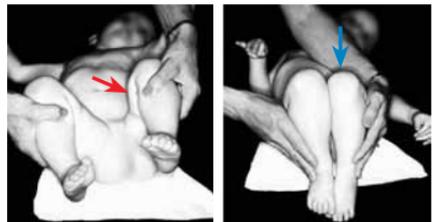
1 Barlow’s sign. Hip instability is demonstrated by attempting to gently displace the hip out of the socket over the posterior acetabulum.



2 Ortoloni’s sign. The thigh is first adducted and depressed to subluxate the hip. The thigh is then abducted. The hip reduces with a palpable “clunk.”



3 Changing signs of DDH. With increasing age, signs change.



4 DDH in older infant. Note the limited abduction (red arrow) and shortening (blue arrow) on the affected left side.

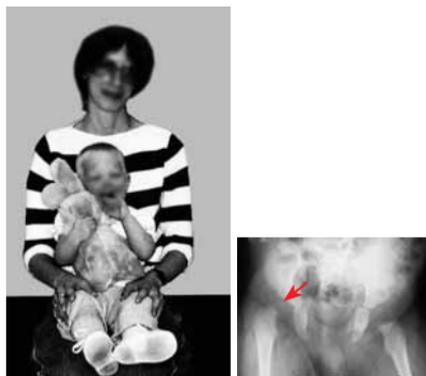
Mother’s intuition Although not proven, a common clinical experience is the accuracy of the mother’s sense that *something is wrong*. Take the mother’s intuition seriously [2].

Hip-at-risk factors The presence of several factors increase the risk of DDH [3]. When risk factors are present, the infant should be examined repeatedly and the hip imaged by ultrasound or radiography.

Hip “clicks” and asymmetrical thigh folds Hip clicks are fine, short-duration, high-pitched sounds that are common and benign. These are to be differentiated from “clunks,” the sensation of the hip being displaced over the acetabular margin. Clicks and asymmetrical thigh folds are common in normal infants [4].



1 Bilateral DDH. This girl has symmetrical bilateral dislocations. The hip symmetry makes early diagnosis more difficult. Note the typical lumbar lordosis (arrow) that occurs with high dislocations.



2 Mother’s intuition. This mother had DDH as a child. She suspected that her son’s hip was abnormal, but the primary care physician found nothing on examination. She insisted on a radiograph. This study demonstrated a dislocation (red arrow). This scenario is not uncommon.

Factor	Comment
Positive family history	Increases risk tenfold
Breech position	Increases risk five – tenfold
Torticollis	Assoc. deformity
Foot deformities	Intrauterine
Calcaneovalgus Metatarsus add.	constraint
Knee deformities Hyperextension Dislocation	Associated with teratogenic type dislocation



3 Risk factors. These factors increase the risk of DDH and signal the need for careful and repeated examinations and imaging studies.

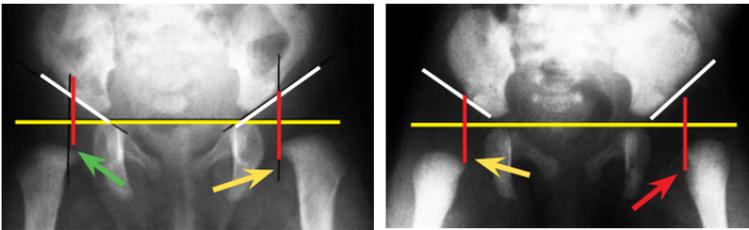
4 Asymmetrical thigh folds. These occur in up to 20% of normal infants.

254 Hip / Development Hip Dysplasia –Diagnosis

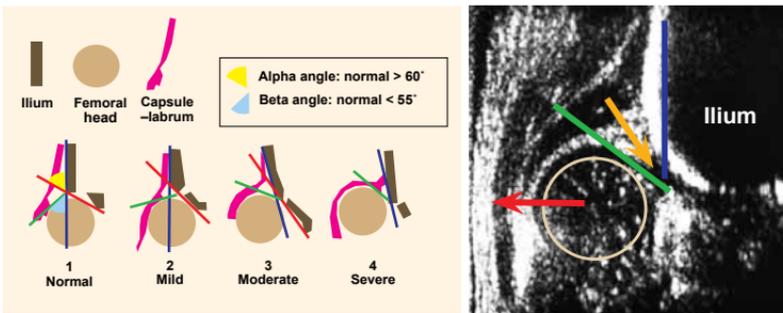
Radiography Radiographs become progressively more diagnostic with increasing age. By 2–3 months of age, radiography is reliable and is the optimum age for screening by this method. A single AP radiograph is adequate. Draw the reference lines and measure the acetabular index (AI). Normally, the AI in early infancy falls below 30° , is questionable in the $30^\circ - 40^\circ$ range, and abnormal if above 40° . Hip subluxation or dislocation may often be demonstrated by the metaphysis of the femur positioned lateral to the lateral acetabular marginal line [1].

Ultrasound imaging The effectiveness of ultrasound imaging depends upon the skill and experience of the examiner. The skillful ultrasound evaluation is an effective screening technique for DDH [2]. The major problem with this screening is the interpretation of the findings. If the hip is unstable, imaging is unnecessary. Imaging is appropriate to evaluate a suspicious finding, when hip-at-risk factors are present, and to monitor the effectiveness of treatment.

Documentation Document your hip evaluation. The failure to diagnose DDH is a common cause of suits against physicians. If the diagnosis is delayed, a record showing that appropriate examinations were made of the hip provides the best defense. DDH may be missed by even the most skilled examiners. Failure to screen for DDH is not acceptable by current standards.



1 Assessing radiographs in early infancy. Before the ossific nuclei are developed, assessing hip dysplasia is more difficult. These two radiographs show how such hips can be assessed. Draw the horizontal reference line (yellow) through the triradiate cartilage space. Construct the acetabular line (white). The inclination of this line measures the acetabular index. Draw vertical lines at the lateral acetabular edge (red). If the proximal femoral metaphysis lies medial to the vertical line, the hip is reduced (green arrow). If on the vertical line, the hip is subluxated (yellow arrows), and lateral to the line, the hip is dislocated (red arrow).



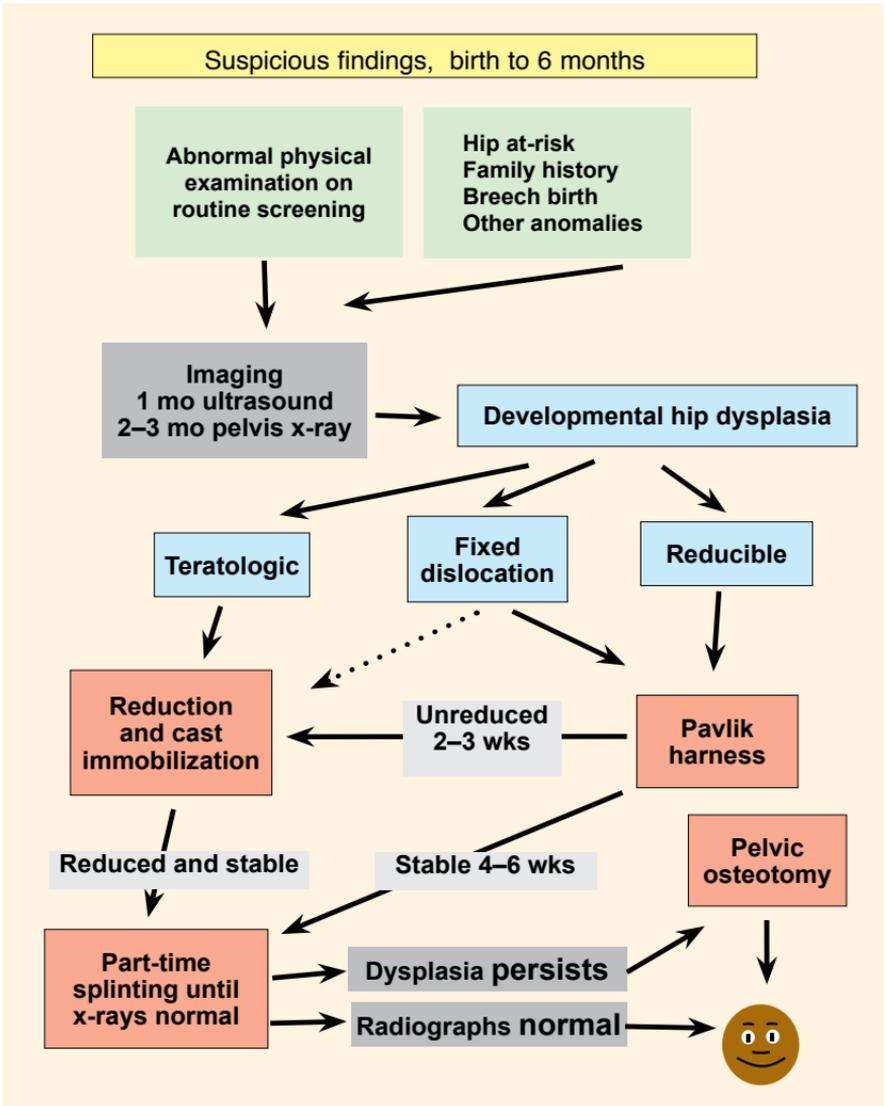
2 Graf grading of DDH by ultrasound. (Left) Drawing shows how the hips can be graded by measurements based on the ultrasound evaluation. The grades shown are divided by Graf into four types. Each is subdivided into subtypes (not shown). Reference lines are drawn to show the iliac margin (blue), the joint inclination (red), and the line head-joint relationship (green). The alpha and beta angles can be constructed to show severity. (Right) The ultrasound image shows a severe displacement (red arrow) of the femoral head (tan circle) in an infant with DDH. The displaced femoral head compresses the labrum and preossified cartilage (orange arrow) against the ilium (blue line).

Management

The management of DDH is challenging. Delays in diagnosis or problems in management often lead to residual anatomic defects and subsequent degenerative arthritis. The objectives of management include early diagnosis, reduction of the dislocation, avoidance of avascular necrosis, and correction of residual dysplasia.

Birth to 6 months

This is the ideal age for management [1]. Treat DDH in this age group first with an abduction orthosis such as the Pavlik harness.



1 DDH management flowchart, birth to 6 months.

Pavlik harness This orthosis is most widely used and allows motion in flexion and abduction. Be certain that it is fitted properly [1] both initially and as applied by the parents. Advise the family on ways of transporting the infant.

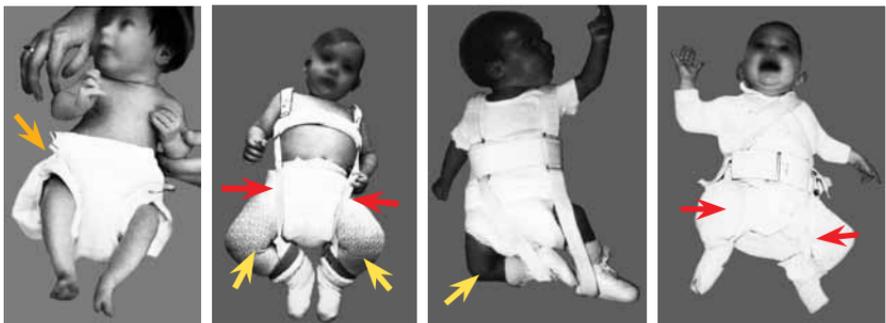
See the infant weekly in the brace. Make certain the brace is being fitted properly [2] and progress is being made. The hip should become progressively more stable.

If harness treatment is successful, continue full-time bracing for 6–8 weeks to allow the hip to become stable. Monitor with ultrasound imaging or by AP radiographs of the pelvis about every 2–4 weeks. Continue the brace at night until the radiographs are normal.

- Failure of Pavlik harness treatment* If a dislocated hip has not reduced by 3–4 weeks, abandon orthotic treatment. Persisting in orthotic management may cause head deformity and posterior fixation, and make closed reduction impossible. Proceed with closed or open reduction. Manage as is described for infants over 6 months of age.



Fig. 7.51 Proper fit of Pavlik harness. The harness should be carefully fitted. Make certain it is the proper size for the infant. The harness must be comfortable. See the fit after the parent applies the harness to assess problems before the parent leaves the clinic.



2 Pitfalls in management. Triple diaper management (orange arrow) is ineffective and gives a false sense that treatment has been initiated. Pavlic harness errors are common. Make certain that the straps are not too tight (red arrows), the calf strap is not too low, the knees are flexed, not the hips (yellow arrows), and the infant is comfortable.

Night splinting After the hip is reduced and stable, continue with night splinting to facilitate acetabular development. Continue until the radiographs are normal. A simple abduction splint is inexpensive and well accepted by the infant.

6 to 18 Months

In this age group, most cases of DDH can be managed by closed reduction and spica cast immobilization [1 on this page and 1 next page].

Traction The need for traction is controversial. The current practice is to omit traction in most cases. Traction may be useful if the hip is stiff and closed treatment is planned. Use home traction when possible. Maintain for about 3 weeks with the legs flexed and abducted about 45° with 2–3 pounds of traction applied to each limb [2].

Scheduling Schedule and obtain consent for a closed, possible open reduction.

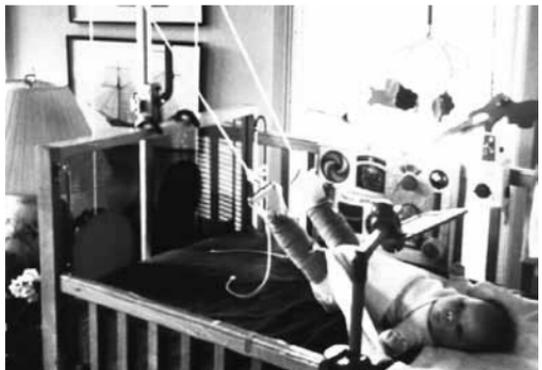
Reduction by closed means is first tried. If unsuccessful, open reduction is required. These procedures are outlined on pages 406 and 407.

Arthrography is useful when the quality of reduction is uncertain or the decision regarding management is difficult.

Follow-up Following reduction, the infant should be followed carefully to assess the effect of time on growth, reduction, and acetabular development. Follow with AP radiographs made quarterly through infancy, yearly through early childhood, and then about every third year during middle and late childhood. The frequency of follow-up studies should be individualized based on the severity of any residual dysplasia.



Fig. 7.55 Spica cast immobilization. Infants require cast immobilization to retain the reduction of a dislocated hip.



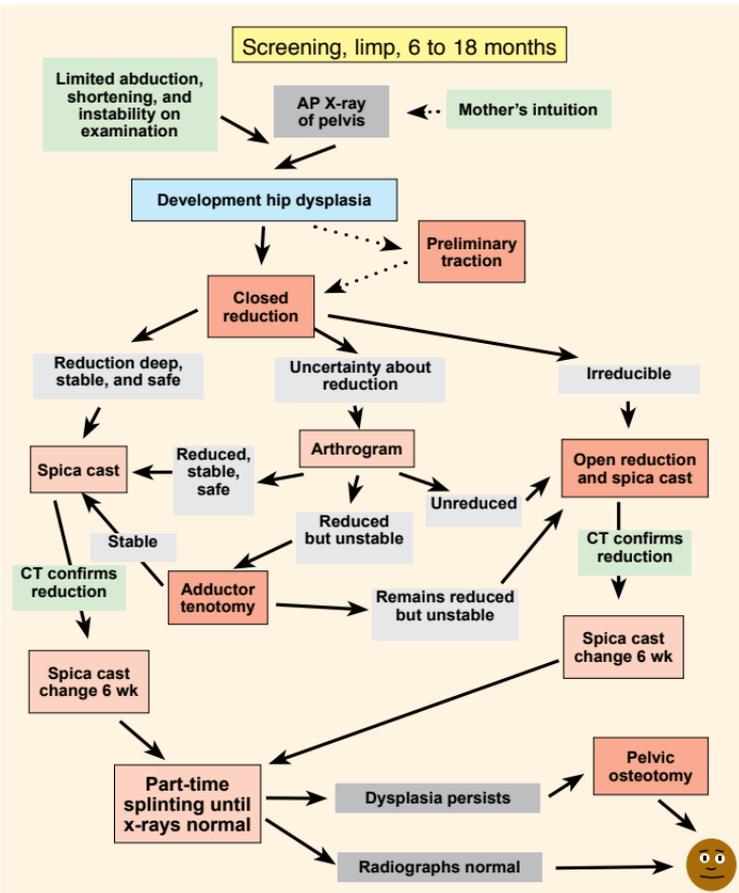
2 Home traction. Setup at home is less expensive, less stressful for the infant, and often more convenient for the family.

18 to 30 Months

In this age group, operative management is usually required [1 next page]. Occasionally, an infant with a “loose dislocation” can be managed as described in the flowchart for infants 6–18 months of age [1]. If the hip is unusually stiff, be prepared to add femoral shortening as described for management of children over 30 months of age.

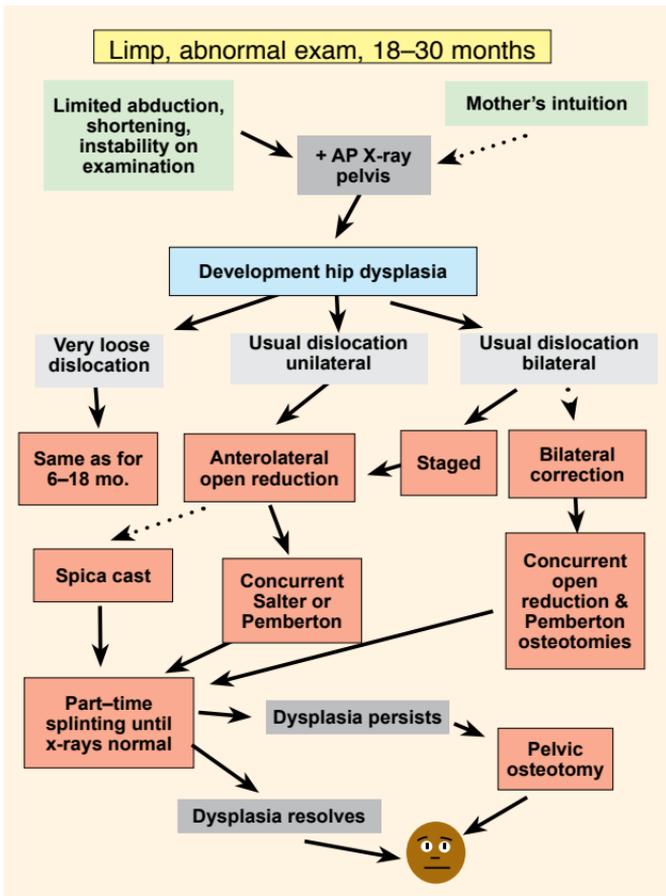
Management Manage with an open reduction through an anterolateral approach and perform a concurrent Salter or Pemberton osteotomy. The open reduction is technically challenging. Add the pelvic osteotomy to improve results and save the child a second procedure.

Open reduction is the most difficult part of the procedure. The pelvic osteotomies are relatively simple, but the reduction can sometimes be difficult. The open reduction requires good exposure, careful dissection to minimize the risk of avascular necrosis, and a concentric reduction. The obstacles to reduction must be corrected [1 on page 260].



1 DDH management flowchart, 6 to 18 months.

- *Iliopsoas tendon* This tendon is interposed between the femoral head and acetabulum and must be released.
- *Capsular constriction* Open the capsule widely to ensure a complete release.
- *Transverse acetabular ligament* This structure lies across the base of the acetabulum and will block a deep concentric reduction unless released.
- *Pulvinar* is fatty fibrous tissue that often fills the depth of the acetabulum. Remove with a rongeur.
- *Ligamentum teres* is elongated and sometimes hypertrophied. Removal is usually required. The vascular contribution through this ligament is minimal.
- *Limbus* is often inverted and hypertrophied. Do not excise this structure. Once the hip is concentrically reduced, the limbus will remodel and form the labrum, an important structure for hip stability and longevity.



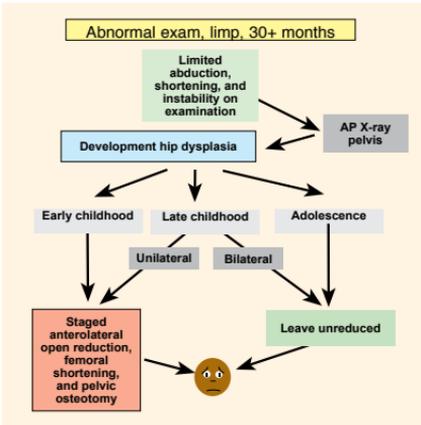
1 DDH management flowchart, 18 to 30 months.

30+ Months

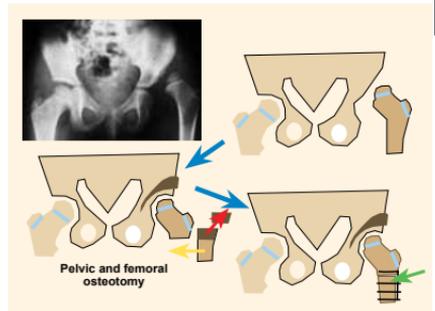
Manage DDH in this age range by open reduction, femoral shortening, and pelvic osteotomy [1 and 2]. If the dislocations are bilateral, correct one side at a time. Allow 6 months between procedures to allow the child to recover. Manage bilateral dislocations on the basis of the age of the child [3].

Selection of pelvic osteotomies Select the type of osteotomy based on the severity of deformity and the age at the time of treatment. Salter osteotomies are good choices because they can be performed at any age, they do not change the shape of the acetabulum, and when combined with a femoral osteotomy, they usually provide adequate correction.

Femoral osteotomy Femoral shortening osteotomy is nearly always necessary. If the deformity is severe, the femoral shortening is performed first, then the open reduction, followed by the pelvic osteotomy. The femoral fragments are then aligned, with gentle traction on the limb. The overlap is then determined and the overlapping distal femoral segment is resected. The procedure is primarily shortening. Slight varus and no rotation are usually appropriate.



1 DDH management flowchart, 30+ months. Outcomes are seldom good or excellent in this age range.



2 Unilateral open reduction and pelvic and femoral osteotomy. This combination of procedures is necessary in the child. Femoral shortening (red arrow) must be added to allow tension-free reduction. The distal fragment is aligned (yellow arrow) and fixed (green arrow).



3 Bilateral DDH in the child. Staged corrections can be done in early childhood. In late childhood or adolescence, leaving the hips unreduced may be prudent.

Avascular Necrosis

Next to achieving a concentric reduction, preventing avascular necrosis (AVN) is of utmost importance. Unless the necrosis is mild, this complication causes altered proximal femoral growth, creates deformity, and often leads to premature degenerative arthritis.

Types The spectrum of AVN [1] includes severe necrosis, extensive physal bridge formation, and shortening of the femoral neck, which leads to degenerative arthritis during adult life. At the other end of the spectrum is the mild resolving form characterized by irregular ossification but without physal bridge formation and subsequent deformity.

Prevention Attempt to prevent AVN by using preliminary traction and open reduction in stiff hips with an obstructing limbus, percutaneous adductor tenotomy, femoral shortening in the child, and immobilization in the “safe” or human position. Despite all precautions, AVN may still occur.

DDH management objectives

- Early concentric reduction
- Avoid avascular necrosis
- Overcome dysplasia

Reduction pitfalls

- Excessive abduction
- Forced positions
- Uncertain value
- Preliminary traction
- Reduction with inverted limbus
- Type of approach for open reduction

Early signs of AVN (Salter)

- Failure of ossific nucleus to appear 1 year postreduction
- Ossific nucleus fragments following reduction
- Ossific nucleus fails to grow following reduction
- Widening of the femoral neck

Temporary changes
 Residual physal bridge



Type 1



Type 2



Type 3



Type 4

1 Classification of AVN patterns. These patterns depend upon the severity and location of the ischaemic necrosis. Based on Kalamchi and MacEwen (1980) classification.



2 Type 2 deformity. This deformity results from a lateral physal bridge (red arrow) causing a lateral tilting of the epiphysis (*tilted hat deformity*).

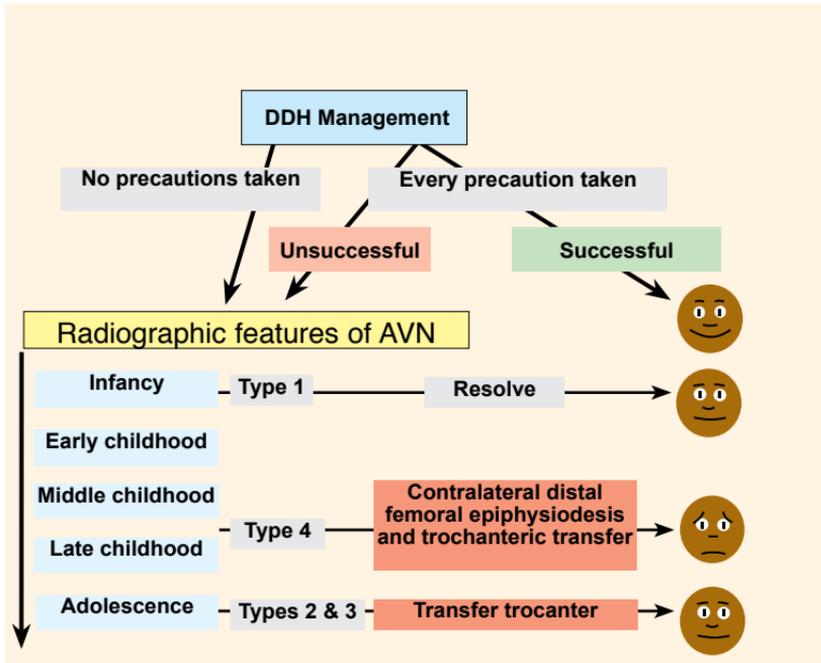
Early signs The early signs of AVN (Salter) are often followed by evidence of a growth disturbance. Type 1 deformity is most common and often mild. Type 2 deformity is more severe, causing a profound shortening of the neck.

Deformity The type and severity of the deformity is related to the location and extent of the physal bridge. Often bridges and deformity are not apparent in early childhood but become obvious toward the end of growth. These bridges cause a tethering of growth and, if eccentric, a tilting of the growth plate [2 opposite page]. Central large bridges cause total arrest with shortening of the femoral neck, relative trochanteric overgrowth, and mild femoral shortening [1].

Management If prevention fails, manage the deformity based on its severity and the type of deformity [2].



1 Type 4 deformity. Note the progressive changes throughout infancy and childhood from a central physal bridge (red arrow) with shortening of the femoral neck and relative trochanteric overgrowth.



1 Management of AVN occurring as a complication of DDH treatment.

Persisting Dysplasia

The third objective in DDH management is the correction of persisting hip dysplasia. Dysplasia should be corrected during growth to prevent osteoarthritis.

Dysplasia may involve the femur, the acetabulum, or both. The most pronounced deformity is in the acetabulum. The most severe dysplasia includes subluxation. Subluxation and dysplasia cause osteoarthritis, which may begin during the teen years. Disability occurs later with simple dysplasia.

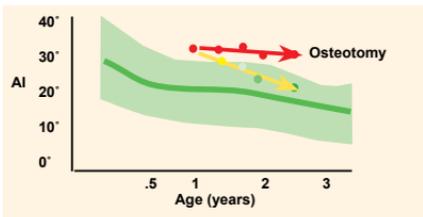
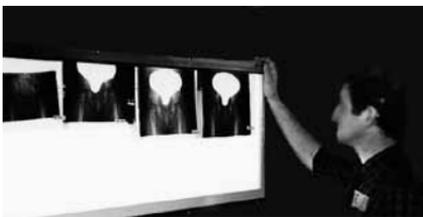
Femoral Dysplasia The proximal femur is anteverted and the head may not be spherical due to the dislocation. The deformity may be due to ischaemic necrosis.

Acetabular dysplasia is the most pronounced deformity and includes shallowness and anterolateral orientation of the socket.

Acetabulofemoral relationship The femoral head is subluxated if not concentric with the acetabulum. The head may also be lateralized following growth with the head subluxated. The acetabulum often becomes saucer shaped, causing instability.

The femoral head may be spherical or aspherical as a result of ischaemic necrosis. The fit with the acetabulum may be congruous or incongruous. Aspherical incongruity is common because over years of growth, the acetabulum assumes a shape to match that of the femoral head.

Timing of correction Correct hip dysplasia as soon as it is evident that



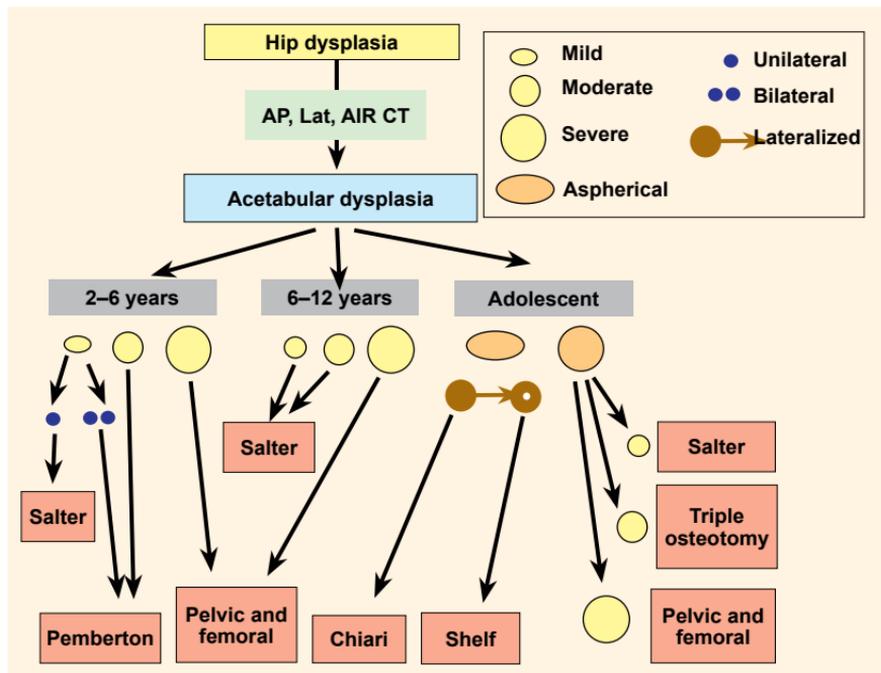
2 Severe acetabular dysplasia. Attempt to correct acetabular dysplasia before it becomes this severe.

1 Effect of growth on acetabular development. Follow acetabular development by placing radiographs in chronologic order and assessing the effect of time. Measure the acetabular index (AI) for each study. Compare this sequence of measurements with the chart of normal AI measurements. If improvement occurs (yellow arrow) and the values become normal, treatment is not required. If AI values remain elevated (red dots and arrow), then pelvic osteotomy will be necessary.

the rate of correction is unsatisfactory, preferably before age 5 years. Establish a time line of a series of AP radiographs [1 opposite page] of the pelvis taken at 4–6 month intervals during infancy and early childhood. Measure the acetabular index, note the smoothness of the acetabular roof (serril), and observe the development of the medial acetabulum (tear drop). Assess by studying the sequence of films. Perform a pelvic osteotomy if the AI remains abnormal and the other features remain dysplastic after 2–3 years of observation. Avoid delaying an obvious need for correction [2 opposite page].

Principles of correction Proper correction of hip dysplasia in DDH follows these certain basic principles:

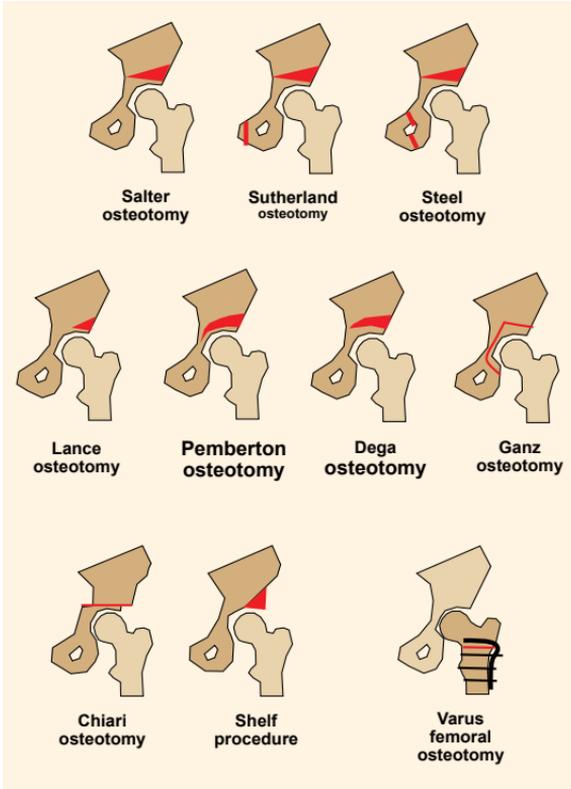
1. Correct the primary or most severe deformity. This is usually the acetabular deformity.
2. Correction should be adequate. If the deformity is severe, combine a pelvic and femoral osteotomy or perform a shelf operation.
3. Avoid creating incongruity. Avoid the Pemberton procedure in the older child. Consider the shelf or Chiari procedure if aspherical congruity is present.
4. Medialize the lateralized hip in the older child with a Chiari osteotomy.
5. Articular cartilage is more durable than fibrocartilage as develops in the shelf and Chiari procedures.



1 Management of acetabular dysplasia. Manage based on age, severity, congruity, and lateralization.

Procedures Select the appropriate procedure based on the site of deformity, age, severity, and congruity [1 previous page]. The choices are numerous [1]. Several procedures are most commonly performed.

- **Femoral osteotomy** Femoral shortening is essential in the older child with unreduced DDH. Remove just enough bone to allow reduction. Reduce the neck–shaft angle by about 20°. Limit rotational correction to about 20°.
- **Salter osteotomy** This is the best choice for correcting mild deformities at any age [2]. The osteotomy will reduce the AI about 10°–15° and the CE angle by 10°.
- **Pemberton osteotomy** This is the best choice for bilateral or moderate to severe dysplasia [1 opposite page] in children under age 6 years.
- **Triple osteotomies** This is the best choice for the adolescent with moderate dysplasia when spherical congruity is present. Procedures are demanding and risk of complications are significant.
- **Chiari osteotomy** This is appropriate when the hip is lateralized and severely dysplastic. It may be used with aspherical congruity. Avoid over-medialization. It covers with fibrocartilage.
- **Shelf procedures** This enlarges the acetabulum with fibrocartilage. It is versatile and the best choice for severe dysplasia without lateralization when aspherical congruity exists. This is the least risky of the major procedures.



1 Options for osteotomies of the hip.



2 Salter osteotomy. This procedure is useful at any age for mild to moderate dysplasia.

Legg-Calvé-Perthes Disease

Legg–Calvé–Perthes (LCP), or simply Perthes, disease is an idiopathic juvenile avascular necrosis of the femoral head. Synonyms include Waldenström's disease and coxa plana. It affects about 1 in 10,000 children. Males are affected four times more often than girls, and it is bilateral in 10–15% of subjects.

Etiology

The cause of LCP disease is unknown. Affected children are small and delayed in maturation, suggesting a constitutional disorder. Vascularity is tenuous in early childhood, and developmental variations in vascular pattern [2] are more common in boys, predisposing some individuals. In addition, trauma, alterations in the coagulability of blood, and endocrine and metabolic disorders may be contributing factors [1 next page]. Possibly several factors combine to cause the disease.

Pathology

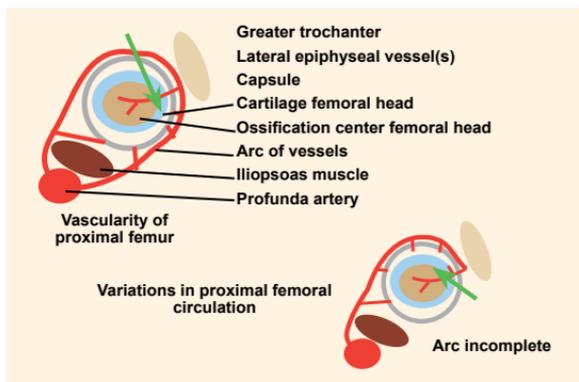
The pathology is consistent with repeated bouts of infarction and subsequent pathologic fractures. Synovitis and effusion, cartilagenous hypertrophy, bony necrosis, and collapse are present. Widening and flattening of the femoral head follow. Most deformity occurs in the "fragmentation phase." If necrosis is extensive and the support of the lateral pillar is lost, the head collapses, mild subluxation occurs, and pressure from the lateral acetabular margin creates a depression, or "furrow" in the femoral head.

Healing requires replacement of dead bone with living bone. Over time in young children, the deformity remodels and the acetabulum becomes congruous. At maturation, the head is reasonably round and the prognosis fair to good. If growth arrest occurs, or the child is older, remodeling is limited. Thus, the capacity of the acetabulum to remodel to congruity is reduced and osteoarthritis is likely in adult life.



1 Pemberton osteotomy.

This infant showed little improvement with time so correction at age 30 months was preformed. Note that the osteotomy extends into but not through the triradiate cartilage.

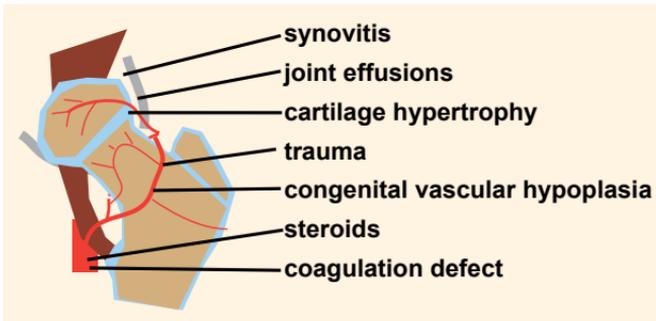


2 Circulation to the proximal femoral epiphysis. This conceptual drawing illustrates the redundant vascular arcade of the proximal femur in the normal child (upper). Congenital or developmental alterations may make the circulation to the femoral head vulnerable to vascular compromise. Note that the circulation is redundant for the proximal femur except for the epiphysis, which is supplied by the lateral retinacular vessels (green arrow).

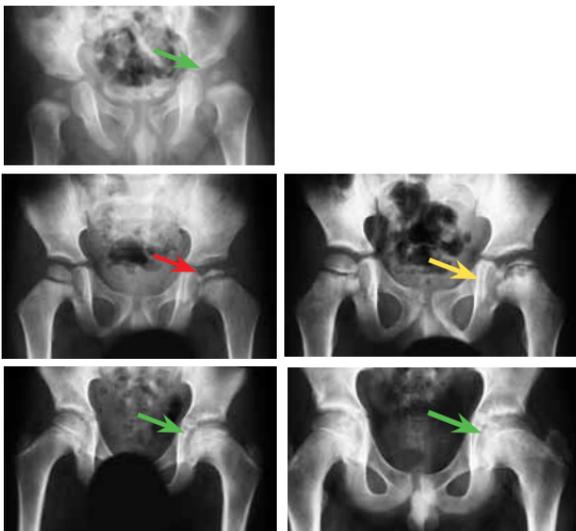
Natural History

The prognosis for LCP disease is fair. The most important prognostic factor is the sphericity of the femoral head at skeletal maturation. This sphericity is related to the age of onset. The younger the age, the more likely the head will be spherical [2 on this page 1 and 2 opposite page]. The longer the period between the completion of healing and skeletal maturity, the longer the period of remodeling. This remodeling cannot occur if a physeal bridge develops [2 and 3 opposite page]. Physeal bridging may occur in young patients and accounts for the occasional poor result seen in these young children. Physeal bridging is most likely in the older child.

Factors affecting prognosis are many, complicating assessment of treatment methods. During late childhood and adolescence, children may experience episodes of pain with vigorous activity. These episodes are transient, often lasting a day or two. More persistent disability may develop during middle to late adult life due to osteoarthritis. The need for joint replacement increases with advancing age and is most likely when the onset of LCP disease occurs after the age of 8 or 9 years [4 opposite page].



1 Causes of LCP disease. These are some of the causes of LCP disease. They may act alone or in combination to cause ischaemic of the femoral head.



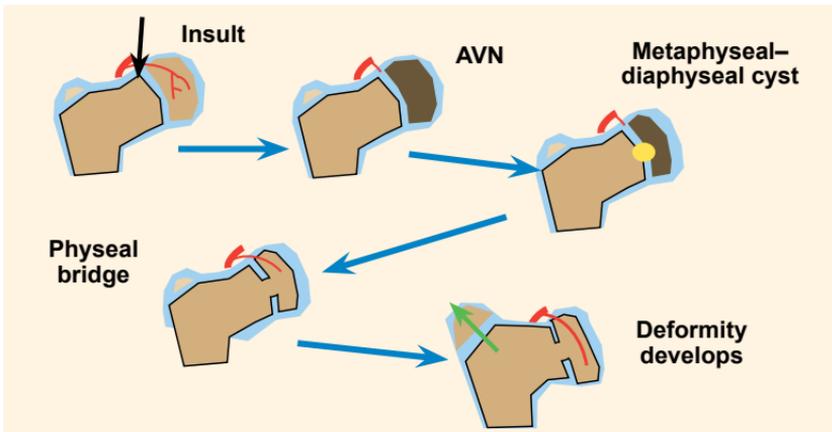
2 Natural history of early onset LCP disease. These radio-graphs were taken at ages 2, 3, 5, 8, and 15 years. Note at age 3 years (red arrow) how total head avascular necrosis is evident. Without treatment, healing (yellow arrow) and remodeling (green arrows) occur.



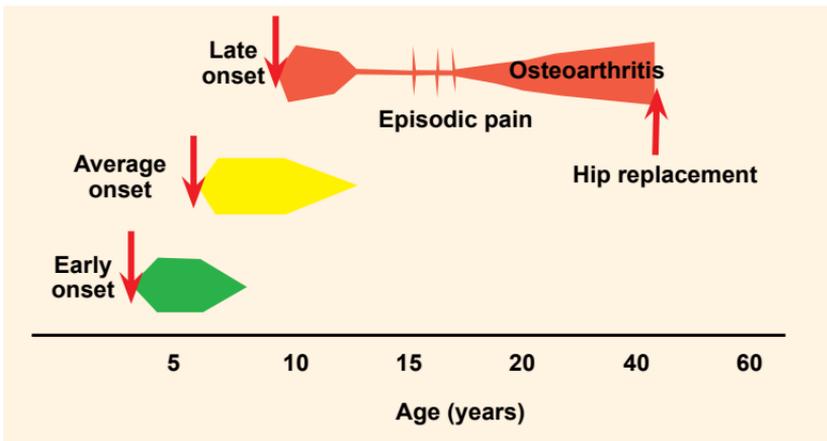
1 Extrusion with residual coxa plana. Note the extrusion in this 7-year-old child. Remodeling improves but does not resolve flattening.



2 Late onset LCP. Note the metaphyseal–epiphyseal cyst at age 11 years (arrow). The head is aspherical and flat at age 18 years (right).



3 Physal bridge formation. Cysts may fill in with bone across the growth plate. This creates a bony bridge that tethers growth and causes progressive deformity. The deformity includes shortness of the femoral neck, relative overgrowth of the greater trochanter, and persistence of flattening of the femoral head.



4 Pain and age of onset of LCP disease. Pain occurs during the acute disease. This lasts for 2–3 years. Pain occurs when deformity persists as episodic incongruity pain during activity and later persistent pain due to osteoarthritis (red).

Diagnosis

LCP occurs between 2 and 18 years of age, but most commonly develops in boys between ages 4 and 8 years. Bilateral involvement occurs with usually more than a year interval between onsets. The disease rarely follows toxic synovitis. An antalgic limp is usually the first sign. Pain may be present but is usually mild. Frequently, the child has recurring pain and a limp for several months before being seen by a physician.

Physical examination The child is comfortable, and the screening examination is normal except for the involved leg. The limp is antalgic, a Trendelenburg sign may be present, and mild atrophy is often present. The most prominent find is stiffness [1]. The loss of hip internal rotation is the earliest sign. The hip rotation test is positive. Abduction is nearly always limited. Flexion is least affected.

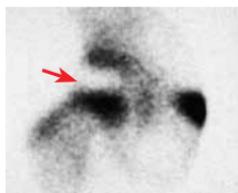
Imaging studies The stage of the disease determines the findings on imaging. Early in the disease, radiographs may be normal, show slight widening of the cartilage space, or often a pathognomonic radiolucent cleft in the femoral head viewed from a lateral position. Radiographic features are largely determined by the stage of the disease at the first visit [2]. Ultrasound will show a joint effusion. The bone scan often shows reduced uptake on the affected side early in the disease [3]. The MRI shows evidence of marrow necrosis, irregularity of the femoral head, and a loss of the signal on the affected side [4]. In the vast majority of cases, only conventional radiographs are necessary to establish the diagnosis and provide management.



1 Limited range of motion. Note the reduction of medial rotation on the affected right side (red arrow) consistent with a positive *hip rotation test*. Examine with one hand spanning the iliac spines as a basis to estimate hip abduction that is reduced in LCP disease (yellow arrow).



2 Radiograph. Various features are shown based on the stage of the disease.



3 Bone scan. Decreased uptake is seen over the region of the femoral head (arrow).



4 MRI. Marrow necrosis corresponding to the extent of the necrosis (arrow) is evident.

Classification

LCP disease is classified by the extent of head involvement and by the stage of the disease.

Extent of involvement Several classification systems are in use for assessing the severity of involvement [1]. Salter–Thompson and Catterall grade the extent of involvement of the epiphysis and Herring grades on the “lateral pillar.” The Salter–Thompson classification is based on showing a cleft in the lateral radiograph [2]. This cleft is a fracture line between the living and dead bone and shows the minimum extent of necrosis. This can be observed early in the disease. The other signs may progress into the fragmentation phase of the disease.

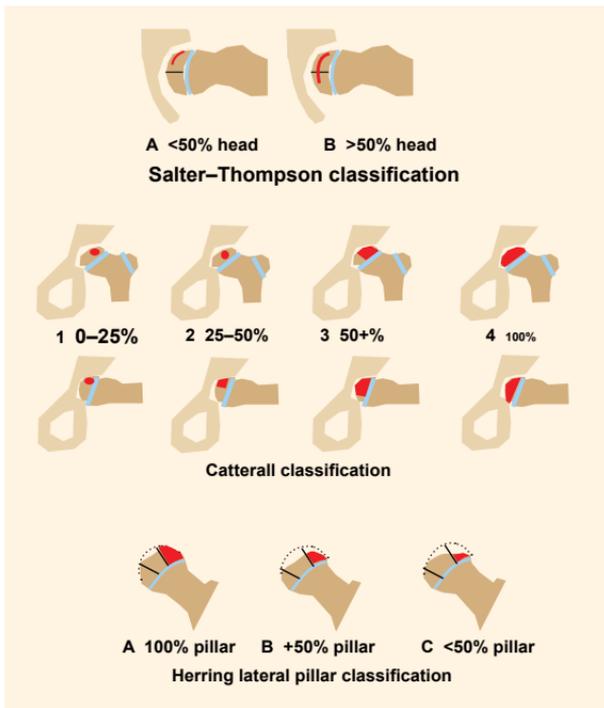
Stage of Disease The disease is divided into four stages: synovitis, necrosis or collapse, fragmentation, and reconstitution [1 next page]. The disease progresses through each stage and part of the healing process. In some classifications, the first stage is omitted.

1. *Synovitis* This stage is of short duration (weeks) and shows the effect of ischemia. Synovitis produces stiffness and pain. Radiographs may show a slight lateralization of the epiphysis (cartilage hyperplasia), bone scans show reduced uptake, and the MRI shows a reduced signal.

2. *Necrosis or collapse* The necrotic portions of the femoral head undergo collapse, and radiographs show a reduction in size and an increased density of the head. This stage lasts 6–12 months.

3. *Fragmentation* In this healing stage, avascular bone is resorbed, producing the patchy deossification seen on conventional radiographs. Deformation of the femoral head often occurs during this stage. This stage persists for 1–2 years.

4. *Reconstitution* New bone is formed. Overgrowth often produces coxa magna and a widening of the neck.



1 Classifications of LCP disease severity.



2 Salter–Thompson classification. Note the extent of the cleft (red arrow), which shows the area of necrosis (yellow arrow), as evident in the radiograph taken 1 year later.

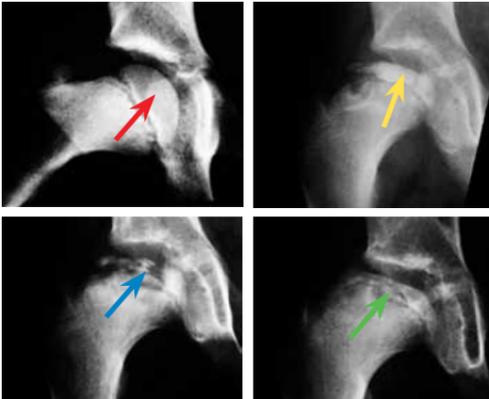
Head-at-risk signs [2] include extrusion or ossification lateral to the femoral head, metaphyseal changes of rarification or cyst formation, and a radiolucency on the lateral aspect of the physis (Gage’s sign).

Differential Diagnosis

Disorders that cause clinical and radiographic changes such as LCP disease are numerous [1 opposite page]. Although these other causes are relatively rare, they should be at least considered before establishing the diagnosis. The most likely diagnoses to miss are hypothyroidism and epiphyseal dysplasia. Dysplasias usually affect both hips with symmetrical degrees of involvement [3]. Bilateral symmetrical involvement is very rare in LCP disease.



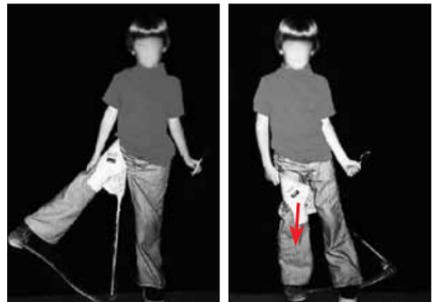
1 Stages of LCP disease. The disease progresses through the stage of synovitis (red), necrosis (yellow), fragmentation (blue), and reconstitution (green).



2 Head-at-risk signs. Extrusion (red arrow), metaphyseal reaction (yellow arrow), and lateral rarification or Gage sign (white arrow).



3 Symmetrical involvement suggests another diagnosis. This child has epiphyseal dysplasia.



4 Adaptive boy. The boy circumvented treatment by dropping the brace (arrow).

Management

The objective of management of LCP is to preserve the sphericity of the femoral head to reduce the risk of stiffness and degenerative arthritis while preserving the emotional well-being of the child.

The management of LCP is very controversial. In the past, treatment regimens have varied from operating on every case to no treatment at all. Children have been subjected to years of hospitalization in recumbency and various types of ineffective bracing [4 opposite page] and operative treatments. Remember our humbling history of management in managing the next patient.

Management Principles The following is a list of currently accepted principles of managing LCP:

1. Avoid treatment of patients who will do well without treatment. The young child and children of any age with minimal involvement do not require treatment.

2. Consider the psychosocial situation. The emotionally dysfunctional child should not be subjected to orthotic management. Due to the long duration of the disease, treatment often imposes severe emotional stress for the child. Be sensitive to the child's overall well-being.

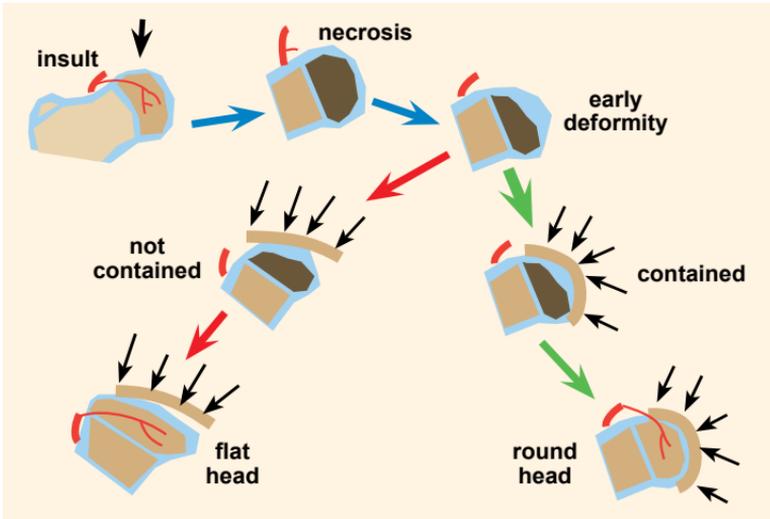
3. Provide "containment" to maintain or improve the sphericity of the femoral head [1 next page]. The acetabulum is used as a mold to contain the plastic femoral head. This requires positioning the hip in abduction in a brace or a surgical procedure that increases acetabular coverage of the femoral head.

Category	Disease	Comment
Syndromes	Gaucher's disease Mucopolysaccharidosis Multiple epiphyseal dysplasia Spondyloepiphyseal dysplasia	Often produce bilateral AVN, which is symmetrical in severity and stage
Hematologic	Sickle cell disease Hemophilia Lupus erythematosus	May be related to steroid treatment
Infection	Septic arthritis Femoral osteomyelitis	Complication in delayed drainage of an infected hip
Metabolic	Hypothyroidism	
Trauma	Femoral neck fractures Hip dislocation Slipped epiphysis	Common causes of AVN
Inflammation	Toxic synovitis	Occurs in 1–3% of cases
Tumors	Lymphoma	

1 Differential diagnosis of LCP. Several disorders may be confused with LCP. Often the primary disease makes the cause of the avascular necrosis clear.

4. Attempt to maintain or gain a satisfactory range of motion (Fig. 7.96). Motion is nearly always reduced. The degree of stiffness is related to the severity of the disease and the activity level of the child. Gaining motion by curtailing activity has its limits. What constitutes a satisfactory range of motion is seldom defined. A minimum is about 20° of abduction.

5. Control the cost of management. Inpatient traction, MRI studies, arthrography, and operative procedures are most expensive. Conventional radiographs, rest at home, and the selective use of imaging and procedures provides optimum care at least cost.



1 Concept of containment treatment. Containment is provided by positioning the leg or the acetabulum to encompass the femoral head. Without containment (red arrows), the head becomes flattened. The contained head (green arrows) becomes round. Both heads become revascularized.



2 LCP disease in a young child. This 3-year-old girl was managed without treatment. The outcome was good.

Management Algorithm

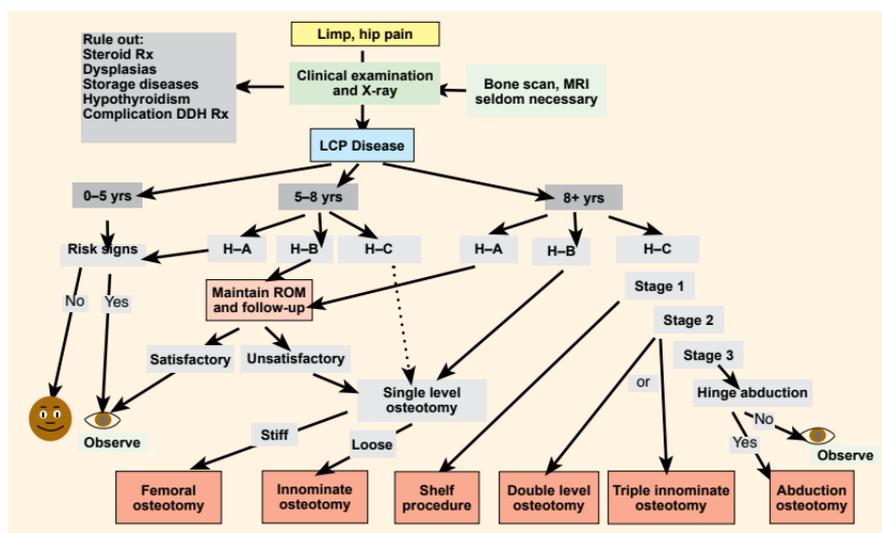
The flowchart below [2] is an approach to management. The management of LCP disease is one of the most controversial in orthopedics. This is one of many approaches. Consider each of the following variables in planning management.

Severity The flowchart [2] is based on the Herring A, B, and C categories. Be aware that the Salter–Thompson cleft sign when visible will predict severity earlier than either the Herring or Catterall methods. The Herring method does not become fully clear until the stage of late necrosis or early fragmentation.

Operative choices These choices [2] demonstrate the many options in management. The choices do not include brace management. This option is still viable but poorly accepted by most children and families due to the long duration required.



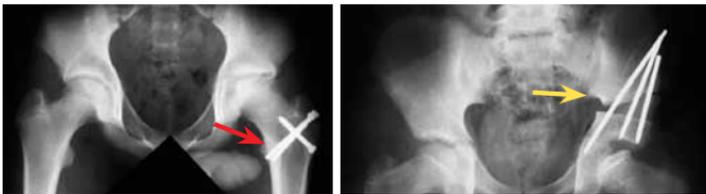
1 Extremes in management. The pattern bottom brace (red arrow) has been shown to increase hip loading and provides no containment. Wide abduction casts (yellow arrow) are effective but very difficult for the child.



2 Management of LCP disease. This flowchart considers age, severity, ROM, and stage of disease in determining appropriate management.

Age is the most important variable and the first consideration. Prognosis is most dependent on the age of onset. Divide ages into the young child (0–5 years), the middle age group (5–8 years), and the older age group (8+ years). This older group has a much poorer prognosis.

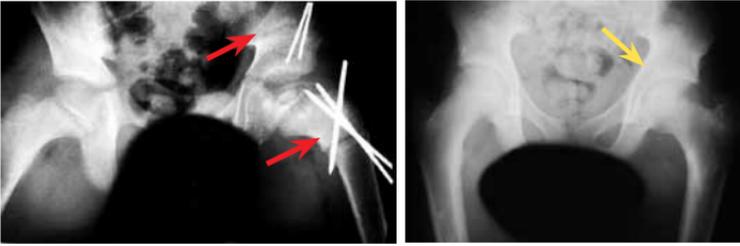
- Early childhood** The prognosis is usually excellent [2 on page 36] unless a physeal bridge develops. The development of bridging is not preventable. In this age group, treatment is not necessary or helpful. Asking the parents to limit the child’s activity is asking the parent to do the near impossible. It is not fair or helpful. Simply ask the parents to redirect the child’s activity when feasible into some activity that is less physical. If metaphyseal cysts develop, follow the child with an AP radiography every 2 years to assess growth, as physeal bridging may occur. If this complication develops, it may be necessary to transfer the trochanter in late childhood or adolescence.
- Middle childhood** Avoid treatments that are either ineffective or present special hardships for the child [1 previous page]. Manage those with H–A and H–B (H = Herring) without containment. Encourage abduction exercises. Provide follow-up. Consider treating H–C by containment [1]. Such treatment is controversial.
- Late childhood** Consider operative containment in H–B and H–C hips if seen during stage 1 or 2. An option is abduction casts or braces, but most children find this treatment very difficult (Fig. 7.98 right). In stage 1, the shelf procedure [2] is effective and least invasive. In stage 2, the double-level osteotomy [1 opposite page] is often necessary. The head is still plastic and will remodel when well contained. In stage 3, the deformity is permanent. If hinge abduction is present, an abduction osteotomy may improve motion and reduce discomfort [2 opposite page]. If motion is satisfactory and not painful, accept the deformity [3 opposite page].



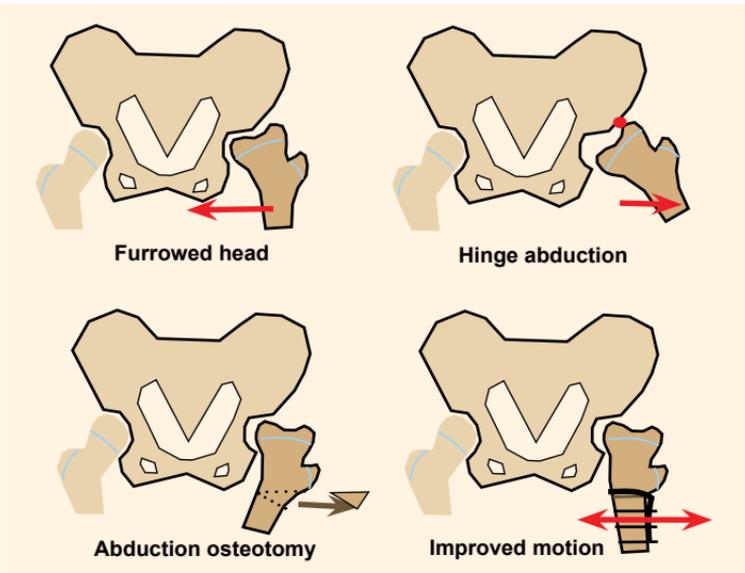
1 Moderate containment. Containment is provided by a varus osteotomy (red arrow) or a single innominate osteotomy (yellow arrow).



2 Shelf containment in LCP disease. This 10-year-old girl is H–C and in stage 1. A shelf procedure was performed. Radiographs are shown at age 10 years (red arrow), 11 years (yellow arrow), and 12 years (orange arrow).



1 Double-level osteotomy for LCD disease. Containment in this 10-year-old child with H–C was provided by an innominate and femoral osteotomy (red arrows). A round head is shown at age 15 years (yellow arrow).



2 Hinge abduction. With flattening or furrowing of the femoral head, abduction is full but adduction causes the lateral margin of the femoral head to hinge on the pelvis, in turn causing widening of the medial joint space and pain. Pain and motion may be improved by an abduction osteotomy.



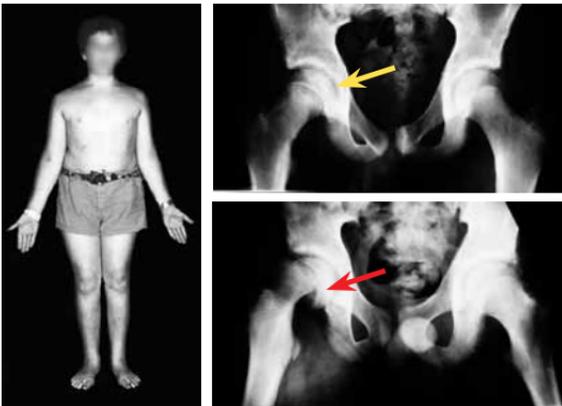
3 Severe deformity. Severe deformity on AP radiograph (red arrow) and by arthrogram (yellow arrow) in an 11-year-old boy in stage 3. The deformity was established and no treatment was recommended.

Slipped Capital Femoral Epiphysis

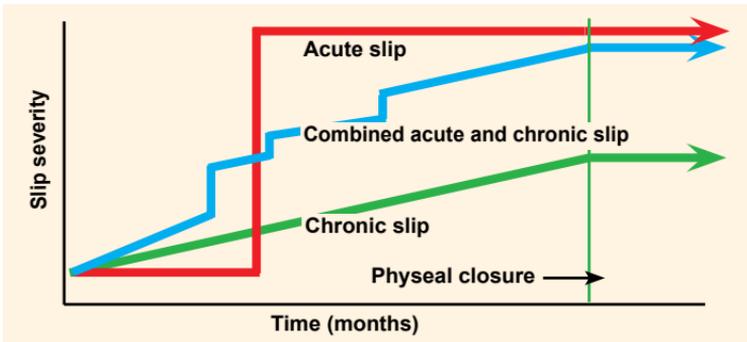
Slipped capital femoral epiphysis (SCFE) is a displacement of the upper femoral epiphysis on the metaphysis [1]. SCFE is the most common adolescent hip disorder. It occurs in about 1 in 50,000, most commonly in obese boys. The peak age for boys is 13 years and for girls 11 years, with a range from middle childhood to maturity. SCFE is bilateral in about one-fourth of cases, with possibly slight silent slippage in even more.

Etiology

The cause of SCFE is complex. In early adolescence, the growth plate is relatively weaker, as evident from the incidence of physeal injuries at other sites at this age. The hip is vulnerable, as it carries about four times its body weight. Retroversion or a reduced neck shaft angle may increase the verticality of the plate, making it mechanically less stable. The risk is further increased by any constitutional disorder that adds to this physeal weakness. Endocrine disorders such as hypothyroidism, hypopituitarism, or hypogonadism and metabolic disorders such as rickets or treatment with radiation or chemotherapy may contribute. If obesity [1 left] or trauma are added to this, the plate may fail, either gradually, acutely, or as a combination of gradual and acute components.



1 Slipped capital femoral epiphysis. Typical habitus (left). Upper radiograph in orthopedist's office where diagnosis of SCFE was made. Acute slip (red arrow) occurred in the parking lot on the way to the hospital.



2 Patterns of slippage. Slipping occurs over a period of months. At physeal closure, progression ceases.

Natural History

Failure of physis and slipping may occur from age 6 years until the plate is fused. Most slips are gradual over a period of many months [2 opposite page]. Often the progress on the slip is variable; acute episodes are superimposed on gradual slipping. Closure of the plate as the result of treatment or as it occurs naturally at the end of growth halts the process. Following slipping, remodeling may reduce the deformity. The risk of osteoarthritis is increased when the slip is more severe, the child is older, and especially if avascular necrosis or chondrolysis complicate management.

Patients with SCFE have a normal acetabulum and the articular cartilage is often preserved. Thus, despite the presence of significant deformity, many do well for many decades. Chondrolysis and avascular necrosis cause early degeneration.

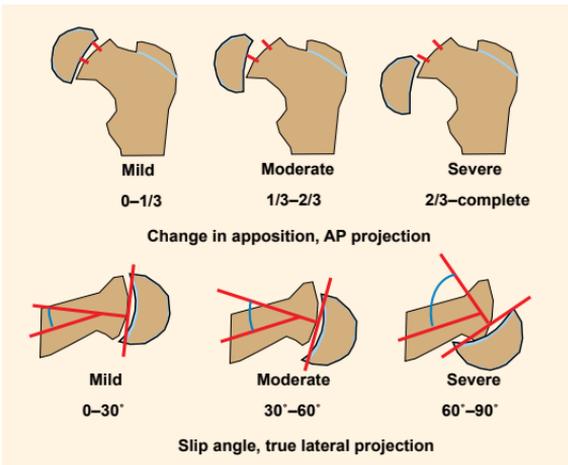
An enigma is the significance of the so-called pistol grip deformity. This deformity is often seen in males who develop osteoarthritis. Speculation suggests that this deformity is secondary to unappreciated SCFE. Why such mild deformity should cause early degeneration is unclear.



1 Classic AP radiographic features. Note that the head is displaced inferior to a line (red) drawn along the superior margin of the neck. Metaphyseal rarification (yellow arrow) and slight widening of the growth plate (orange arrow) are seen.



2 Very early slip. Note that on the AP view, no change is seen in the head-neck relationship. The slight slip is clearly evident on the lateral view.



3 Grading severity of SCFE. Severity can be expressed as a grade based on the displacement seen in the AP projection. A more accurate measurement is the slip angle measured from a true lateral radiograph.

Diagnosis

The diagnosis of SCFE is made more difficult because the onset of the common chronic slip is insidious and the pain is often referred to the knee. Knee pain occurring between the ages of 6 years and maturity should promote an evaluation of the hip. Long-standing slips will produce an out-toeing gait, an abductor lurch, and limb atrophy.

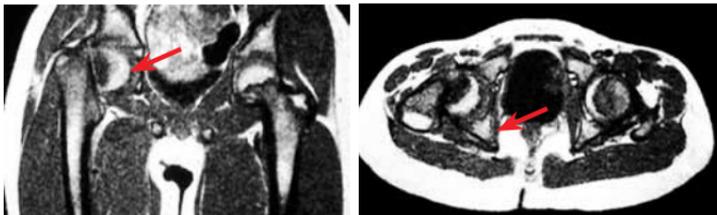
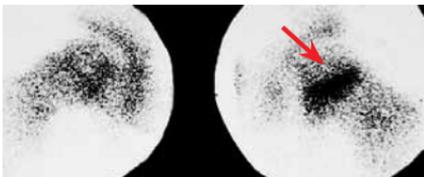
Screening is done with a hip rotation test. The loss of medial hip rotation is due to inflammation of the joint and to the posterior inferior slippage of the femoral head, causing a deformity similar to femoral retroversion. A positive finding requires further evaluation with a “frog-leg” lateral radiograph of the pelvis.

Radiography The diagnosis of SCFE can nearly always be made on conventional radiographs of the pelvis. The frog-leg lateral best shows the posterior slippage of the epiphysis. The AP radiograph usually shows widening of the growth plate and rarefaction of the adjacent metaphysis [1 previous page]. Sometimes these are the only findings, and the condition is called a “preslip.” Subtle displacement is identified by a loss of the normal relationship at the epiphysis–neck interphase. On the AP radiograph, the head lies above and lateral to a line drawn along the superior margin of the neck. On the lateral radiograph, any slipping will disrupt this alignment [2 previous page]. On the AP radiograph, assess severity by the percentage of contact between the head and neck. For a more accurate assessment, obtain a true lateral view and measure the slip angle [3 previous page]

Other imaging Pinhole (high-resolution) lateral bone scans of both femoral heads will show increased uptake in preslips [1]. Ultrasound imaging will demonstrate the “step off” at the site of displacement. The MRI shows AVN or altered head position [2].



1 Bone scan in preslip. The diagnosis of a preslip can be confirmed by a high-resolution bone scan. Increased uptake of the physis (red arrow) is noted as compared to the opposite uninvolved side.



2 MRI view of SCFE. This study shows a severe slip (red arrows).

Management

The objective of management is to stabilize the growth plate to prevent further slippage and to avoid complications [1 on page 283]. Achieve this by a screw, pins, epiphysiodesis, or immobilization with a spica cast.

Stable SCFE Fix mild and moderate stable slips *in situ* with a single screw. This prevents further slippage and leads to fusion of the growth plate [1]. In the child under age 8 years, fix with smooth pins to allow growth.

For severe slips, the choice is *in situ* fixation or osteotomy. *In situ* fixation is sometimes difficult, and it is necessary to place the entry point of the screw far forward on the neck of the femur. If motion is unsatisfactory, an osteotomy can be performed later. The other choice is an osteotomy to correct deformity and stabilize the slip.

Osteotomy The procedure may be performed at the neck, the base of the neck, intertrochanteric, or subtrochanteric location.

- **Cervical osteotomy** Osteotomies of the neck include shortening, are correct at the site of deformity, but carry a significant risk of AVN. Unless the surgeon has considerable experience with the technique it is wisely avoided.
- **Base of the neck osteotomy** This provides safety with good correction. Through an anterolateral approach, the capsule is opened. An anterolaterally based wedge of bone is removed at the base of the neck. Any prominences are shaved off. Fixation is simply achieved with screws or Steinmann pins.



1 Pinning stable SCFE *in situ*. This mild slip was pinned *in situ* with a single pin (red arrow).

- **Intertrochanteric osteotomy** This is safe, and involves fixing with a nail plate.
- **Subtrochanteric osteotomy** Far from the deformity, this fixation is more difficult.

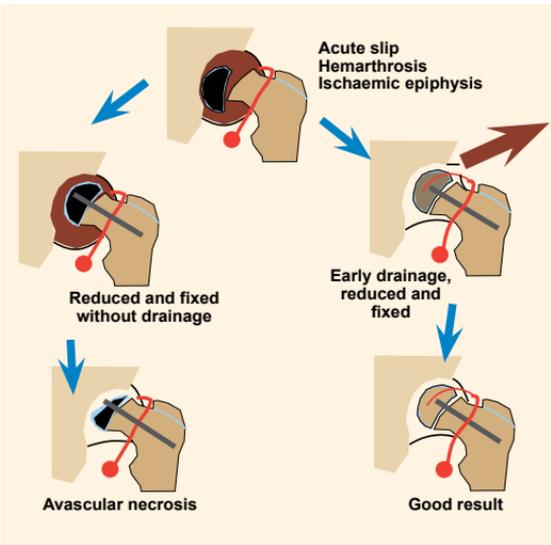
The base of the neck or the intertrochanteric level osteotomy levels are preferred because correction is good and risks are fewer.

Osteoplasty Residual prominence of the anterior portion of the femoral neck is a common cause of loss of hip flexion. Removal is simple and safe.

Prophylactic pinning Bilateral slips occur in about one-fourth of patients. Always carefully evaluate the apparently uninvolved side. Pin the other side if you are suspicious that an early slip is present or if some underlying metabolic disorder such as renal osteodystrophy is present.

Unstable Slips Acute slips (5–10% of all slips) cause instability and increase the risks of avascular necrosis (AVN). They occur suddenly, causing an inability to walk. As the slip is unstable, any movement of the leg causes pain.

Unstable slips are often more severe than gradual slips. Management is difficult and controversial, and the outcome is sometimes poor. Acute management choices include traction, manipulation, cast immobilization, acute decompression, reduction, and fixation. Mounting evidence suggests that early decompression and fixation reduces the risk of AVN [1].



1 Prevention of AVN by early drainage. This concept of Parsch and Swinkowski recommends early emergency decompression, reduction, and fixation to prevent AVN in acute SCFE.

Admit the patient. Arrange for pin fixation. If the procedure is delayed, consider applying skin traction with the limb supported on a pillow. Reduction may occur from traction or when the limb is in position in the operating room for fixation. Fix as with a stable slip. Supplement the fixation with a second screw if the first pin is not optimal or if the patient is obese or even more unreliable about self-care than most adolescents. Encourage bedrest for 3 weeks and then non-weight-bearing activity until early callus is seen. Follow-up to observe for AVN.

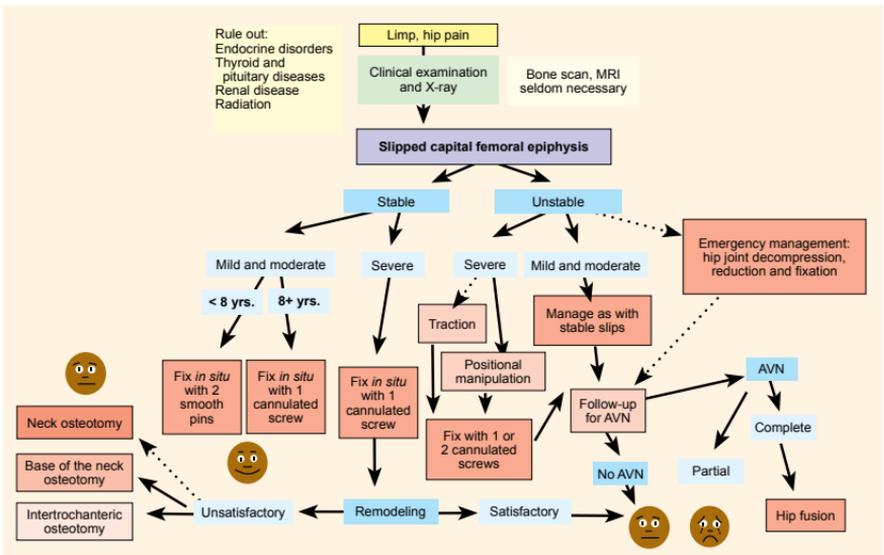
Complications

Complications are common in SCFE. Some can be avoided.

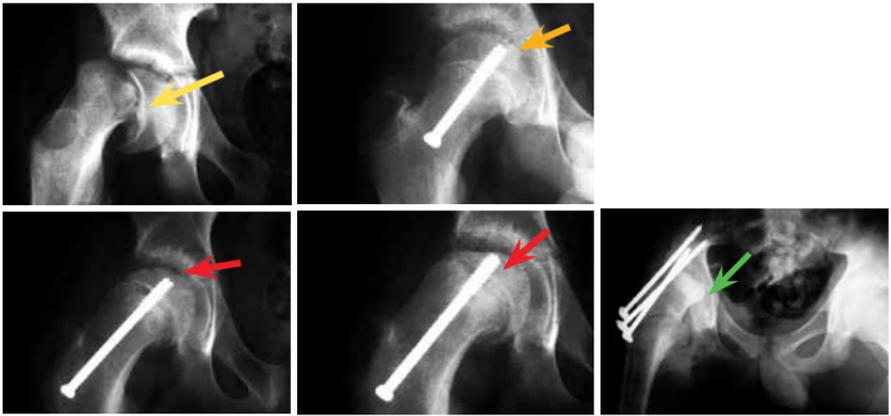
Avascular necrosis AVN is a serious complication that often follows management of unstable slips [1 next page]. Do everything possible to prevent this disastrous outcome. Avoid manipulative reductions. In unstable slips, consider emergency drainage and pin in the position that results from traction or operative positioning. Follow the patient’s progress clinically [3 next page]. Be suspicious if hip rotation becomes guarded or progressively more restricted. Necrosis is usually clear radiographically in 6–12 months or earlier on MRI studies.

If AVN occurs, remove or exchange protruding pin(s), prescribe crutch walking, and encourage motion with activities such as swimming. If the AVN is only partial, attempt to salvage function. Procedures such as drilling or coring are not effective. Fuse the hip if pain and disability are unacceptable. Avoid fusion techniques (such as cobra plates) that jeopardize the outcome of conversion to a prosthetic joint later in life.

Chondrolysis may occur with or without treatment [2 next page]. Joint penetration by guide pins or screws is a doubtful cause. The joint space narrows, hip motion decreases, and an abduction contracture often develops [1 on page 285]. Relieve weight bearing and encourage motion. The value of aspirin, hospital traction, and capsulotomy is uncertain. Most improve with time. Rarely the disease progresses to joint destruction and arthrodesis. The combination of chondrolysis and AVN is devastating and usually ends in hip fusion.



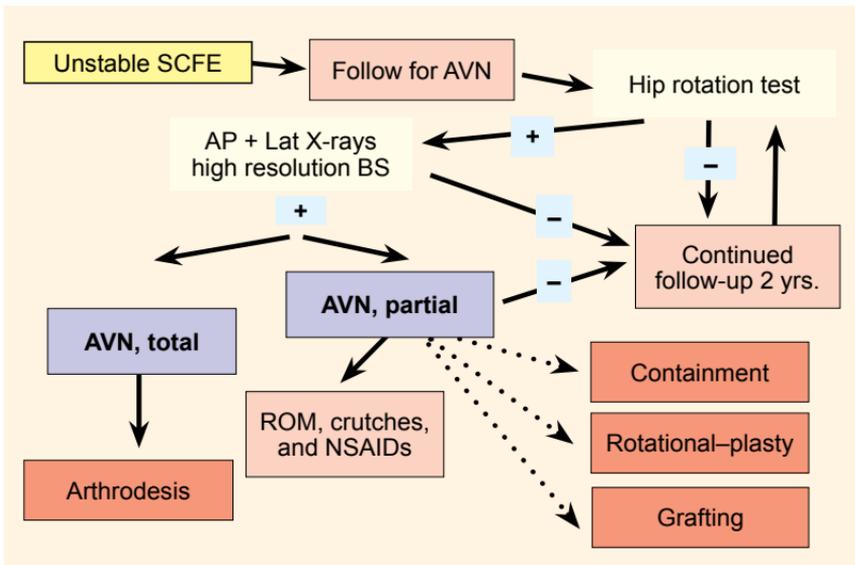
1 Management flowchart for SCFE.



1 Unstable slip with avascular necrosis. This acute slip (yellow arrow) was reduced and pinned (orange arrow) without drainage. The hip underwent AVN (red arrows) and eventually required a hip fusion (green arrow).



Fig. 7.117 Chondrolysis. Note the narrowing of the cartilage space as demonstrated by this arthrogram.



3 Management of AVN in SCFE.

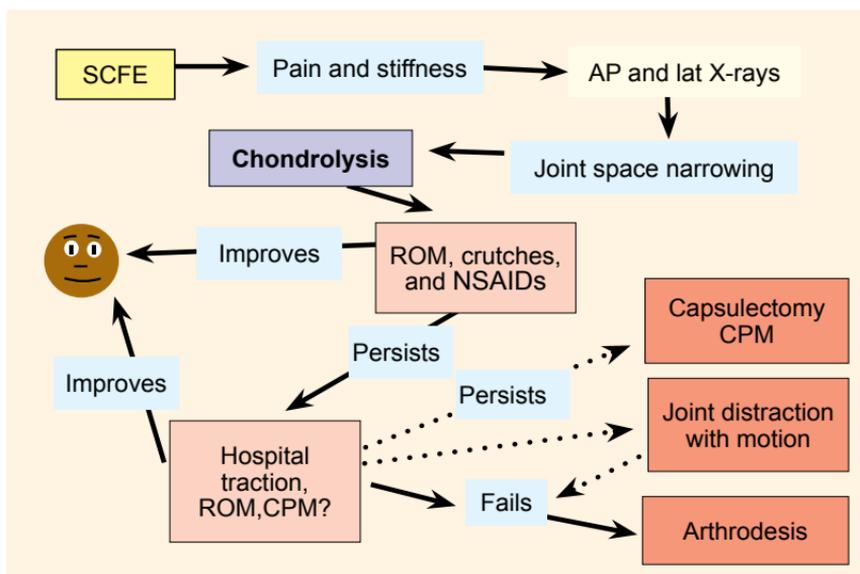
Uncommon Hip and Femoral Disorders

Coxa Vara

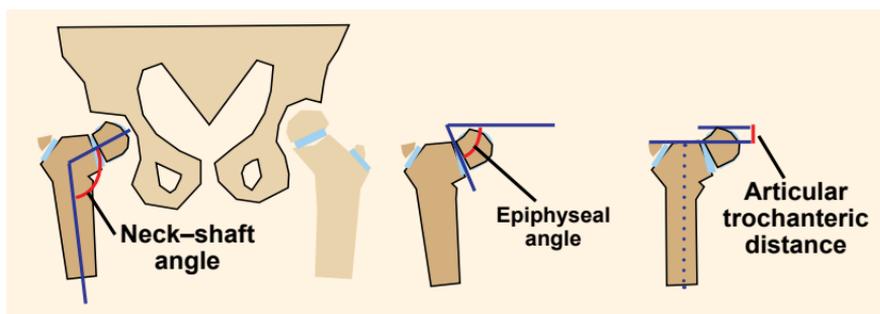
Coxa vara (CV) describes a deformity in which the neck–shaft angle is reduced below 110° [2]. CV has many causes [1 next page] and can be a primary isolated deformity or associated with other disorders.

Measurements Measurements of the shape of the upper femur in addition to the neck–shaft angle include the epiphyseal angle and the articular trochanteric distance (ATD). The ATD is an important measurement in assessing abductor muscle deficiencies. Normally the center of the femoral head lies at the level of the tip of the trochanter. The ATD is positive. A reduction in the ATD may be due to coxa vara or relative trochanteric overgrowth from loss of growth of the capital femoral epiphysis.

Disability Coxa vara causes limb shortening and abductor muscle weakness. As the deformity increases the epiphyseal angle, the deformity is sometimes progressive.



1 Management of chondrolysis in SCFE.



2 Measurements of the proximal femur. These measurements are often used in discussion of changes in the neck–shaft angle.

Congenital coxa vara is often associated with a short femur [2] or it can be associated with a skeletal dysplasia such as spondyloepiphyseal dysplasia.

Developmental coxa vara may develop over time [1 on page 288].

Secondary coxa vara may be associated with other problems, as is common in fibrous dysplasia, or it may be iatrogenic, as may occur after a varus osteotomy treatment of LCP disease.

Management is based on the category of deformity.

- *Progressive coxa vara* may be congenital or developmental. Perform a valgus osteotomy [1 opposite page] that reduces the epiphyseal angle to less than about 40°. This more horizontal position provides stability and reduces the risk of recurrence.

Primary coxa vara

Congenital:

- Isolated defect
- Bone dysplasias
- Developmental

Secondary coxa vara

- Trauma, malunion
- Avascular necrosis, DDH management
- Tumors
 - Fibrous dysplasia
 - Bone cysts
- Osteopenic
- iatrogenic, post-varus osteotomy

Functional coxa vara, coxa breva

- LCP disease
- Total physeal arrest, DDH treatment

1 Causes of coxa vara.



2 Congenital coxa vara. This deformity is often associated with congenital short femur (red arrow) or may occur as a feature of a bone dysplasia (yellow arrows).

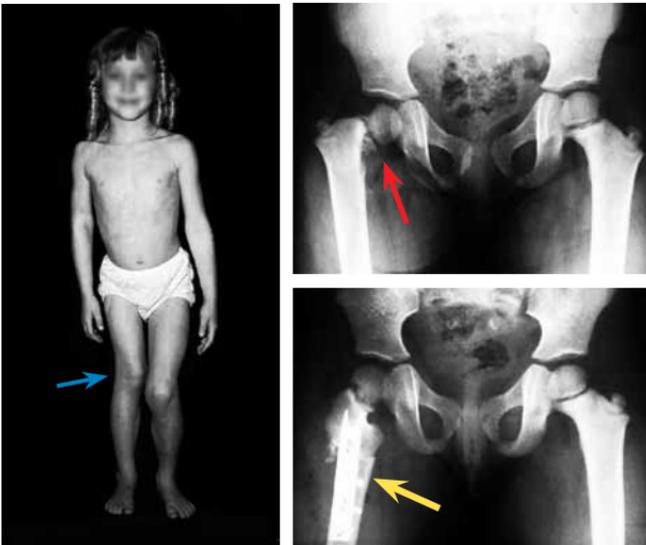
- *Varus osteotomy* Residual deformity is most common when the procedure is performed late in childhood when the time for remodeling is limited [2 next page]. The effect of this osteotomy is sometimes minimized by a concurrent greater trochanteric arrest.
- *Fibrous dysplasia* The deformity is often progressive [3 next page], producing a shephard's crook deformity. Manage early with a valgus osteotomy, stabilized by permanent intramedullary fixation.

Coxa Valga

An increased neck shaft angle is seldom a problem. Coxa valga is seen in cerebral palsy and other neuromuscular disorders. This deformity may be confused with antetorsion because both can increase the *apparent* neck shaft angle [1 on page 289]. The *true* neck shaft angle can be assessed by rotating the hip under image intensification until the proximal femur is in profile.

Idiopathic Chondrolysis

This idiopathic disorder is characterized by chondrolysis with a spontaneous onset resulting in pain, stiffness, and narrowing of the joint space [2 on page 289]. Its natural history is variable. Often restoration of the joint space occurs over several years. In others, the hip becomes ankylosed and fusion is required.



1 Valgus osteotomy. Note the shortening of the right leg (blue arrow). The varus deformity (red arrow) was corrected with a valgus osteotomy (yellow arrow).

Manage by weight release with crutches while encouraging active motion through activities such as swimming.

Protrusio Acetabula

Protrusio is rare in children. It occurs in Marfan syndrome, seronegative spondyloarthropathy, and conditions that weaken bone. Pain, stiffness, medial displacement of the acetabulum [3 opposite page] and an increased CE angle are typical features. Manage the underlying disease. Consider early fusion of the triradiate cartilage in Marfan syndrome or in severe deformity. An osteotomy of the pelvis to shift the loading more laterally (reverse triple innominate osteotomy) may be required.

Snapping Hip

Snapping hip may occur in the adolescent. The iliopsoas tendon subluxates, creating a snap and causing pain. The subluxation can be demonstrated by ultrasound. Manage with rest, injection, or rarely with tendon lengthening.

Labral Tears

Labral tears can occur in adolescents spontaneously following trauma or associated with acetabular dysplasia. The diagnosis can be made by MRI, arthroscopy, or arthrography. Management is difficult. Limited debridement may provide relief.



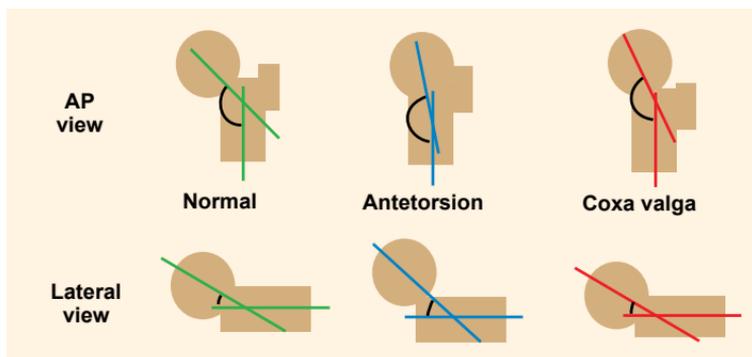
1 Developmental coxa vara. Radiographs show progressively increasing epiphyseal angle (yellow angles) at 3 months (top), 12 months (middle), and 3 years (bottom).



2 Postoperative coxa vara. This varus deformity followed a varus osteotomy for treatment of LCP disease.



3 Secondary coxa vara. This varus deformity is secondary to fibrous dysplasia..



1 Effect of rotation on neck–shaft angle. Note that the neck–shaft angles (black arcs) on the AP projections are increased both by antetorsion (blue lines) and coxa valga (red lines).



2 Idiopathic chondrolysis. Note the narrowing of the joint space



3 Acetabular protrusion. Note the deepening of the acetabulum.

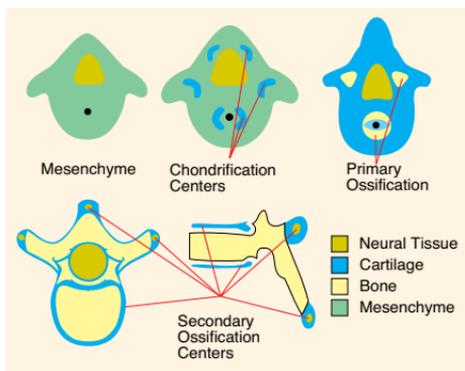
Chapter 8 – Spine and Pelvis

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Spine problems in children have the potential to cause considerable disability and must be taken seriously. Whereas the majority of adults have back pain at times, back pain in children is less common and often due to some specific organic disease that requires treatment. Deformity is of greater concern in the child because of the potential for progression with growth. Conversely, minor truncal asymmetry is common in children and may cause undue concern leading to unnecessary apprehension and treatment.

Normal Development

The vertebral bodies form in the usual sequence from mesenchyme to bone [1]. In the frontal projection, the spine is relatively straight throughout growth. In the lateral projection, the spine evolves from a single curve at birth to a triple curve pattern in the child [1 next page]. Although this triple curve pattern is necessary to assume an upright posture, the obliquity imposes an added load on the lumbar spine. This load contributes to development of spondylolysis in the child, intervertebral disc herniation in the adolescent, and degenerative arthritis in the adult.



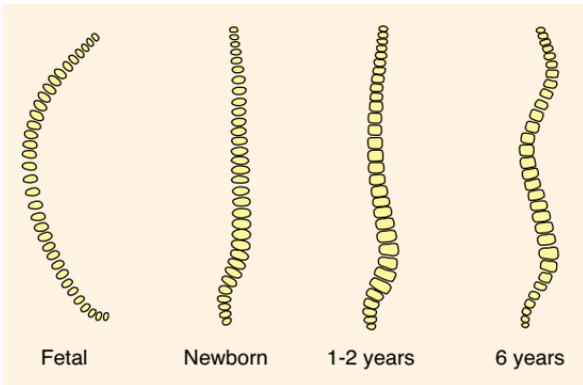
1 Vertebral development.

Vertebrae develop first as mesenchyme, then cartilage, and finally bone. Secondary ossification centers develop during childhood and fuse during adolescence or early adult life. From Moore (1988).

Terminology and Normal Variability

Sagittal plane The normal range for dorsal kyphosis falls between about 20° and 45° . Kyphosis between 45° and 55° is marginal. Kyphosis below 20° is referred to as *hypokyphosis*, and above 55° as *hyperkyphosis*. Hyperkyphosis is sometimes referred to as a “round back” deformity. Normal levels for lumbar lordosis fall between 20° and 55° . Likewise, reduced lordosis is termed *hypolordosis*, and increased lordosis, *hyperlordosis*. Hypolordosis is called a “flat back” and hyperlordosis either a “lordotic deformity” or a “swayback.”

Frontal plane Mild curves that cause truncal asymmetry are usually normal variants [2]. These variations are $<10^\circ$ by Cobb and $<5^\circ$ by scoliometer measure. These asymmetries have not been shown to cause any disability in childhood or adult life.



1 Normal spine development (sagittal plane). The spine changes from a single curve at birth to a triple curve during childhood.



2 Mild truncal asymmetry. These mild asymmetries are variations of normal, do not require treatment, and cause no disability.

Evaluation

The spine is evaluated as part of a screening examination or to assess pain or deformity. The screening examination was detailed in Chapter 2.

History and Physical Examination

Screening examination Is there some underlying disorder? Marfan syndrome, neurofibromatosis, osteochondrodystrophies, or mucopolysaccharidoses are readily obvious in the older child but may not be so apparent in the infant.

History Inquire about the onset, progression, disability, and duration. The family history is of great importance, because scoliosis and hyperkyphosis are often familial. Back pain is also familial.

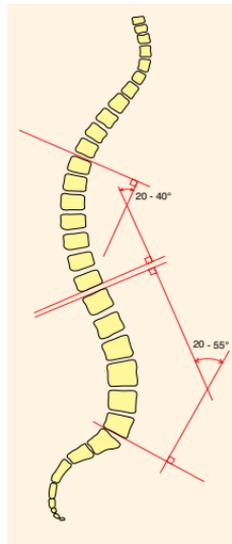
Posture Note asymmetry of shoulder height, scapular prominence, flank crease, or asymmetry of the pelvis. Note any skin lesions, especially those in the midline. The presence of midline skin lesions such as dimples, hemangioma or hair patches, cavus feet, or leg atrophy are often associated with underlying spinal lesions. Café au lait spots are associated with neurofibromatosis, a cause of scoliosis.

Be aware that minor truncal asymmetries occur in about 10% of children. These are benign, cause no disability, and require no treatment. Avoid calling attention to such normal asymmetries because it only worries the patient and family.

Forward bending Perform the forward bending test [1]. This is best done with the examiner seated in front of the child. Control her forward bend by holding her hands together. Slowly guide her forward bending while observing the symmetry of each level of her spine. Any significant scoliosis will be readily apparent. Assess asymmetry with a scoliometer [2 next page], which measures inclination. Minor degrees of asymmetry are usually only a variation of normal, but require follow-up examination. If any abnormalities are found, a detailed physical and screening neurological examination is essential to avoid diagnostic errors. Hesitation, a list to one side, or restricted motion is abnormal. Lesions such as spinal cord tumors, spondylolisthesis, disc herniations, or discitis limit the mobility or symmetry on forward bending.



1 Limited forward bending. Limited forward bending (red arrow) is seen in a variety of diseases. It is an important sign that suggests the need for additional studies.



2 Method of measuring spinal alignment. Select the endplate of the upper and lower vertebrae with greatest deviation from the horizontal plane. Construct an endplate and right angle line (red). The enclosed angle is the degree of kyphosis or lordosis (red lines).

Side view As viewed from the side, the back should curve evenly without any sharp angulation. A sharp angular segment of the spine is seen in Scheuermann kyphosis.

Neurological examination should be part of the examination. In addition to the routine assessment, assess abdominal reflexes. Abdominal reflexes are assessed by gently stroking each quadrant of the abdominal wall [1]. Absence or asymmetry suggests a subtle neurological abnormality that may indicate the need for more intensive neurological investigation such as MRI.

Imaging Studies

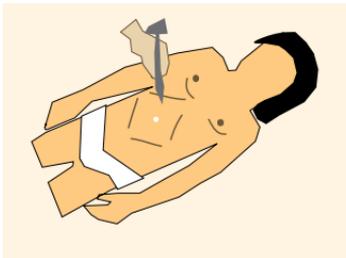
Radiographs and other imaging studies are indicated to measure the vertebral curves and to further assess specific problems identified by the physical examination [2 previous page and 1 opposite page].

Radiographs Make PA and lateral spine films in the upright position on 36-inch film using shielding and techniques that avoid excessive radiation exposure. Order oblique lumbosacral views to assess the pars if spondylolysis is suspected and not seen on lateral view.

Bone scans are usual in assessing back pain when radiographs are negative or equivocal. SPECT imaging is useful to assess subtle pars reactions.

CT studies are useful to detail bony deformities or lesions.

MR imaging is used to study patients with neurological findings, those with unexplained progression of deformity, and certain types of deformity as well as preoperatively for children with neurological impairment. These studies are helpful in evaluating tumors, congenital abnormalities such as Chiari malformation, various cysts, tethered cords, and filum terminale anomalies.



1 Abdominal reflexes. Stroke each quadrant of the abdomen with the base of a reflex hammer to assess the symmetry of the reflex.



2 Truncal inclination. Asymmetry may be assessed with an inclinometer or scoliometer. Measures above 5-7° are an indication for radiographic studies.

Spine and Pelvis Congenital Deformities

Diastematomyelia

This is a congenital defect with a central cartilagenous-bony projection that divides the spinal cord [2].

Diagnosis Cutaneous lesions occur in most with a hairy patch, dimple, hemangioma, subcutaneous mass, or teratoma at or near the level of the diastematomyelia. Other deformities are common. Nearly all have some associated anomaly like spinal dysraphism, asymmetry of the lower extremities, club foot, or a cavus foot. Two-thirds have congenital scoliosis. Two-thirds are located in the lumbar spine. Half have neurological abnormalities.

Management Resect the spur in patient with progressive neurological findings. Follow the others and consider resection should neurological findings develop or if correction of spinal deformity is planned.

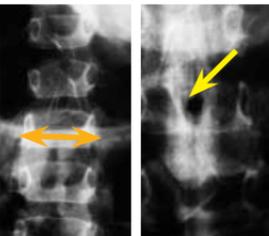
Sacral Agenesis

Caudal regression or sacral agenesis includes a spectrum of abnormalities [1 next page] with hypoplasia or aplasia of the sacrum, which is most common in offspring of diabetic mothers.

Clinical features include knee-flexion contractures with popliteal webbing, dislocations and flexion contractures of the hips, scoliosis, equinovarus deformities of the foot, and instability at the spinal-pelvic junction. These deformities vary in severity with the level of the agenesis and the resulting loss of motor power. Neurological features may be most predictive of progression and MR imaging is helpful in assessment.

Imaging Method	Condition
Radiography	Initial study
PA	36-inch standing for scoliosis
Lateral	36-inch standing for kyphosis or lordosis
Oblique lumbar	Spot L-S spine for spondylolysis
CT scans	Fracture, tumors
Bone scans	Back pain, infection, tumors
MRI	Spinal dysraphism, cord lesions, tumors, abscesses

1 Uses of imaging methods for spinal disorders. Avoid ordering a battery of studies, as this is expensive and often exposes the child to unnecessary radiation.



2 Spinal dysraphism.

Diastematomyelia and other congenital spine defects should be considered in children with cavus feet or limb hypoplasia (red arrow). The interpedicular distance is widened (orange arrow) and a midline bony bar bisects the spinal cord as shown on myelography (yellow arrow).

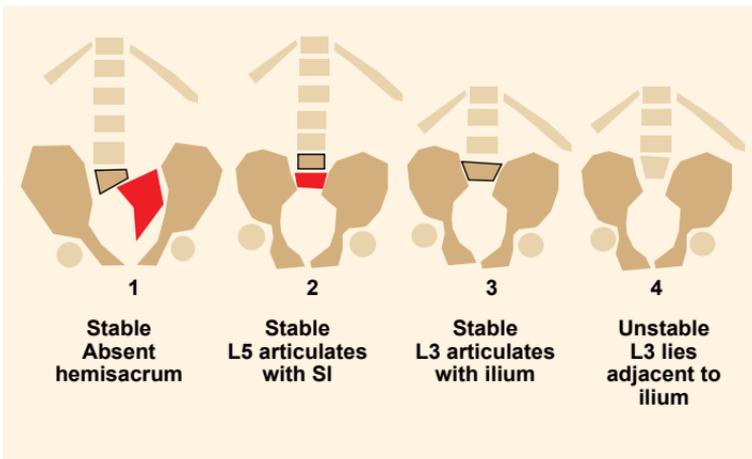
Management is often difficult and depends upon the deformity, motor, and sensory status. Knee flexion deformities are difficult to correct, and recurrence is common. The combination of limited operative procedures, orthotic or mobility aids are tailored to the child. Spine–pelvic instability and hip dislocations are often better tolerated than the stiffness caused by surgical stabilization or reduction.

Exstrophy Bladder

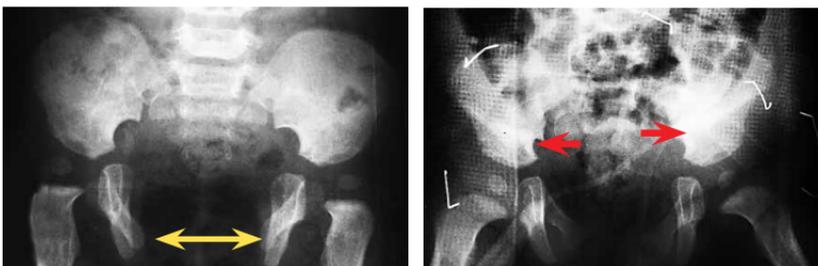
A failure of anterior closure of the pelvis results in pelvic diastasis and an open bladder [2].

Clinical features include pelvis diastasis, acetabular retroversion, and lateral rotation of limbs with out-toeing gait. This out-toeing tends to improve with age.

Management Orthopedic disabilities are insufficient to require correction. Pelvic osteotomy may be required during bladder reconstruction to facilitate closure. Perform bilateral supra-acetabular osteotomies and stabilize with a spica cast following urological repair.



1 Sacral agenesis classification by Renshaw. The sacrum may be hypoplastic or completely absent (red). The spine–pelvic relationship may be stable or unstable. Radiographs show a type 3 deficiency (yellow arrow). Based on Renshaw (1978).



2 Bladder exstrophy. This is associated with separation of the pubic bones (yellow arrow) and retroversion of the acetabula. Bilateral iliac osteotomies (red arrows) were performed to facilitate bladder reconstruction.

Back Pain

Back pain in children is usually caused by some significant organic disease.

Prevalence

Back pain becomes increasingly common during childhood [1]. By mid-teens recurrent or chronic pain occurs in about a quarter of boys and a third of girls.

Evaluation

Be concerned about a history of back pain in children. Sometimes the presenting symptoms of serious conditions may be misleadingly mild, and the spectrum of causes and mode of presentation differ from adults.

Worrisome features include onset before age 4 years, symptoms persisting beyond 4 weeks, interference with function, systemic features, increasing pain, neurological findings, and recent onset of scoliosis.

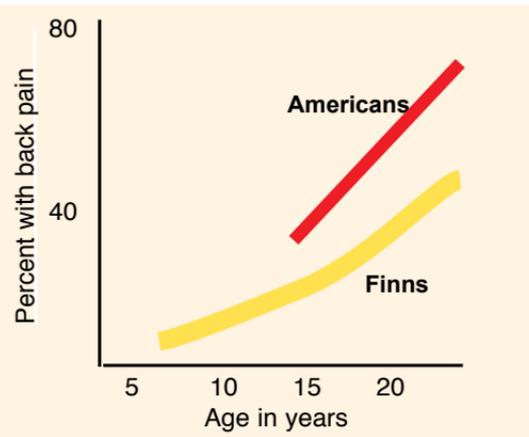
Examine with a focus on mobility, symmetry, tenderness, neurological status, and hamstring tightness.

Image first with conventional radiographs. Supplement with a bone scan as necessary. High-resolution SPET imaging may be useful in assessing adolescent stress injuries. Add MRI if tumor or infection is suspected.

Idiopathic Back Pain

Adolescent benign back pain Back pain without physical abnormalities accounts for an increasingly large proportion of the category with advancing age. Overall about half of children's and adolescents back pain falls into this category.

- **Management** may be difficult. Some suggest limiting backpacks to less than 20% of body weight (not evidence based). Encourage activity, a healthy lifestyle, and weight control. Provide reassurance. Consider this as a *backache* that is common, requires no treatment, and is best ignored.
- **Prognosis** If back pain is present in adolescence together with a positive family history of back pain, nearly 90% of adolescents will have back pain in adult life. Psychosocial problems are more significant than
- **structural abnormalities in determining the likelihood that back pain will become chronic** [1].



1 Percentage of children with back pain.

Back pain increased from about 1% at 7 yrs. to nearly 20% in adolescents of Finnish children and 56% in adults. From Taimela (1997). In Americans incidence is about 30% in adolescence, and about 75% in adults. From Olson (1992) and Balague (1995).

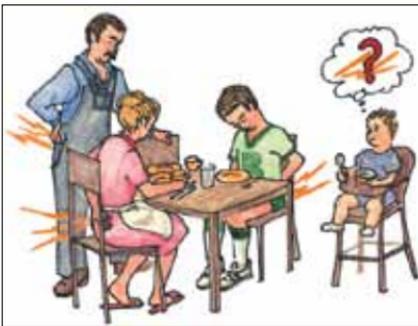
Conversion reaction Reflex sympathetic dystrophy or conversion hysteria may underlie back pain. The typical patient presents with gross, bizarre, and disabling symptoms. Most are adolescent girls. As this type of back pain is very difficult to manage, consider referring to an adolescent medicine specialist or pediatric rheumatologist who has experience in managing this problem. Management often includes physical therapy, psychotherapy, and supportive measures.

Rheumatoid Spondylitis

Rheumatoid disorders may cause back and pelvic pain. The age of onset is usually between 4 and 16 years. More than 90% of patients are HLA-B27 positive with the absence of RF and ANA. About a third will have a family history. Symptoms include peripheral arthritis, usually pauciarticular and asymmetric, involving big joints of the lower limbs. Many complain of heel, back, or sacroiliac pains. An acute iridocyclitis may occur. Most patients develop radiographic sacroiliitis. Refer to a rheumatologist.

Cervical Disc Space Calcification

Cervical disc space calcification is a rare, idiopathic, inflammatory condition with clinical manifestations of fever, neck pain and stiffness, and eventual disc space calcification [2]. The pain and fever resolve spontaneously; calcification is seen at the end of the inflammatory phase. Often residual narrowing and irregularity of the disc space is seen if radiographs are made. Manage with rest, a cervical collar, and a non-steroidal antiinflammatory agent. Resolution of the acute symptoms usually occurs within 7–10 days.



1 Familial back pain. The child may learn about back pain from siblings and parents.



2 Cervical disc space calcification. Note the calcium deposits in the disc space (arrow).

Vertebral Tumors

Benign

- Eosinophilic granuloma
- Osteoid osteoma
- Aneurysmal bone cyst
- Osteoblastoma
- Neurofibromatosis
- Osteochondroma

Malignant

- Ewing sarcoma
- Lymphoma bone
- Leukemia

Spinal Cord Tumors

Benign

- Neurofibroma
- Lipoma
- Spinal cysts

Malignant

- Astrocytoma
- Ependymoma
- Mixed glioma
- Ganglioglioma

3 Vertebral (bone) and spinal cord tumors.

Tumors

Tumors may be metastatic or primary. Primary tumors may arise from the cord or bone [3 opposite page].

Metastatic Tumors

These tumors are most common in the thoracic, lumbar, then cervical spine [1]. Manage with chemotherapy and radiation. Mortality is high. Those who survive are likely to have deformity. Early stabilization may prevent progression of the deformity.

Primary Tumors

Primary tumors may occur in the vertebrae or cord. Most vertebral tumors are benign, most cord tumors are malignant. Either type may cause spinal cord compression [2].

Spinal cord tumors cause diagnostic difficulties. They may present to the orthopedist with torticollis, scoliosis, gait disturbances, foot deformities, or back pain. Often forward bending is limited and asymmetrical. Perform a careful neurological examination. Study with plain radiographs first. Look for changes in intrapedicular distance. MRI studies are usually diagnostic.

Metastatic tumors

Ewing sarcoma
Rhabdomyosarcoma
Adenocarcinoma
Neuroblastoma
Misc. other tumors

Cord compression

Neuroblastoma
Sarcomas
Astrocytomas
Lymphomas

1 Metastatic tumors to spine. From Freiberg (1993).

2 Tumors causing cord compression in children. From Conrad (1992).



3 List on forward bending. This boy with an osteoid osteoma shows asymmetrical forward bending. Bending is restricted on the right side (arrow).

Vertebral tumors are more common. Most are benign. Most present with pain. Duration of symptoms from benign tumors is usually longer than those from malignant tumors. Most may be diagnosed by conventional radiographs.

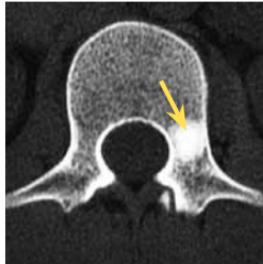
- *Osteoid osteoma and osteoblastoma* cause classic night pain, usually secondary scoliosis, limited spinal mobility [3 previous page], often tenderness, and sometimes classic radiographic features [3]. Bone scans are often diagnostic. Excision is often necessary. Exactly localize with preoperative imaging.
- *Eosinophilic granuloma* causes pain, tenderness, limited mobility, and usually a focal lesion. The classic vertebrae plana [1] is often absent. For solitary, uncomplicated lesions, observational management is appropriate. If lesions are multiple or if neurological involvement is present, operative resection may be necessary.
- *Aneurysmal bone cysts* cause pain, rarely cord or root compression, sometimes deformity, and limited mobility. Radiographs are often diagnostic with expansion and ballooning of the cortex [2]. Management is often difficult. Manage with preoperative selective arterial embolization, intralesional excision curettage, bone grafting, and fusion of the affected area if instability is present.



1 Disc collapse from eosinophilic granuloma. Note the vertebral collapse. The appearance is classic.



2 Aneurysmal bone cyst in 15-year-old boy. Note the expansile cystic lesion (red arrows).



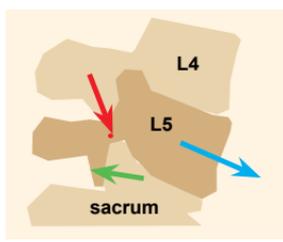
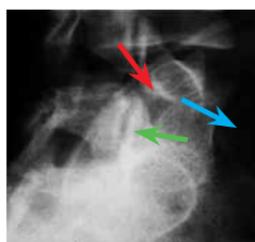
3 Osteoid osteoma spine. This lesion caused severe night pain. Enlargement of pedicle is seen on radiographs (red arrow). The bone scan showed a focal hot spot (yellow arrow) and the CT scan shows the sclerotic lesion (orange arrow). Excision was curative.

Spondylolysis and Spondylolisthesis

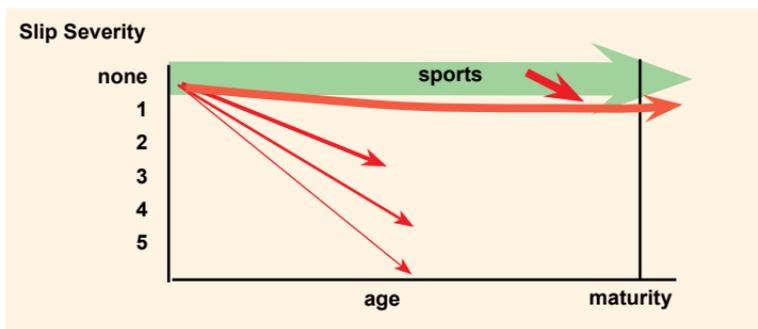
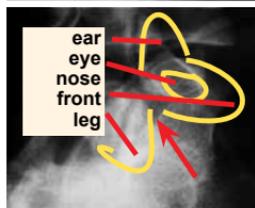
Bilateral or unilateral defects of the pars interarticularis cause spondylolysis [1]. This defect may allow displacement of the vertebrae which is called spondylolisthesis. These lesions are the most common cause of structural back pain in children and adolescents.

Pathogenesis

In children these conditions are usually due to a stress fracture through a congenitally dysplastic pars interarticularis [2]. This inherent weakness occurs more commonly in certain races (such as Inuit peoples), families, or individuals. Often the defects are associated with spina bifida occulta. Spondylolisthesis occurs in about 4% of 4-year-old children and increases to about 6% by maturity. Spondylolisthesis occurs in about a third of those with pars defects, especially in those with mechanical instability. These lesions occur more commonly in children with abnormal bone or connective tissue, as occurs in conditions such as Marfan syndrome and osteopetrosis. Lesions are common in children who participate in certain sports that cause hyperextension of the lumbar spine with rotation such as gymnastics, wrestling, diving, and weight lifting. Progression after adolescence is unusual.



1 Spondylolysis. The fracture through the pars is shown by red arrows. Note the fracture through the pars as shown on the oblique radiograph (upper left) and diagram (upper right). The scotty dog analogy is often used (lower left) to describe the vertebral elements (yellow lines). The neck is the site of fracture. On the model (lower right), the site of fracture is shown.



2 The natural history of pars defects. Most defects develop during early childhood and remain mild. Others develop in late childhood usually due to repetitive trauma from certain sports or less commonly acute trauma.

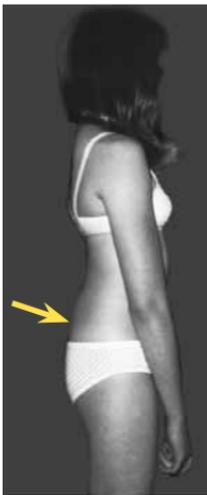
Clinical Features

History and physical examination The child usually complains of back pain. Tenderness may be present at the L5-S1 level. If the displacement is severe, a prominence is palpable over the defect [1]. Straight leg raising and forward bending may be limited. The neurological examination is usually normal. If the condition is acute, secondary scoliosis may be present.

Imaging First, order a standing lateral radiograph of the lumbosacral spine. A forward displacement of the body of L5 or L4 establishes the diagnosis. If no displacement is present, order oblique radiographs of the lower lumbar spine to assess the status of the pars. Spina bifida occulta is common in children with the defect. A bone scan may show reaction [2] before radiographs show a defect and may be used to determine the activity and healing potential of the lesion [3]. Even more sensitive is the SPECT scan or MR imaging in demonstrating bone reaction. Rarely the slippage is severe and a noticeable deformity is present.

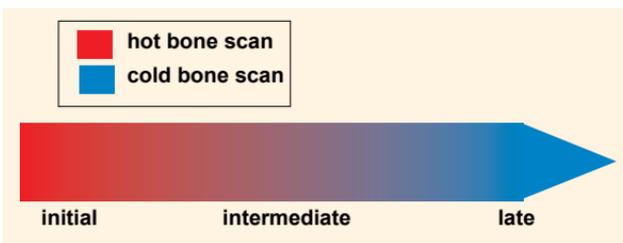
Classification Wiltse classifies spondylolisthesis into two types:

- *Dysplastic* is a congenital facet deficiency allowing slippage.
- *Isthmic* allows slippage due to a defect in the pars interarticularis. These lesions may be due to a fatigue fracture, a stress fracture, or elongation without fracture.



1 Grade 5 spondylolisthesis. The severe slip produces a flattening of the back (yellow arrow) and complete forward displacement of L5 on the sacrum (red outlines).

2 Unilateral spondylolysis. This bone scan shows (arrow) an active unilateral defect.



3 Activity of spondylolysis. Stages of activity. Acute stage following trauma (red), intermediate stage, and late stage (blue).

Grade the degree of slip in severity and activity (duration).

- *Severity* Is usually assessed by the the amount of displacement (slip grade) and rotation (slip angle). Rotation usually occurs in slips when displacement is greater than 50% [1].
- *Activity* Grade on the duration or activity [2]. Recent fractures are active and show increased uptake on bone scan. Cold lesions are chronic, inactive, and less likely to heal.

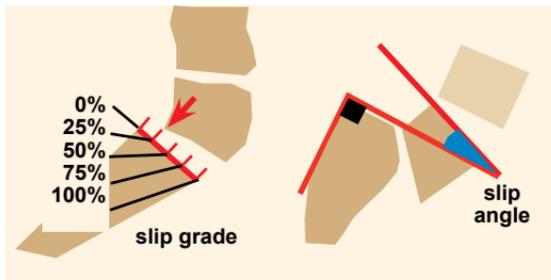
Progression Pain is most pronounced at the time of onset or fracture. Most isthmic lesions become stable and painless with time. Pain is aggravated by activity, especially competitive sports. Often lesions are symptomatic in adolescence but become painless in adult life when activity levels are reduced. The incidence of back pain is comparable to normal population levels.

Management

Management is based on the patient’s age, degree of deformity, type of lesion, activity, and physical activity level.

Spondylolysis management depends upon the activity of the lesion.

- *Acute lesions* from an acute injury or recent overuse experience are managed by reduction of activity and usually an under-arm brace. Often these lesions will heal.
- *Established lesions* Manage symptoms with NSAIDS and activity modification. Operative stabilization is seldom necessary.



1 Slip grade and angle. Slip severity is usually quantified using the slip grade (displacement) and slip angle (rotation) together.



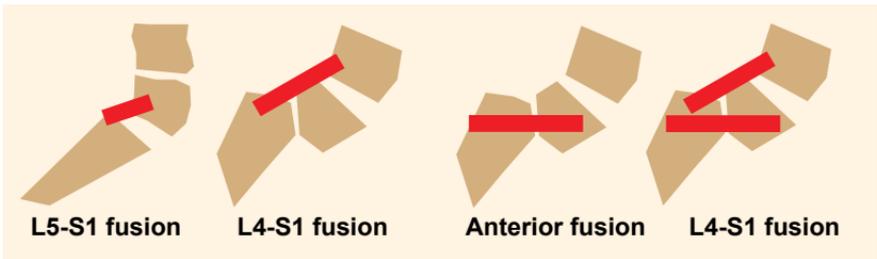
2 Slip severity. The severity of the slip is assessed by the degree of displacement of L5 relative to the sacrum in 5 grades. Note that the slip angle increases progressively through grades 3 to 5.

Spondylolisthesis is managed based on severity of the slip considering the displacement and slip angle. If fusion is required it is often performed without reduction [1].

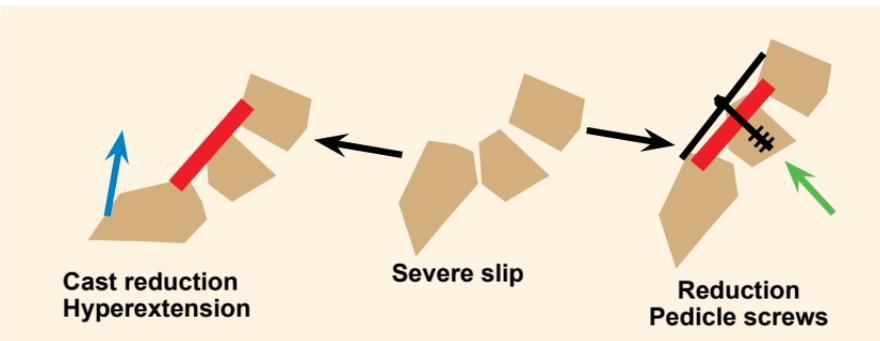
- *Grade 1–2 slips* Manage with NSAIDS, activity modification, and TLSO as necessary to control symptoms. Follow with standing lateral radiographs.
- *Grade 3 slips* Most require operative stabilization in children. Fuse L4–S1 level with posterolateral autogenous grafting. See page 411.
- *Grade 4 slips* These slips may require fusion of L4–S1 as the displacement may be significant making identification of the transverse process of L5 difficult. If slip angle is severe, reduction is sometimes elected [2 this page and 1 opposite page]
- *Grade 5 (spondyloptosis)* management is controversial. *In situ* fusion provides pain relief and safety but the deformity remains. Reduction incurs greater risk but improves appearance and posture [1 on page 302].

Special situations requiring tailoring of management.

- *L4 spondylolisthesis* is less common, more mechanical in etiology, often causes more symptoms, and is more likely to require operative stabilization.
- *Spondylolysis* with persisting symptoms may be managed by repair of the pars defect with grafting and fixation.



1 **In situ fusion.** The fusion is based on the severity of the slip.



2 **Reduction techniques.** Reduction of the severe slip may be done with hyperextension casting (blue arrow) or with pedicle screws (green arrow). Once reduced, a two-level fusion is usually performed.

Scheuermann Disease

Scheuermann disease is a familial disorder of the thoracic spine producing vertebral wedging and kyphosis greater than about 45° [3].

Clinical Features

A history of heavy physical loading from athletics or work is common. Often the deformity is familial [2].

Clinical Features

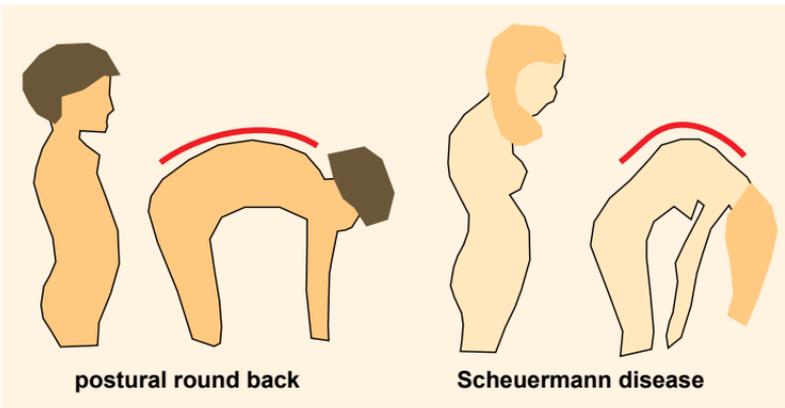
Patients often complain of deformity, fatigue, and sometimes pain. The normal even contour of the spine is lost with an abrupt kyphotic segment at or above the thoracolumbar level. Tenderness over the apex may be present. Radiographs show anterior body wedging. Mild scoliosis is common. The strict definition requires a wedging of at least 5° involving three vertebrae [1 next page].



1 Reduction and fixation. This grade 3 slip was reduced, fixed with pedicle screws, and fused.



2 Familial Scheuermann. This father and son have the same fixed deformity.



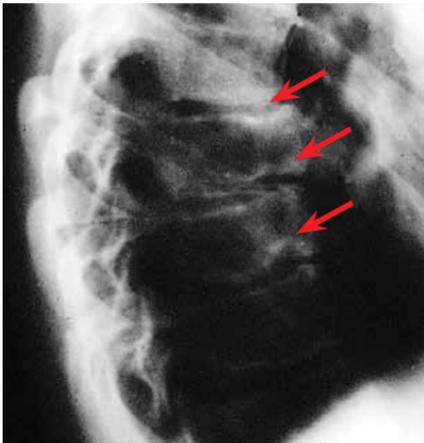
3 Differentiating postural round back and Scheuermann disease. Note the smooth contour of the back on forward bending in the child with round back as compared with the angular pattern in the child with Scheuermann disease.

Manage

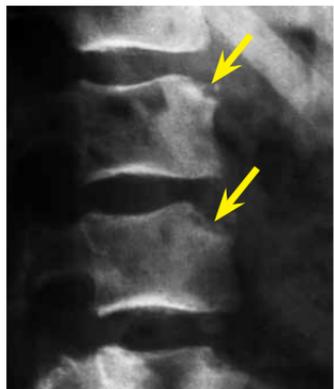
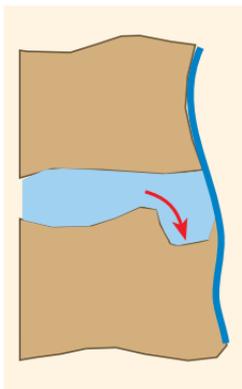
Treat the pain by NSAIDs, rest, and stress reduction. Sometimes a TLSO will be helpful in controlling the pain. Management of the deformity is discussed on page 176.

Schmorl Nodes

These nodes are vertical herniations of the intervertebral disc through the vertebral endplate causing narrowing of the disc space [2]. Sometimes the condition is referred to as *lumbar Scheuermann disease*. This herniation is most common in adolescents, is often associated with trauma, and may be the cause of back pain. The lesions may be seen on plain radiographs, although MR imaging is most sensitive and may be indicated when the diagnosis is uncertain. Manage by rest, NSAIDs, and sometimes a TLSO.



1 Painful kyphosis. This 16-year-old male has pain and tenderness over the lower thoracic spine. Note the narrow disc spaces, and erosion and deformity of the vertebral bodies (red arrows).



2 Schmorl nodes. With vertical loading, the nucleus may herniate into the vertebral body (red arrow) producing pain and atypical radiographic defects (yellow arrows).

Discitis

Discitis is an inflammation (probably infection) of disc space that involves the lower thoracic or upper lumbar disc spaces in infants and children. Unlike other musculoskeletal infections, discitis usually resolves spontaneously.

Clinical features

The clinical features of discitis are age related. Discitis in the infant is characterized by fever, irritability, and an unwillingness to walk. The child may show constitutional illness with nausea and vomiting. The adolescent may complain of back pain. Because the symptoms are vague and poorly localized, the diagnosis is often delayed. The findings of fever and malaise, a stiff back, unwillingness to walk, and an elevated ESR and CRP are suggestive of discitis.

Imaging Early in the disease, a bone scan may show increased uptake over several vertebral levels [1]. After 2–3 weeks, narrowing of the disc space is seen on a lateral radiograph of the spine. MRI often shows worrisome features and may lead to overtreatment [2].

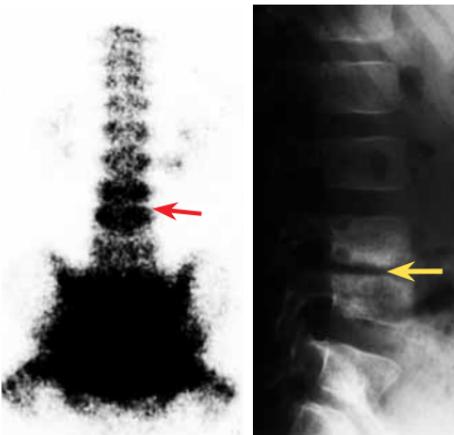
Aspiration or biopsy Disc space aspiration is not necessary unless the disease is atypical.

Management

Manage based on the stage and severity of the disease. If the child is systemically ill, antistaphylococcal antibiotic treatment is appropriate. If the child is acutely ill, an intravenous route is appropriate. Otherwise, oral medication is adequate. Continue antibiotics until the ESR returns to normal. For comfort, consider immobilization in a “panty spica” or brace for a period of several weeks.

Prognosis

Long-term studies show a variety of abnormalities that include residual narrowing, block vertebrae, and limited extension, but the likelihood of back pain is not increased.



1 Discitis L4-5. The typical features of discitis are shown on different imaging studies. The bone scan shows increased uptake (red arrow), and later the lateral radiograph shows narrowing (yellow arrow) of the disc space.



2 MRI of discitis. The typical intense inflammatory reaction seen on MRI may lead to over-treatment.

Disc Herniation

Disc herniations occur rarely in adolescents. Predisposing features include positive family history, recent trauma, facet asymmetry, spinal stenosis, transitional vertebrae, and spondylolisthesis.

Clinical Features

Herniations usually occur at L4–5 or L5–S1 levels, often producing radicular pain and secondary spinal deformity. The patient may be seen because of scoliosis or a list. Straight leg raising is limited, and neurological changes are variable. Radiographs are usually normal. Occult spina bifida is more common in these patients. MRI studies or myelography show the lesion [1]. Disability is increased if the herniation is associated with spinal stenosis. Be aware that fracture of the lumbar vertebral ring apophysis may be confused with disc herniations.

Management

Manage first with with NSAIDs, rest, limited activities, and a TLSO. Persisting or increasing disability are indications for MR imaging and operative disc excision. Endoscopic or open discectomy are successful in 90% of cases.



1 MRI in disc herniation. The posterior bulging disc at L4-5 is clearly demonstrated on MRI.

Scoliosis

Scoliosis is often defined as simply a frontal plane deformity of the spine $>10^\circ$. The deformity is much more complex and includes significant transverse and sagittal plane components. The causes of scoliosis are numerous [1]. Mild truncal asymmetry occurs in as much as 10% of the population and may be considered as a variation of normal. Curves greater than 10° are abnormal and in the growing child may progress to cause a significant problem. Scoliosis is the most common back deformity.

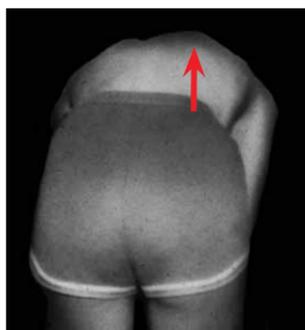
Evaluation

The evaluation should establish the diagnosis, determine the severity, and allow an estimation of the potential for progression of the scoliosis.

History Inquire about the age of onset, progression, and previous management. A family history of deformity [2] or pain is important as both run in families. Painful scoliosis in the child suggests an inflammatory or neoplastic basis for the scoliosis.

Screening examination Start with a screening examination. Look for conditions such as Marfan syndrome or the café au lait spots of neurofibromatosis. Assess the child's limb lengths and gait, and perform a neurological examination.

Category	Disease
Secondary	Muscle spasm Leg length inequality Functional disorders
Congenital	Failure formation or segmentation Neural tissue disorders
Neuromuscular	Upper neuron, such as cerebral palsy Lower neuron (polio) Myopathic, such as muscular dystrophy
Constitutional	Syndromes Metabolic disorders Arthritides
Idiopathic	Infantile (0–3 years) Juvenile Adolescent
Miscellaneous	Traumatic Neoplastic Secondary to contractures Iatrogenic, such as radiation and thoracoplasty



1 Classification of scoliosis. Scoliosis is classified into general categories.

2 Familial scoliosis. Scoliosis runs in families. Perform a forward bending test on the parents and siblings. This mother (right) was unaware of her scoliosis.

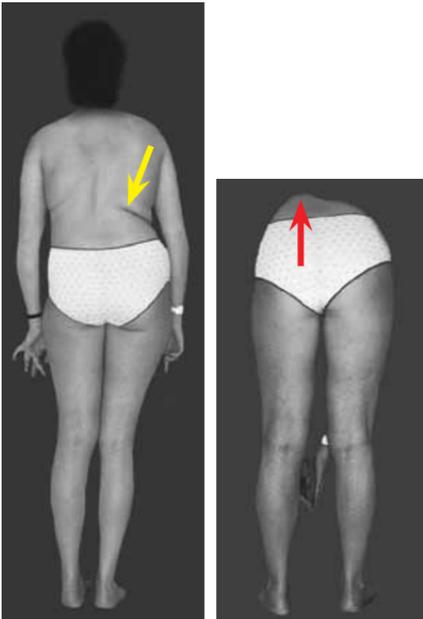
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Back examination Note truncal symmetry [1]. Note differences in shoulder height, scapular prominence, flank crease, and pelvic symmetry. Ask the patient to bend forward. Be concerned about stiffness or a list as these suggest an underlying neoplastic or inflammatory process.

Perform the forward bending test. Visually scan each level of the spine to assess symmetry. If a "rib hump" is present, measure it with a scoliometer. This simple device measures the tilt of the rib hump. Assess the balance of the spine using a plumb line [2]. The displacement of the weight from the buttock crease is recorded.

Radiographs Radiographs are indicated if the scoliometer reading is greater than 7° or if progression is likely. Progression is more likely if the child is under 12 years, when others in the family have significant curves, or if any findings suggest that the curve may not be simply idiopathic. Radiographs should be made on 36-inch film and taken standing with shielding. A single PA radiograph is satisfactory for screening or a baseline study.

Measure the curve by the Cobb method [1 opposite page]. Measure the level with the greatest tilt. Note the "apical vertebra" as this defines the level of the curve [2 on page 313]. Curves greater than 10° are considered significant.



1 Adolescent idiopathic right thoracic left lumbar scoliosis. The flank crease (yellow arrow) and thoracic prominence (red arrow) are shown.



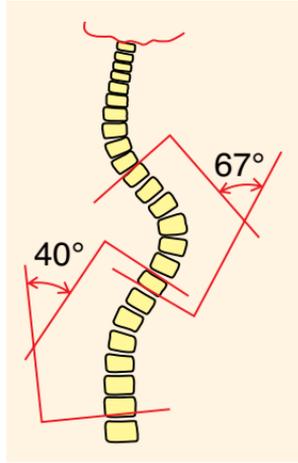
2 Classification of scoliosis. Scoliosis is classified into general categories.

Secondary or Functional Scoliosis

This type of scoliosis can also be described as “functional” because it is secondary to some other problem. The scoliosis usually resolves when the underlying problem is corrected. The scoliosis is usually flexible and nonstructural. There are no bony changes and the rotational elements are minimal. The common causes of functional scoliosis are leg length inequality and muscle spasm.

Leg length discrepancy Differences in limb length produce a transient functional scoliosis. As discussed in Chapter 4, this type of scoliosis seldom becomes rigid or structural, presumably because the scoliosis is present only when the child is standing on both feet. Thus with lying, sitting, and walking, the spine is straight. The fear of causing a structural scoliosis or other back problems is not a valid reason for ordering a shoe lift or for performing limb length equalization procedures.

Muscle spasm Scoliosis may be the presenting sign for several inflammatory or neoplastic disorders [2]. The spinal curvature often functions to relieve discomfort. Thus, the back is curved to reduce pressure on a nerve root from a herniated disc. Management is directed at the underlying disorder. Scoliosis will disappear once the underlying problem is corrected.



1 Cobb method for measuring curves.

The endplate of the most deviated vertebrae are marked and a right angle line drawn. The angle created by the intersecting lines indicates the degree of curvature.

Disease	Comment
Spondylolisthesis	Only with severe displacement
Herniated Disc	Commonly causes scoliosis
Osteoid Osteoma	Focal benign lesion
Intraspinal Tumor	Most serious cause
Discitis	Older child



2 Underlying causes of scoliosis due to muscle spasm. These conditions should be ruled out if the scoliosis is atypical, associated with pain, list, stiffness, tenderness, or obvious muscle spasm.

Infantile Scoliosis

Idiopathic scoliosis is the most common spinal deformity. Idiopathic scoliosis is often divided into three subgroups based on time of onset infantile, juvenile, and adolescent categories or simply into either *early* or *late* onset types. Each group has a different natural history and potential for disability [1]. Scoliosis is often classified simply by the site of the most severe curve [2 opposite page].

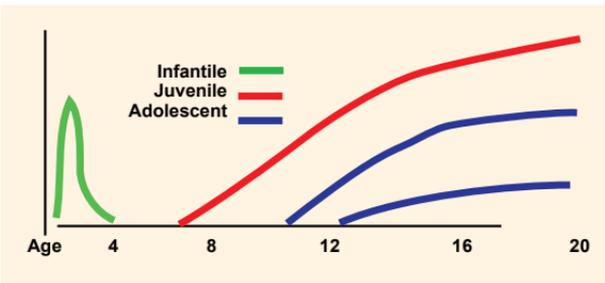
Infantile idiopathic scoliosis occurs in infants and children under 3 years of age. Because the deformity often is associated with plageocephaly and hip dysplasia it is thought to be a positional deformity. Like other position deformities, spontaneous resolution usually occurs. In some cases, the scoliosis is secondary to an underlying spinal abnormality. These cases progress to become severe. Infantile scoliosis is rare in North America.

Evaluation

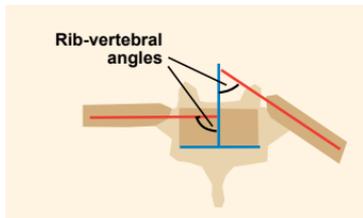
Truncal asymmetry and scoliosis by radiography establish the diagnosis. Most are boys with left thoracic curves. Measure the apical-rib-vertebral angle difference or RVAD [2]. If the RVAD exceeds 20°, study with an MRI as about a quarter will show a significant neuroanatomical abnormality such as Chiari-1 malformations.

Management

Curves with angles of <20° resolve and require only observation. Follow closely curves >20°. If curves progress and exceed a Cobb angle of about 25° manage with a brace. Curves uncontrolled by bracing that exceed 40° may require operative correction. This correction may include instrumentation without fusion to preserve growth, or anterior and posterior fusion to arrest progression and prevent crankshaft deformity. Be aware that following fusion trunk height will be lost at about 0.03 cm per level fused times the years of remaining growth. Operative decisions are often difficult.



1 Natural history of idiopathic scoliosis. Progression is related to the age of onset of the scoliosis.



2 Rib-vertebral angle difference. This is the angle between the axis of the ribs (red lines) and a right angle to the body of the vertebrae (blue lines). The differences is the RVAD.

Juvenile Scoliosis

This form of scoliosis is identified between the 3 and 10 years of age [1]. The course of this early-onset scoliosis is more progressive than the adolescent form and most require bracing.

Etiology

This early-onset scoliosis is more likely to be secondary to some underlying pathology such as a Chiari I malformations [1 next page] or tumors than curves with onset at puberty.

Evaluation

Hypokyphosis with values $<20^\circ$ suggests a poorer prognosis and complicates orthotic management. In addition to the standard measures, for children with curves $>20^\circ$ study with a full spine MRI as 20–25% will show a significant spinal abnormality that accounts for the early onset and progressive course of this type of scoliosis. Measure the RVAD. Curves $<10^\circ$ are usually benign.

Management

Follow for progression. A few curves resolve spontaneously. Institute orthotic management for progressive curves that exceed 20° .

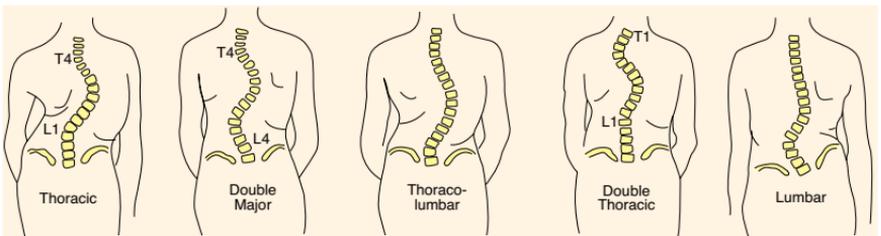
Bracing Manage curves with an apex below T7 with a TLSO. A Milwaukee brace is necessary for more proximal curves. Considering the long duration of bracing necessary, balance brace time with tolerance. Avoid bracing for too many years as the child must endure many years of brace treatment as well as the final surgical correction.

Operative correction is indicated for curves exceeding 40° – 50° . Anterior and posterior fusion are necessary for young children to prevent the crankshaft deformity. Be certain to correct or maintain normal sagittal alignment. Instrumentation without fusion may be considered in very young children but this is controversial.



1 Juvenile scoliosis. This girl shows an elevated right shoulder and thoracic asymmetry.

2 Classification of scoliosis. Scoliosis is classified into general categories.



Adolescent Scoliosis

Idiopathic scoliosis with an onset after age 10 years is the most common and classic form.

Etiology

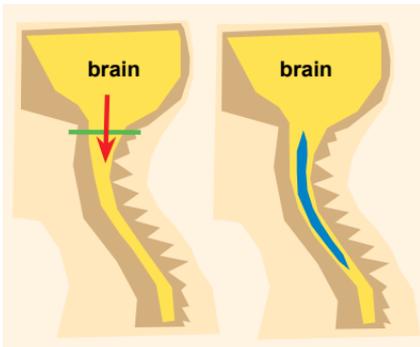
The causes of scoliosis are probably multiple. Individuals with progressive curves show vestibular, height, and gender differences from unaffected controls. A genetic component is present but the mode of inheritance is uncertain.

Prevalence

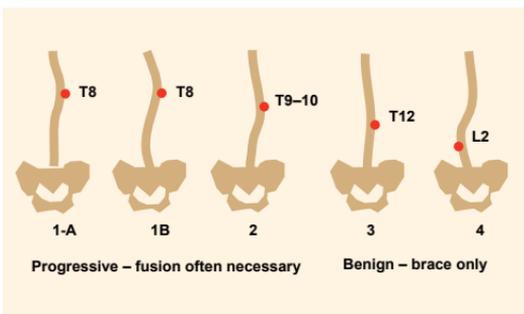
Mild truncal asymmetry occurs in about 10% of the population and is a normal variant. The diagnosis of scoliosis is reserved for curves $>10^\circ$ and this occurs in 2–3% of children, with boys and girls equally affected. Progressive curves are more common in girls by 4–7:1 and prevalence of 0.2% with $>30^\circ$ and 0.1% $>40^\circ$. About 10% of children identified with scoliosis require treatment.

Natural History

The potential for progression depends upon curve type [2], severity, level of maturation [1 opposite page], and Risser sign [2 opposite page]. In adults, curves $<30^\circ$ progress little, curves $30\text{--}50^\circ$ progress about $10\text{--}15^\circ$ over a lifetime. Curves $50\text{--}76^\circ$ progress about 1° a year. Individuals with scoliosis have normal mortality rates. Curves $>100^\circ$ may reduce pulmonary function. Back pain occurs in about 80%, which is comparable to the general population. Curves at the lumbar and thoracolumbar regions are most likely to be painful.



1 Chiari malformation and syrinx. This malformation is a displacement of the cerebellum into the spinal canal (red arrow). These lesions may be associated with a syrinx (blue).



2 Progressive curve patterns in juvenile idiopathic scoliosis.

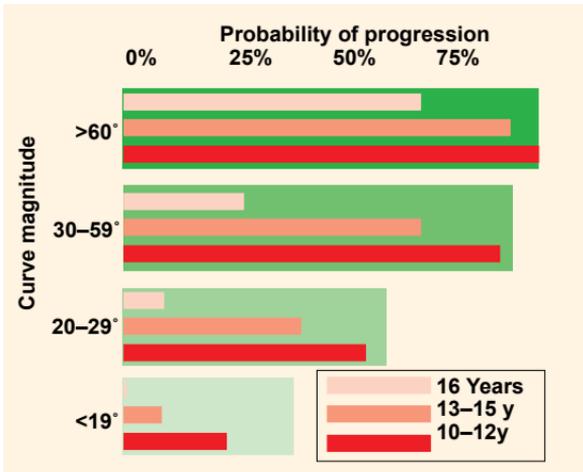
Pattern of curves and apex (red dot) level are shown. Curves 1 and 2 are common, often progressive, and usually require fusion. Curves types 3 and 4 are less common, more benign, and usually managed by bracing. Based on Robinson and McMaster (1996).

School Screening

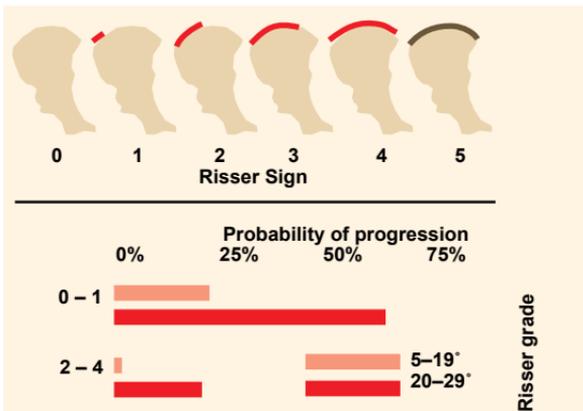
The value of school screening is controversial. The advantage is the earlier detection of deformity. The disadvantage is the large numbers of children with *schooliosis*, those with minimal truncal asymmetry that are referred to physicians, often studied radiographically, and subjected to the anguish of having *scoliosis*. Proposals to be more efficient have included establishing a threshold of 7° scoliometer reading and biannual screening.

Evaluation

Perform an orthopedic evaluation, measure the the Cobb angle [1 next page], and obtain bending films [2 next page] if operative correction is planned. Assess the psychosocial situation. Be aware that scoliosis has been shown to increase the risk of suicidal thought, worry, and concern over body image. Try to estimate the tolerance of the child and family to long-term treatment. Exceeding this tolerance results in noncompliance and problems that are sometimes preventable. Support groups, counseling, and special help from family members may be essential.



1 Probability of progression (>5°) based on magnitude of curve at age of initial detection. From data of Nachemson, Lonstein, and Weinstein (1982).



2 Probability of progression based on curve magnitude and Risser grade. From data of Lonstein and Carlson (1984).

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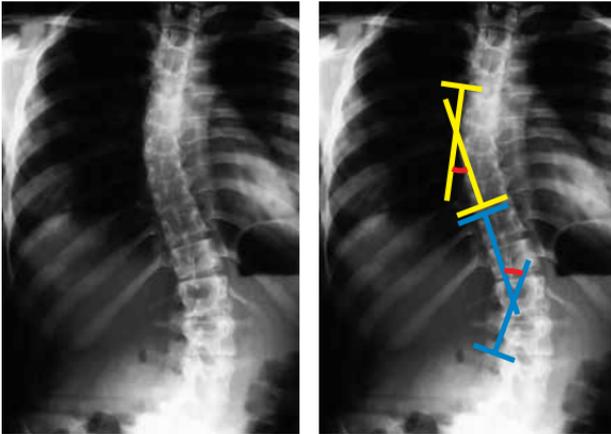
Management Principles

Manage scoliosis by observation, bracing, or surgery. Exercises, electrical stimulation techniques, and manipulation are ineffective and should be avoided. Ninety percent of curves are mild and require only observation.

Reassurance is an important part of management. Avoid the term *scoliosis* for mild curves and simply refer to the deformity as a “mild truncal asymmetry.” This reduces the apprehension that is associated with the diagnosis of *scoliosis*. This diagnosis often causes apprehension as scoliosis is usually equated with treatment either by bracing or surgery.

Indications for treatment should be individualized; however, some generalization can be made.

- **Brace treatment** is indicated for immature patients (Risser 0 or 1) with curves between 25° – 40° . Boys may be treated with Risser 2–3 if the curve exceeds 30° and is progressive. Observe smaller curves for progression. Progression is defined as a documented increase of 5 or more degrees.
- **Operative treatment** is usually indicated for immature patients with curves of $>40^{\circ}$ and mature patients with curves $>50^{\circ}$.



1 Cobb measure of curve. The degree of scoliosis (red arcs) is the angular difference between right angle lines drawn to the most tilted vertebral bodies. Note the double curve with the thoracic apex at T8 and the lumbar curve at L4.



2 Bending study. To assess the stiffness of the curve, bending films are sometimes used. The degree of correction is a measure of curve flexibility and predictive of correction possible with surgery.

Brace Treatment

Although the value of bracing is still questioned, evidence suggests that bracing slows or arrests progression of most spinal curvatures in immature patients with progressive curves between 25° and 40° .

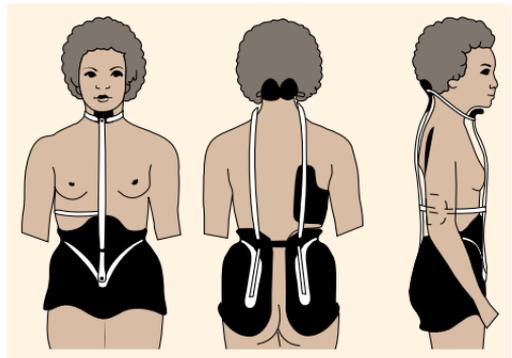
Bracing principles apply to most braces. *Immediate effect* should show a reduction of the curve by $>50\%$. *Introduce the brace* over a period of several weeks. *Encourage acceptance* as quickly as possible. *Discomfort* in the brace should be corrected by making necessary modifications early. Continued discomfort reduces compliance, increasing the risk of bracing treatment failure. *Modifications* in the brace will correct this problem. *Encourage normal activities* while being braced [1]. *Schedule follow-up* visits every 4–6 months to assess fit, size, compliance, and curve progression. Obtain a standing PA radiograph out of the brace to assess progress. *Discontinue bracing* about 2 years post-menarcheal or Risser 4 for girls and 5 for boys. Progression while bracing may indicate the need for operative stabilization.

Bracing options Select the orthosis based on the type and level of curve and the anticipated tolerance of the patient. The most effective bracing types and protocols are most restrictive and cause greatest psychosocial disability. Select a balance that is best for the patient.

- **Milwaukee brace** For upper thoracic curves, the Milwaukee brace is often prescribed. This brace is most restrictive and is compatible with limited activity [2].



1 Milwaukee brace. Encouraging an active lifestyle is especially important in these more restrictive braces.



2 Milwaukee brace. This brace is necessary for upper thoracic curves. It is the most poorly tolerated of the spinal orthoses.

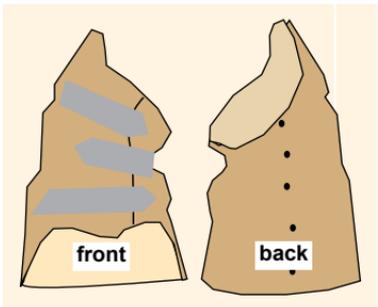
- *TLSO brace* is the most commonly used orthosis. It is appropriate for curves with an apex in the midthorax and below [1]. The Boston brace is prefabricated with custom pads applied by the orthotist. Most include a 15° lordosis correction. The brace may be worn on a 16 or 23 hour per day protocol.
- *Nighttime braces* are best tolerated but effectiveness is controversial. The Charleston bending brace [2] is most widely used. The brace is worn only at night allowing the child freedom during the day.

Dealing with compliance Bracing is uncomfortable, often adversely affects self-image, and imposes some difficulties with social and athletic activities. All of these problems further complicate an already difficult time in life. The physician must not exceed the “tolerance limit” of psychological stress on the girl. If this tolerance limit is exceeded, the girl will become noncompliant and may not return for follow-up. She may simply ignore the problem or seek nonconventional methods of treatment that are less demanding.

- *Improving acceptance* Several methods can be used to reduce the adverse effects of brace treatment of scoliosis [3]. The patient should participate in most of her prebracing activities. The bracing schedule may be tailored to the patient. Some patients are already at or beyond their tolerance limit. It may be best to maintain a relationship with the patient and family and to follow the patient without treatment. If the curve is advanced, it may be best to elect an operative option earlier than is normally appropriate.



Fig. 8.59 Boston brace. These underarm braces are useful for low thoracic and lumbar curves.



2 Charleston bending brace. This underarm bending brace is made for nighttime use. Based on Price, et al (1997).

Technique	Comment
Education	Reassurance
Support Groups	Organize or arrange with other patients wearing braces.
Type of Brace	Use underarm braces when possible
Duration: daytime	Part-time bracing per day Out of brace for school?
Duration: total	Start weaning early if necessary
Activities	Encourage activities in the brace Allow brace-free time for activities
Time out of brace	Be flexible to allow out of brace for special occasions

3 Adverse effects of bracing. These are techniques that may be used to keep the management within the tolerance limit of the patient.

Operative Treatment Principles

Indications Operative management is the most definitive and effective method of management of scoliosis. It is appropriate for curves that exceed 40–50°.

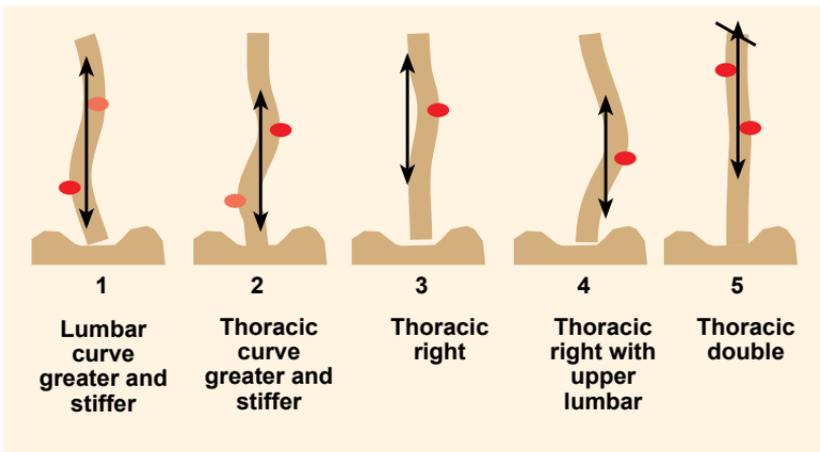
Risks are early and late.

- *Early complications* include usual operative complications and neural injury. Neural injuries occur in about 0.3% of those with standard posterior fusion.
- *Late complications* include pseudoarthrosis in about 2%, progression in about 1%, late degenerative arthritis at vertebral levels below lumbar fusions, post-fusion back pain, and the *crankshaft phenomenon* if fusions are performed in Risser 0 children.

Fusion levels are important to establish thoughtfully. Fusion too short may result in progression; fusion too long increases the risk of pain and degenerative changes. Inappropriate fusion levels may cause spinal malalignment, changes in posture, and post-fusion back pain.

- *Classification of curve patterns* was established to aid in assessment and determining the appropriate levels for instrumentation and fusion. The curves were classified into five types [1]. Types 1, 2, and 5 are double curves. The level of the apex is established by the degree of angulation of the vertebrae and apex of the curve. The rigidity of the curve may be assessed clinically or by bending radiographs. This classification is becoming less commonly used.
- *Extent of fusion* includes the neutral (or stable) vertebrae above and below the primary curve(s). Attempt to reduce the required number of vertebral levels by considering anterior or selective fusions.

Spinal monitoring is designed to monitor spinal cord function during the operative procedure. The wake-up test has been traditional. Somatosensory evoked potential monitoring is currently being supplemented by monitoring motor function.



1 Curve pattern classification. This classification is sometimes still used for classifying curves. Curves may be single or double with the curve apex (shown by red dot or dots) being thoracic, lumbar, or combined. The extent of fusion (arrows) varies with the curve pattern. Based on King, et al (1983).

Operative Technique

Instrument to reduce the scoliosis and maintain or improve sagittal alignment. Avoid excessive distraction and incorporate solid fixation. Decorticate carefully, excise facet joints when feasible, and add supplemental bone. This supplemental bone may be autogenous, bank bone, or agents that induce osteogenesis.

Harrington instrumentation was the initial standard that incorporated distraction and compression of the ends of the curves [1]. This technique provided little control of sagittal alignment and has been largely replaced.

Luque fixation utilizes sublaminar wires fixed to posterior rods.

Drummond fixation employs spinous processes to posterior rods fixation.

Cotrel and Dubouset (CD) introduced a universal system that provides translation and rotation in addition to distraction that permits a solid 3-dimensional correction. Many modifications of this form such as the Isola and TRSH systems have been developed.

Anterior fixation provides excellent stability when extended to or just beyond the neutral vertebrae [2]. This fixation allows correction with the least number of fused segments.

Video-assisted thoracoscopy These procedures allow anterior releases, rib resection and harvesting, and insertion of correctional implants and fusion with reduced morbidity.



1 Spinal Instrumentation. Many forms of spinal instrumentation are used. The classic Harrington instrumentation (red arrow) and more modern CD form (yellow arrow) are examples.



2 Anterior fixation. This lumbar curve was instrumented and fused involving only 5 vertebrae.

Congenital Scoliosis

Congenital structural defects may cause a variety of spinal curves [1]. Such curves are often complex and may require special imaging techniques for assessment. Because these malformations are due to an abnormality of the fetal somite formation, associated lesions in the same somite are common. Thus, the finding of congenital scoliosis, especially one involving the thoracolumbar region, should prompt an ultrasound evaluation of the urinary system and consideration about syndromes such as the VACTERL association.

Pathogenesis

Congenital scoliosis is usually caused by a failure of formation or segmentation [2 on this page and 1 next page]. The progression of the curve is related to the type of bony defect. Curves that are most likely to progress are those with unilateral unsegmented bars that restrict growth on one side while the opposite side grows normally.

Evaluation

Note the severity, symmetry, and flexibility of the curve. Screen the child for additional disorders of the urinary and cardiovascular systems. Murmurs should be evaluated by a pediatric cardiologist. Order a renal ultrasound as 10–20% will have congenital urinary abnormalities, some of which are life-threatening.



1 Congenital scoliosis. This child has upper thoracic congenital scoliosis with severe deformity. This type of deformity should be prevented by early surgery.



2 Grades of severity. The hemivertebrae (green arrow) often produces little deformity. On the other extreme, unilateral fusions of the vertebrae (red arrows) and ribs (yellow arrow) causes progressive severe deformity. This curve was fused in infancy to prevent further progression.

Imaging Study the pattern of the curve on AP and lateral radiographs of the entire spine and additional imaging methods for special situations [2]. Categorize the curve pattern to assess the likelihood of progression. If the curve pattern is ambiguous, CT scans of the apical region are sometimes necessary. MR studies are indicated if neurological abnormalities are found. Plan follow-up and repeat the radiographs in 3–6 months.

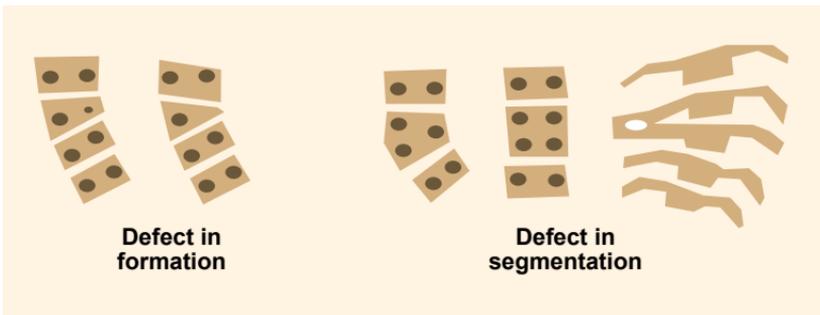
Management

The management of congenital scoliosis depends upon the pattern and severity of the curve and rate of progression.

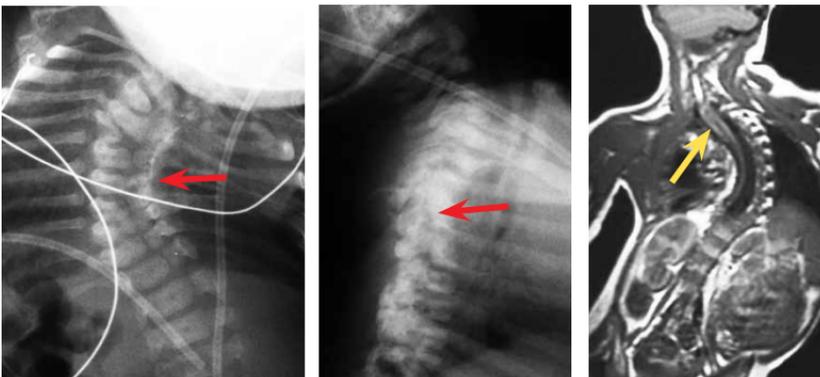
Observation is appropriate when the potential for progression is uncertain. Evaluate every 3 months during the first 3 years and again during puberty when spinal growth is greatest.

Operative treatment is indicated for curves due to unilateral bars. Early *in situ* fusion prevents progression, or anterior and posterior hemifusion on the convex side may result in some correction of the curve with growth. Problems with growth are less severe than for idiopathic curves as growth is already limited due to the underlying deformity. Operative treatment is required in about half of children with congenital scoliosis.

Orthotic treatment of congenital scoliosis is controversial and less effective than for idiopathic curves. Congenital curves that are long and flexible are most likely to respond to brace treatment.



1 Types of congenital scoliosis. The common defects are a failure in formation or segmentation. Complex deformities may show mixed patterns.



2 Special imaging in congenital scoliosis. This congenital scoliosis (red arrows) in a newborn was imaged with MRI because of a neurological deficit. Note the hydromyelia (yellow arrow).

Neuromuscular Scoliosis

Most neuromuscular disorders are associated with scoliosis [1].

Natural History

This scoliosis often occurs early, is rapidly progressive throughout growth, and continues to progress in adult life. Scoliosis often parallels the severity of the neuromuscular disease. Primary curves may interfere with sitting and nursing care. Severe curves may cause cardiopulmonary compromise.

Evaluation

These children have a systemic illness, and a total evaluation is essential.

Physical examination

General evaluation should be thorough. Be certain the diagnosis is accurate to better understand the natural history and potential for disability. Assess the child's motor and mental status, family situation, nutritional status, pulmonary status and general health.

Back examination Observe the child sitting, standing, and walking. Note balance, sagittal alignment, and severity. Examine prone to assess pelvic obliquity [1 next page]. If the scoliosis is secondary to infrapelvic obliquity, focus attention on the hips rather than the spine. Assess sagittal alignment.

Image with PA and lateral 36-inch radiographs. If the hip examination is abnormal, add an AP film that includes the pelvis on the same film to assess the relationship of the hip and spine deformity. Making the spine radiographs with the child sitting is often most helpful.

Laboratory evaluation is essential prior to any surgical procedure. Assess albumen levels (should be > 3.5 g%) for nutritional status, pulmonary function (vital capacity), and lymphocyte levels (above 1500).

Neuropathic	
Upper motor neuron	cerebral palsy spinocerebellar degen. syringomyelia spinal cord tumors spinal cord trauma
Lower motor neuron	poliomyelitis trauma spinal muscular atrophy dysautonomia
Myopathic	
	arthrogryposis muscular dystrophy congenital hypotonia myotonia dystrophica

1 Classification of neuromuscular scoliosis.

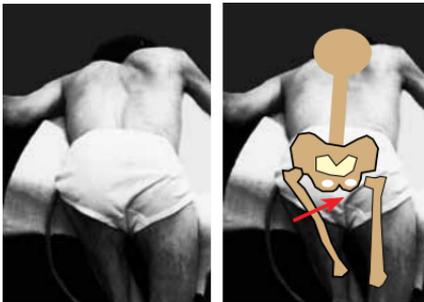
Management Principles

Manage with an understanding of the natural history [2], the potential for disability, and the effectiveness of the various treatment options. As management is often complex, controversial, and long-term, consider what the family values.

Observation is the initial and often primary mode of management for most curves. At each clinic visit, screen for scoliosis with forward bending tests as part of the general physical examination.

Orthotic treatment is controversial [3]. Orthotics are not useful in myelodysplasia or muscular dystrophy and of questionable value in cerebral palsy. Orthotics are often uncomfortable, may cause skin breakdown, decrease pulmonary function, and are expensive for the family. Orthotics may slow progression of the curve in some children and allow a delay in operative correction.

Surgery may be required for progressive curves to provide stability, improve sitting balance, maintain hand function, maintain pulmonary function, facilitate care, and reduce discomfort.



1 Assess type of pelvic obliquity. Position the child prone over the edge of the exam table. Note that in this patient the spine-to-pelvis relationship becomes neutral or normal. The obliquity is infrapelvic from a dislocated hip.



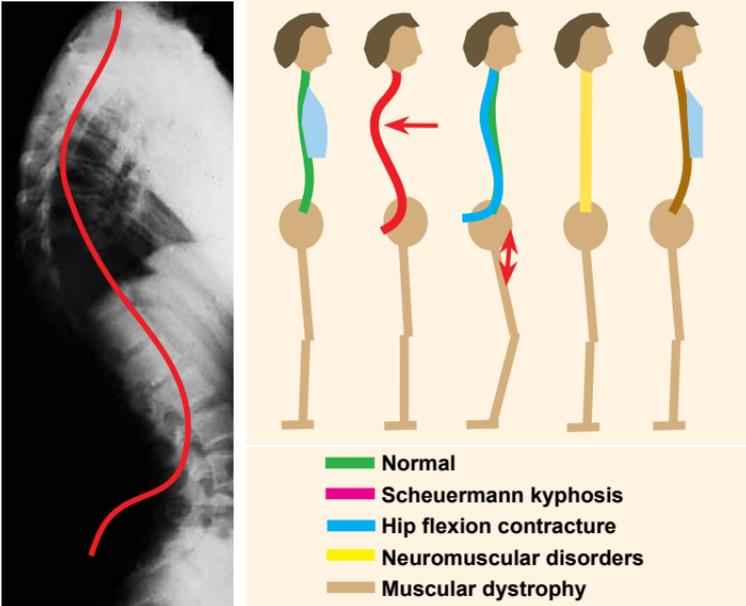
2 Progression of neuromuscular scoliosis. This curve progressed significantly between ages 14 and 18 years.



3 Bracing in cerebral palsy. Bracing in cerebral palsy is controversial.

Sagittal Alignment

Sagittal alignment [1] is affected by our upright posture and significantly affects appearance, cardiopulmonary function, and potential for degenerative arthritis of the spine. As the spine has greater mobility in flexion and extension than side bending, sagittal deformities are not complicated by a rotational component as occurs with scoliosis. The spine has three curves, cervical lordosis, thoracic kyphosis, and lumbar lordosis. Upright posture requires that these curves be balanced; they are interrelated. Furthermore, lower extremity alignment affects the spine. For example, excessive lumbar lordosis is usually compensated by hip flexion.



1 Patterns of sagittal deformity. Normal (green), Scheuermann kyphosis (red); hyperlordosis secondary to hip flexion contracture (blue); flat back (yellow), and thoracic lordosis (brown) with pulmonary compromise.



2 Congenital kyphosis. Vertebral hypoplasia may lead to paraplegia (red arrow). Kyphosis in spina bifida (yellow arrow) can be very severe, causing skin breakdown over the apex and difficulty in positioning.

Kyphosis

Kyphosis is a posterior convex angulation of the spine. Kyphosis is normal for the thoracic spine with normal range from about 20°–50°.

Postural Round-back

This is a normal variation. The major problem is cosmetic. It is flexible as the posture can be improved by asking the child to *straighten-up* and does not cause a permanent deformity.

Congenital Kyphosis

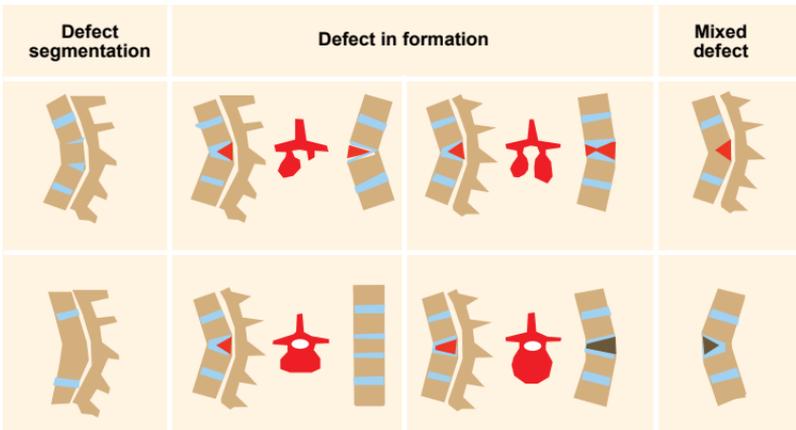
Congenital kyphosis may be due to a failure of formation, segmentation, or mixed types [2 previous page and 2 this page]. The apex of the curve is most common between T10 and L1. Deformities secondary to a failure of formation are usually progressive and may lead to paraplegia. Assess the apex with high-quality radiographs and a CT study if necessary. Classify the type of deformity. For progressive deformities under about 55°–60° fuse posteriorly. More severe deformities may require anterior and posterior fusions.

Scheuermann Kyphosis

This disease often causes both pain and deformity [1]. The diagnosis is discussed on page 166.



1 Scheuermann kyphosis. Note the round back deformity and anterior wedging of vertebrae (red arrows).



2 Classification of congenital kyphosis and kyphoscoliosis. Based on McMaster and Singh (1999).

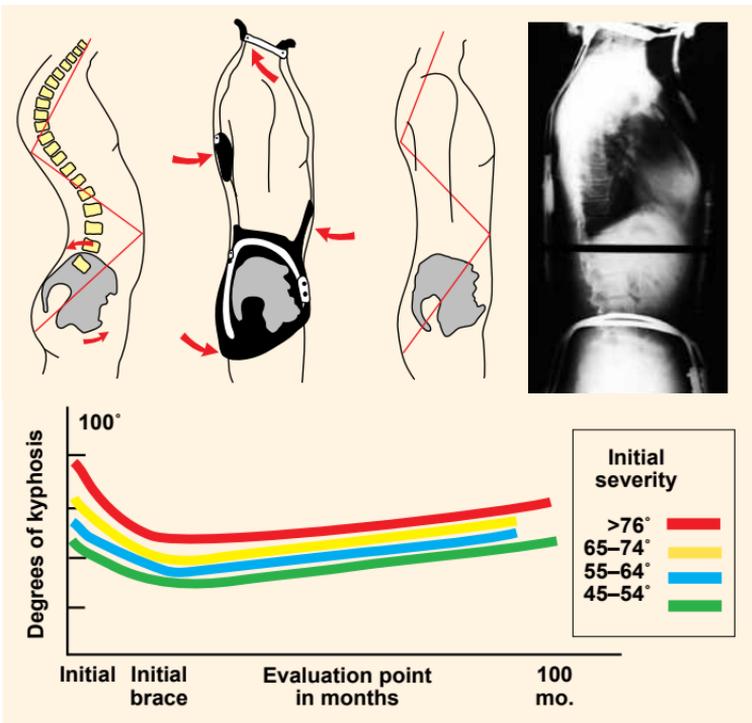
Manage the pain with NSAID and immobilization. Management of the deformity is controversial as long-term disability is mild and effective treatment difficult.

- *Manage mild deformity* curves $<60^\circ$ with observation and encouragement to be physically active.
- *Curves $>60^\circ$* in skeletally immature children (Risser sign <3) may be improved by brace treatment. Consider applying a preliminary hyper-extension plaster cast to improve flexibility. For curves above T7 use a Milwaukee brace [1]. For lower curves use an underarm brace. Brace initially for 20 hrs daily. Once the curve is controlled taper the brace to nighttime use.
- *Curves $>80^\circ$* uncontrolled by bracing may require operative correction with posterior instrumentation and fusion.

Natural history of this condition is usually benign except in individuals with kyphosis that was upper thoracic and $>100^\circ$ who were likely to have restrictive lung disease.

Postoperative Hyperkyphosis

This serious deformity is common following laminectomy in children for conditions such as tumors or trauma. This deformity is best prevented by decompression or exposures that save posterior elements or early posterior fusion in wide excisions in growing children.



1 Milwaukee brace management of juvenile kyphosis. The Milwaukee brace is effective in managing kyphosis. The outcome is related to the severity of the curve at the beginning of treatment. Based on Sachs et al. (1987).

Lordosis

Lordosis is anterior convex angulation of the lumbar spine. The normal range of lordosis is from about 30°–50°.

Developmental Lordosis

This developmental variation is common in the prepubescent child [1]. Parents are concerned. The deformity is flexible, the screening examination normal. Radiographs are not necessary. Resolution occurs with growth.

Functional Hyperlordosis

This deformity is functional, a compensation for fixed deformity above or below the lumbosacral level.

Hyperkyphosis is the primary deformity and the hyperlordosis is compensatory. This compensatory deformity remains flexible and this flexibility is demonstrated by correction of the lordosis on forward bending.

Hip flexion contracture cause a functional increase in lordosis, usually >60°. This deformity is very common in cerebral palsy. Assess with the prone extension test [2]. Lordosis is also common in children with bilateral developmental hip dislocations or coxa vara.

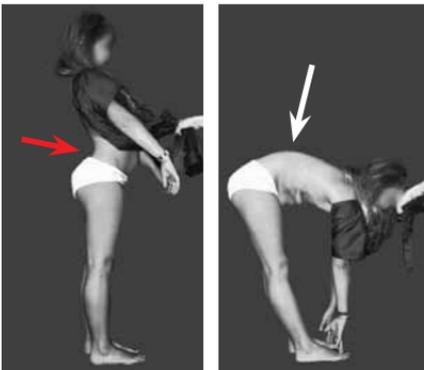
Structural Hyperlordosis or Hypolordosis

Operative procedures that arrest growth of the posterior lumbar vertebrae such as shunting or rhizotomy may result in increasing lordosis with growth.

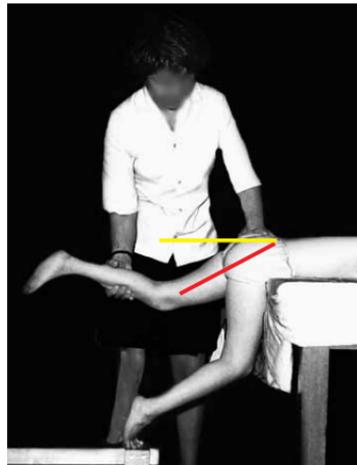
Spondyloptosis causes a secondary hypolordosis with flattening of the buttocks.

Neuromuscular disorders such as muscular dystrophy may cause hypolordosis.

Fractures with malunion may cause an increase or decrease in lordosis.



1 Physiologic lordosis of puberty. This form of lordosis (red arrow) is seen during late childhood just prior to puberty. The spine is flexible and the lordosis disappears on forward bending (white arrow).



2 Prone extension test for assessing hip flexion contracture. The thigh is gradually lifted until the pelvis starts to extend. This indicates the limit of hip extension. The contracture is the angle between the thigh (red line) and the horizontal (yellow line).

Cervical Spine

Cervical spine problems that often present with neck complaints will be covered in the next chapter.

Radiographs

Conventional radiographs remain the most valuable method of imaging the neck and shoulder.

Pseudosubluxation at C2–3 and less commonly at C3–4 is common in children under the age of 9 years [1].

ADI (atlanto-dens interval) is the distance between the odontoid and anterior arch of axis [2]. This measure is most important in children. This distance is <4–5mm in children. When the ADI >10–12 mm all ligaments have failed. Flexion-extension lateral radiographs [3 next page] demonstrate instability most graphically.

SAC (space available for the cord) is between the odontoid and the posterior arch of the axis.

Occiput–C1 relationship is often assessed by McRae and McGregor lines [2].

Special Studies

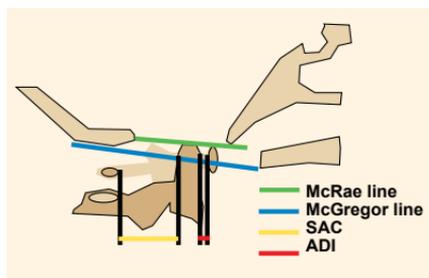
Additional imaging studies may be appropriate depending upon the evaluation. Look for associated defects. For example, order a renal ultrasound evaluation if the diagnosis of Klippel-Feil syndrome is made. In children with disproportionate dwarfism, prior to any surgical procedure requiring anesthesia, order a screening flexion–extension lateral radiograph of the cervical spine. If instability is demonstrated, special intubation techniques will prevent injury to the cervical spinal cord.

Basilar Impression

Basilar impression is a congenital or acquired deformity in which the cervical spine extends into the foramen magnum. The deformity may be congenital or secondary to osteopenia due to conditions such as rickets or osteogenesis imperfecta. This deformity may cause symptoms during adolescence.



1 Pseudosubluxation. The normal alignment of the cervical spine is usually well demonstrated by a lateral radiograph. Pseudosubluxation is common in younger children with C2 displaced forward on C3 (yellow arrow).



2 Cervical measures. These lines and measures are commonly used. The SAC, or space available for the cord (yellow), and ADI, or atlanto-dens interval (red line), are expressed in mm.

Occipital-Atlantal Instability

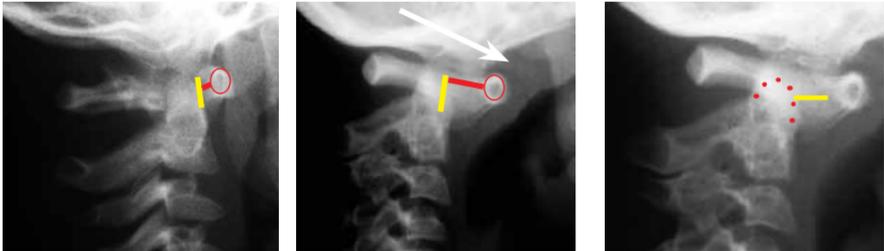
Instability at the occiput-C1 level is rare and usually due to a congenital bony defect or marked ligamentous laxity as seen in Down syndrome. Seldom is operative stabilization by fusion necessary.

Atlantoaxial Instability

Instability at the C1-C2 level is relatively common [1]. Instability is due to abnormalities of the odontoid [2 and 3] or to ligamentous laxity. Instability results from rupture or attenuation of the transverse atlantal or alar ligaments [4]. Such ligamentous deficiencies are common in Down syndrome and in rheumatoid arthritis. Instability is also common in disproportionate dwarfism. Children with these problems should avoid activities that cause cervical spine stress and have evaluation prior to being administered a general anesthetic.

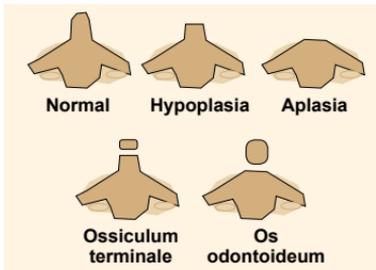
Polyarticular Juvenile Rheumatoid Arthritis

Clinical stiffness and radiographic changes in the cervical spine occur commonly in polyarticular-onset and systemic-onset disease. Neck problems are rare in pauciarticular-onset disease. Although stiffness and radiographic changes are common, children seldom complain of neck pain.

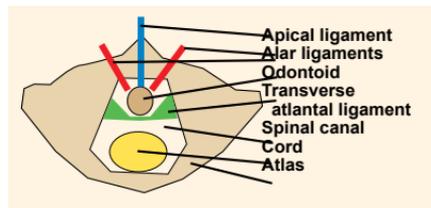


1 Neutral and flexion views of cervical spine. These studies show the relationship between arch of atlas (red ring) and the front of the odontoid (yellow line). The distance between is the ADI (red line). This relationship changes with neck flexion (right) demonstrating C1-C2 instability with the ADI increasing from 2 to 10 mm due to rupture of the transverse atlantal ligament.

2 Odontoid hypoplasia. Note the hypoplastic odontoid and the instability as demonstrated by ADI of 8 mm.



3 Odontoid types. These varied types contribute to varying degrees of instability. Based on Copley and Dormans (1998).



4 Constraining ligaments. These multiple ligaments usually prevent the odontoid from compressing the cord.

Spinal Cord Tumors

Tumors of the cervical spine are similar to those of the rest of the spine. Neurofibromatosis may cause grotesque deformity. Tumors may present with torticollis or cause clumsiness and upper extremity weakness. Some lesions are so slow growing that they remain relatively silent for many years [1].

Spine in Generalized Disorders

Many constitutional disorders such as the osteochondrodystrophies and metabolic and chromosomal abnormalities are associated with scoliosis. In these children, during each clinic visit, screen for spinal deformity.

Achondroplasia

This is a rhizomelic short-limb dwarfism, which is usually readily recognized at birth. Major and disabling spine deformities occur often in these children [2].

Stenosis of the foramen magnum causes increased hypotonia, sleep apnea, and sudden infant death syndrome. Foramen magnum decompression, duroplasty, and cervical laminectomy may be necessary if symptoms are severe.



1 Cervical spinal cord tumor. This myelogram demonstrates an extensive cervical spinal cord tumor (arrows). The boy became progressively weaker over many years before the diagnosis was made.



2 Achondroplasia. Note the kyphosis in the infant (red arrow) and the narrow lumbar canals in the adolescent (yellow arrows).

332 Spine and Pelvis / Spine in Generalized Disorders

Thoracolumbar kyphosis is common in most infants. The deformity is usually flexible. Treat rigid curves $>30^\circ$ with an orthosis. If deformity exceeds 40° after age 5, anterior and posterior fusion may be required.

Spinal stenosis is common and often becomes symptomatic in early adult life. The stenosis may be aggravated by thoracolumbar kyphosis. This deformity is usually treated in adulthood.

Diastrophic Dysplasia

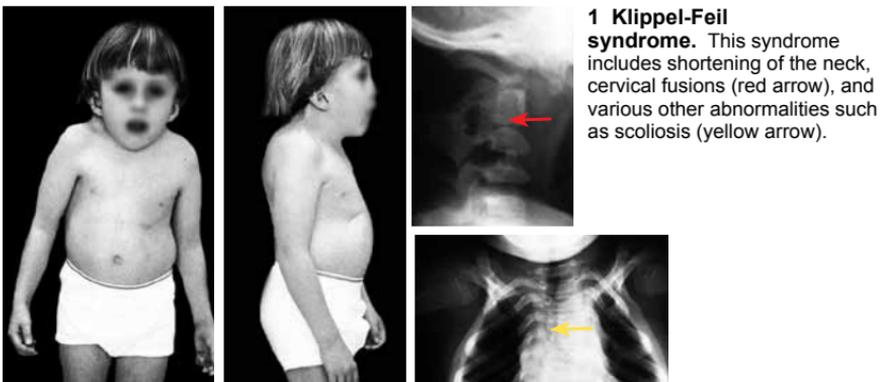
This is an autosomal recessive disorder with short-limb dwarfism. Spine deformities include generalized cervical spina bifida, cervical spine kyphosis, and thoracolumbar kyphoscoliosis. These deformities may be severe and require instrumentation and fusion.

Down Syndrome

Trisomy 21 syndrome includes characteristic faces, congenital heart disease, mental retardation, and excessive joint laxity. Upper cervical instability involving the occipito-cervical and the atlantoaxial levels develop in many children. This instability results from joint and ligamentous laxity.

Clinical manifestations of cord compromise from instability include disturbances in gait, exercise intolerance, and neck pain. Mild weakness and hyperreflexia may be found. Screen with flexion-extension radiographs by ages 5–6 years.

Management Be concerned if ADI >5 mm. Follow yearly with examination and every several years with radiographs. Some recommend fusion with ADI >10 mm.



1 Klippel-Feil syndrome. This syndrome includes shortening of the neck, cervical fusions (red arrow), and various other abnormalities such as scoliosis (yellow arrow).

Disease	Associated Disorders
Klippel-Feil Syndrome	Scoliosis Renal abnormalities Sprengel deformity Deafness Synkinesis Congenital heart disease
Disproportionate Dwarfism	C1-C2 disorders causing instability

2 Associations. Disorders about the neck are often associated with other congenital defects. Renal and cervical instability problems may not be diagnosed unless special studies are ordered.

Klippel–Feil Syndrome

Classically the syndrome includes cervical fusion, low hairline, and stiffness of the neck [1 opposite page]. The syndrome is now known to be much more generalized.

Clinical features About half have the classic findings of fusions, low hairline, and stiffness. Classify the condition by the levels of fusions. Other clinical associations include congenital scoliosis, renal anomalies, Sprengel deformity, synkinesia, congenital heart disease, and impaired hearing [2 opposite page]. Other deformities include odontoid abnormalities, occipito-cervical fusion, and basilar impression.

Evaluate carefully with full spine examination, neurological, cardiac, renal, and hearing screening. Make radiographs of the entire spine. Order a renal ultrasound. If neurological findings are present, study with MR imaging.

Management includes advising family of the risks and avoiding activities such as diving, football, and gymnastics, which place excessive loads on cervical spine. Arthrodesis of unstable segments may be required if excessive instability and neurological abnormalities are present.

Natural history Affected individuals have instability problems above and degenerative problems below the levels of fusion. Adults have disability from this syndrome.

Neurofibromatosis

Spine involvement in neurofibromatosis is common [1]. Look for bony dysplasia associated with the scoliosis. If dysplastic features are present, consider MRI or CT studies. Follow carefully as rapid progression may occur with growth.

Nondystrophic scoliosis Manage like idiopathic scoliosis.

Dystrophic scoliosis is often characterized by short angular progressive curves. Brace treatment is ineffective. Correct by combined anterior and posterior spinal fusion. Include the entire structural levels in both the fusion masses.



1 Neurofibromatosis. Curves tend to be sharp and progressive (red arrow).



Marfan Syndrome

This is an autosomal dominant disorder of connective tissue.

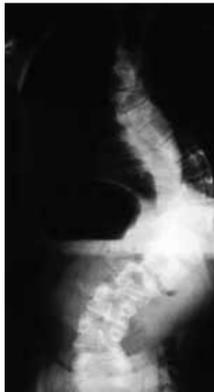
Scoliosis develops in most patients [1]. Curve patterns are often double major structural right thoracic, left lumbar. Some curves are triple. Curves usually start earlier, and are more progressive, refractory, and rigid.

- *Brace management* is less effective than for idiopathic scoliosis but is used with similar indications and protocols.
- *Operative management* is indicated for curves $>50^\circ$ with segmental fixation using sublaminar wires. Be certain to balance the spine and restore normal sagittal alignment.

Other spinal deformities have included atlantoaxial instability, spondylolysis, etc.

Morquio Syndrome

This mucopolysaccharidosis type IV is one of a spectrum of lysosomal storage diseases. The spine is normal at birth but deformities develop with growth [2]. Odontoid dysplasia is common and life-threatening. Odontoid aplasia, hypoplasia, or os odontoideum may cause instability. This instability combined with accumulation of mucopolysaccharides within the spinal canal may compromise the cord, causing sudden death or quadraplegia. Manage instability with neurological compromise first with evaluation by dynamic MRI studies. Fuse occiput to C3 or more proximal if posterior elements are adequate. Consider prophylactic stabilization if instability is severe.



1 Marfan syndrome. Note the severe right thoracolumbar curve. This curve is not improved by bracing, making instrumentation and fusion necessary.



2 Morquio syndrome. Vertebral body changes (yellow arrows) are useful in evaluation. Odontoid hypoplasia (red arrow) is a serious defect.

Osteogenesis Imperfecta

Deformity is due to osteopenia [1], and scoliosis and basilar invagination are serious problems. Bracing is inappropriate as it may cause chest and rib deformity and is unlikely to arrest progression of the curve. Operative stabilization and fusion is indicated for curves exceeding 35° – 45° . Instrument with posterior sublaminar segmental fixation and fusion. Add anterior fusion if the deformity is severe and/or associated with kyphosis.

Pseudoachondroplasia

This autosomal dominant short-limb dwarfism causes several spinal problems.

Atlantoaxial instability from odontoid deficiencies and generalized laxity is demonstrated by flexion–extension radiographs and MRI if unstable. Decompression and fusion may be required.

Thoracolumbar deformities include kyphosis and scoliosis.

Hyperlordosis may result from hip flexion contracture.

Spondyloepiphyseal Dysplasia

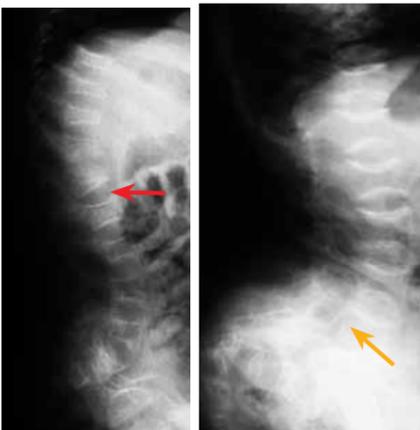
This is a group of short-trunk dwarfism with dysplasia of the spine and long bones.

Atlantoaxial instability occurs in about 40% from odontoid deficiencies, and generalized laxity is demonstrated by flexion–extension radiographs and MRI if unstable. Decompression and fusion may be required.

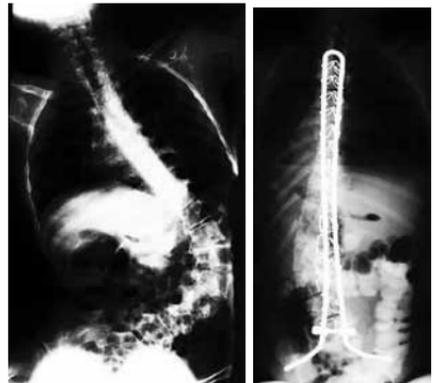
Thoracolumbar scoliosis and kyphosis are common and may cause back pain in adults. Manage as with idiopathic scoliosis.

Rett Syndrome

Rett syndrome is a progressive encephalopathy observed only in girls, who are apparently normal until 6 to 12 months of age. It is characterized by autism, dementia, ataxia, stereotypic hand movements, hyperreflexia, spasticity, seizures, and scoliosis [2]. Scoliosis is usually progressive and seldom responds to brace management. Most require posterior fusion with segmental instrumentation.



1 Osteogenesis imperfecta. Note the vertebral deformity (red arrows) and accentuated lumbar lordosis (orange arrow).



2 Rett syndrome. Deformity is severe and progressive, often requiring long fusion.

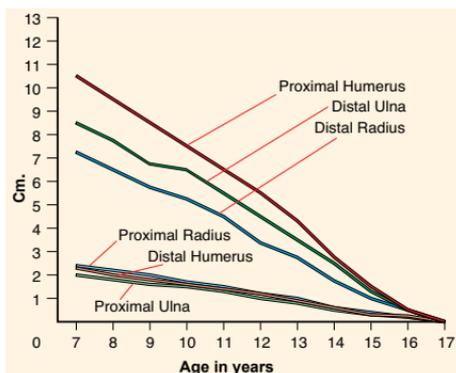
Chapter 9 – Upper Limb

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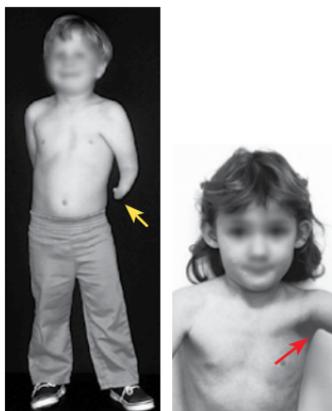
Because upper limb disorders are less common than those of the lower limbs the topic is covered in one chapter.

Development

The upper limb bud develops between the fourth and eighth fetal week. Most congenital upper limb defects have their origin during this period [2]. During the seventh week, the upper limb flexes at the shoulder and elbow and rotates around a longitudinal axis to account for the dermatomal pattern of the upper extremity. The scapula migrates caudad during development. A failure of the descent of the scapula is a feature of Sprengel deformity. Ossification of the clavicle develops from two centers. A failure of coalescence of these two centers may be the cause of the congenital pseudarthrosis of the clavicle. Upper limb growth occurs most rapidly in the proximal femoral and distal forearm epiphyses [1].



1 Growth rates for upper limb. The majority of growth of the upper limbs occurs from physes about the wrist and shoulder compared to the elbow. From Pritchett (1988).



2 Congenital anomalies. Limb deficiencies (yellow arrow) cause considerable disability. Others, such as Poland syndrome or absence of the pectoralis (red arrow), cause only a cosmetic disability.

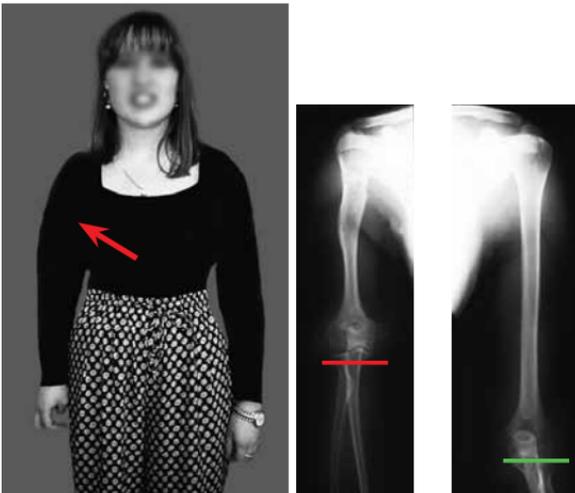
338 Upper Limb / Development

During infancy, hand function progresses in an orderly fashion [1]. Bimanual function becomes refined during the second year. Both fine and gross motor skills improve with age.

Upper limb orthopedic problems are less common than those of the lower extremity for several reasons. The upper limb is not subjected to the stresses of load bearing. Upper limb vascularity is less vulnerable than that of the lower limb. For example, the upper humeral circulation is less easily interrupted as that of the proximal femur. The function of each upper limb is more independent than that of the lower limbs. Thus, a short arm causes less functional disability than a short leg [2].

Age	Hand Function
1 Month	Hand clenched
2 Months	Opens hands
3 Months	Holds objects
5 Months	Primitive finger grasp
9 Months	Early finger pinch
12 Months	Picks up large objects
18 Months	Piles blocks
3 Years	Buttons clothing
4 Years	Can throw a ball
5 Years	Can catch a ball

1 Hand function by age. Hand function becomes progressively more skilled with advancing age.



2 Upper limb length inequality. This girl's right arm is shortened due to a cyst of the proximal humerus. Her disability is minimal.

Evaluation

The physical examination should follow the standard sequence of inspection, palpation, range of motion evaluation, and a careful neurological examination. Imaging should start with conventional radiographs.

Observation

Head and neck Observe the head and neck for abnormalities and asymmetry. The head is normally held in a vertical position by the vestibular and ocular righting mechanisms. Head tilt is common in “wryneck” or torticollis. Describe the deformity in terms of the three planes, flexion-extension, lateral head tilt, and rotation. Observe the shape of the head. Plagiocephaly is common in torticollis and includes a flattening of the malar prominence and a lowering of the position of the eye and ear on the involved side. The ipsilateral occiput is flattened.

Limbs Start with observation of the relationship of the neck and limbs. Note any asymmetry. Observe differences in spontaneous movement. Loss of movement may be due to true paralysis from a nerve injury or more likely from pseudoparalysis due to trauma or infection. The infant with a clavicular fracture or septic arthritis of the shoulder or elbow will spontaneously limit arm movement.

Observe the *carrying angle*, the alignment of the arm and forearm as viewed with the child in the anatomic position. The carrying angle is normally in 0–10° of valgus. A varus carrying angle causes the so-called *gunstock deformity*, which is usually due to malunited supracondylar fracture [3]. An increase in carrying angle is seen in Turner syndrome.

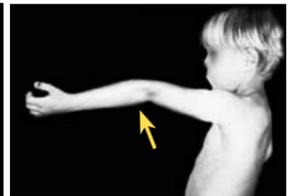
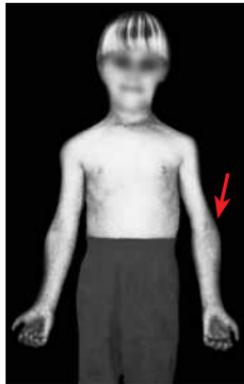
Look for asymmetry or masses and note any finger or nail abnormalities [1]. Nail dysplasia is seen in the nail-patella syndrome. Other syndromes have characteristic finger deformities such as the “hitchhiker’s” thumb in diastrophic dysplasia.



1 Nail-patella syndrome. Nail dysplasia is seen in the nail-patella syndrome.



2 Localize tenderness. Correlate tenderness with anatomic structures.



3 Cubitus varus. This child has a malunion following a supracondylar fracture. In the anatomic position, the child has a cubitus varus deformity (red arrow). Hyperextension deformity (yellow arrow) and limited elbow flexion (orange arrow) are also present.

Palpation

Palpation is most important if the child complains of pain. Exact localization of the point of maximum tenderness is very important in establishing the cause of pain. This is most feasible about the elbow, wrist [2 previous page], and hand, where the bone and joints are subcutaneous.

Range of Motion

Describe the motion of the neck in three planes. The normal child is able to flex the chin to the chest. Lateral head tilt should allow the ear to touch the shoulder. Normal head rotation allows about 90° of motion to the right and left. Assess forearm rotation with the elbow flexed to a right angle. Supination and pronation are each about 90° in the normal child.

Joint Laxity

The upper limb is readily examined to assess joint laxity. Assess the elbow, wrist, and fingers for the ability to hyperextend [1].

Pain

Pain is usually due to trauma [2], infection, or neoplasms [3]. Pain is often manifest by pseudoparalysis in the infant and young child. Localization of the site of tenderness is very helpful in narrowing the diagnostic possibilities and in deciding what should be studied radiographically. Sometimes, a bone scan is necessary, localize the problem.



1 Joint laxity. Joint laxity is commonly assessed in the upper limb.



2 Overuse syndrome shoulder. Note the widening and sclerosis adjacent to the proximal humeral epiphysis in this baseball pitcher.



3 Eosinophilic granuloma. These lesions present in unusual locations such as the scapula (red arrows). They typically cause pain.



4 chronic osteomyelitis of the clavicle. Note the swelling and sclerosis of the clavicle (arrow). A low-grade staphylococcal infection was found.

Associations

Certain deformities of the upper extremity are often associated with specific syndromes [1, 2 and 3] Examples include nail dysplasia in nail-patella syndrome and the various conditions associated with radial and ulnar deficiencies and syndactyly. Carefully examine the whole child. Look for dysmorphic features, and shortness of stature, and assess the child's general health. Inquire about medical problems in the family. Certain findings indicate the need for additional studies. For example, the finding of torticollis is an indication for a radiograph of the pelvis to rule out hip dysplasia. The finding of radial dysplasia is an indication for a hematologic and cardiac evaluation.

Unique Upper Limb Conditions

Chronic clavicular osteomyelitis The response of the clavicle to infection is unique [4 opposite page]. The clavicle becomes enlarged, sclerotic, and tender suggesting a neoplastic origin. Evaluate with CT scans or MRI to establish the primary focus of the infection. Drain, culture, and biopsy all suspicious portions of the lesion.

Reflex sympathetic dystrophy may occur in the upper extremities of children and adolescents. The condition occurs most commonly in adolescent girls who complain of pain, stiffness, and limited function. Radiographs often show osteopenia and bone scans may show normal, increased, or decreased uptake. See page 58 for a more detailed description.

Syndrome	Comment
Craniosynostosis	Form associated with radial aplasia
Fanconi anemia	Bone, skin, hematologic defects
Holt-Oram	Bone and cardiovascular defects
Ladd	Bone and craniofacial
Nagar	Bone and craniofacial
Thrombocytopenia (Tar syndrome)	Associated with radial aplasia

1 Syndromes associated with radial defects. These syndromes should be considered if radial defects are present.

Syndrome	Comment
Goltz	Bone, skin, eye, anus, retardation
Mammary aplasia	Associated with ulnar hypoplasia

2 Syndromes associated with ulnar defects. These syndromes should be considered if an ulnar defect is present.

Syndromes with syndactyly

Apert
Carpenter
Noack
Pfeifer
Poland
Summit
Waardenburg
Oculodentodigital
Orofaciodigital

3 Syndromes associated with syndactyly.

Upper Limb Deficiencies

Upper limb deficiencies may be due to malformations [1] or disruptions, such as amniotic bands from trauma or result from resections of malignant tumors.

Frequency

Limb deficiencies are most common in the lower limb and in boys [2]. Proximal transverse forearm amputations are the most common congenital upper limb deficiency.

Classification

Classify the congenital limb deficiencies as either intercalary or transverse [1 opposite page] and the segmental defect as either longitudinal or transverse. This differentiation has additional diagnostic implications. For example, nearly 90% of children with longitudinal defects of the radius have additional malformations, whereas less than a third of those with transverse defects of the radius have other similar defects.

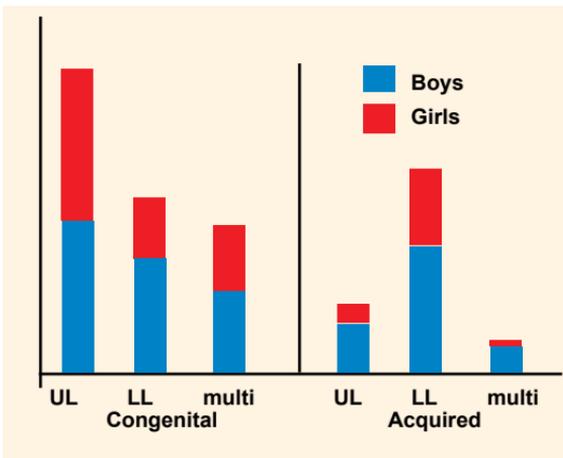
Evaluation

Although the diagnosis can usually be made by the physical examination, make radiographs to document and classify the deficiency.

Screening examination is necessary to identify other abnormalities such as radial head dislocations or radioulnar synostoses.



1 Complete phocomelia. This child has bilateral deficiencies.



2 Distribution of pediatric amputees. Distribution for girls (red) and boys (blue) for UL (upper limb), LL (lower limb), or multi (multiple levels) are shown in 1400 cases. From data of Krebs and Fishman (1984).

Family situation should be evaluated carefully. Make certain that counseling is available for parents who are having difficulty dealing with the grief and guilt common in parents of limb deficient children. Make a special effort to develop a warm and supportive relationship with the family as management is often difficult. Good rapport improves the child's compliance with treatment and the parent's acceptance of recommendations for management.

Management Principles

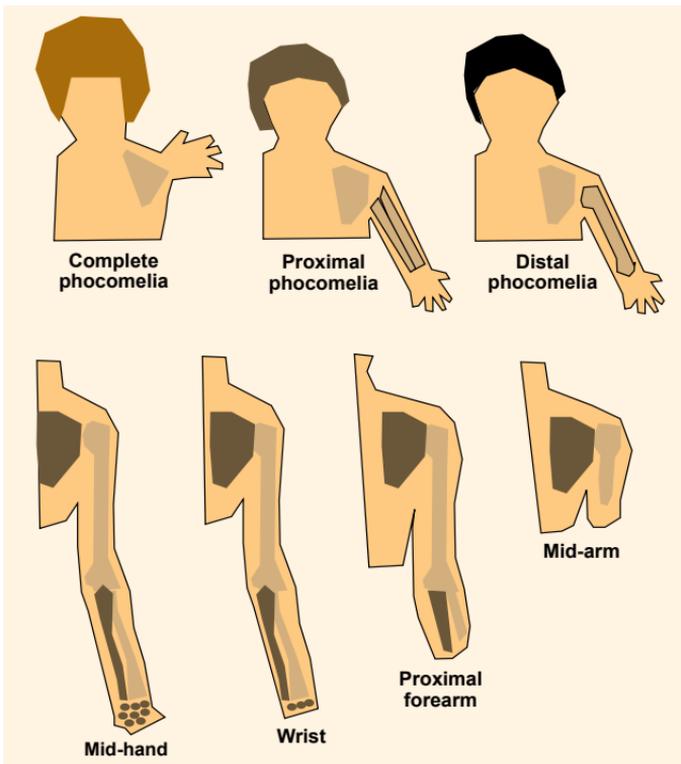
The following principles may be helpful in planning management.

Early prosthetic fitting is controversial Some physicians believe that covering the limb with a prosthesis prevents sensory feedback and slows development of bimanual function. Others recommend the fitting of a passive prosthesis between 3 and 6 months of age to promote the development of a more normal self-image by the infant.

First prosthesis is usually passive Convert to an active prosthesis based on the infant's developmental age.

Acceptance is usually less for upper than for lower limb prostheses. The lack of sensibility and fine movement control makes upper limb prostheses less useful than those for the lower limb. Children are most likely to accept an upper limb prosthesis when a specific functional need is recognized. This awareness usually occurs at about 8 years of age.

Myoelectric power is inherently attractive to parents. Because these electrically powered limbs are expensive and difficult to maintain, long-term acceptance is poorer than for the simpler, body powered prostheses.



1 Classification of upper limb deficiencies.

Congenital and acquired amputations are different. Congenital amputees are more accepting of their disability, develop techniques of compensation, and have fewer painful stumps than those with acquired deficiencies.

Modify prosthesis to facilitate activities of daily living. Make available an experienced occupational therapist to access the child's needs, and make recommendations for modifications that enhance self-care.

Family support groups are extremely valuable for both the parents and the child. Most childhood amputee clinics have ready access to these support groups and can help families make the necessary contacts.

Allow child natural adaptations Such adaptations are usually practical, effective, and energy efficient [1].

Replace prosthesis when destroyed, causes discomfort, or becomes sub-optimally functional.

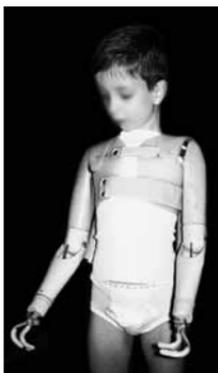
Discarding of prosthetics is most common when deficiencies are extensive, prosthetic devices are complex in design, and natural adaptations without a prosthesis are effective.

Operative Procedures

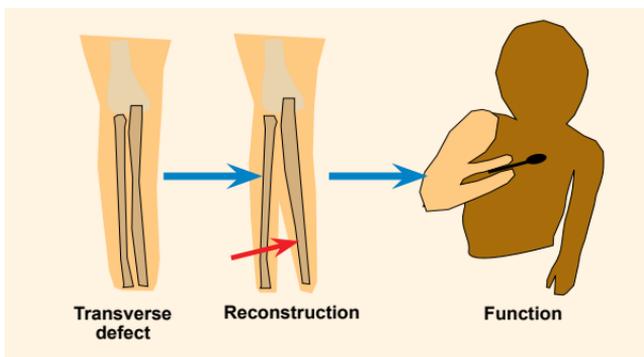
Procedures have limited indications.

Krukenberg procedure separates the radius and ulna to allow grasp with sensibility [2]. The outcome is usually functionally good but cosmetically poor. The procedure is appropriate for blind children who cannot visually position items in their prosthetic hands or hooks or for other children with special needs.

Revisions for overgrowth may be necessary in both congenital and acquired transdiaphyseal amputations [1 opposite page].



1 Upper limb amelia. These children become surprisingly proficient in using the feet to duplicate hand function. The prosthetic fitting was marginally successful.



2 Krukenberg reconstruction. This separation of the distal radius and ulna (red arrow) and repositioning of forearm muscles provides the child with a functional grasp with sensibility.

Prosthetic Options

Terminal devices options include several alternatives.

- *CAPP* (child amputee prosthetic project) includes a closing spring and a frictional resilient covering that enhances control.
- *Hooks* with elastic closures and plastic covering are durable and can be fitted with body-powered opening mechanisms.
- *Cosmetic hands* may be passive, body powered, or myoelectrically controlled.

Powering devices include several options:

- *Body power* is commonly used for both opening of a terminal device and elbow flexion [2].
- *Myoelectric power* may be provided by single or double electrodes placed over flexor or extensor muscles. Single controls are usually applied during the second year with sensors placed over extensor muscles to activate the opening device. The terminal device stays open as long as the muscle is contracted. A second sensor over the flexors may be applied about age 3 for active flexion.



1 Diaphyseal overgrowth. This is a complication of through-bone amputations.



2 Upper limb prosthesis. The artificial limb aids function, especially for tasks that are bimanual.

Torticollis

Torticollis, or *wryneck* includes a variety of conditions, that require different management [1].

Acute Torticollis

Acute torticollis, or *wryneck*, is relatively common, may occur spontaneously, follow minimal trauma, or occur after an upper respiratory infection. Why the head tilts is uncertain. The tilt may be due to muscle spasm secondary to cervical lymphadenitis or possibly due to a minor subluxation of the cervical vertebrae.

Clinical features Acute torticollis causes the head to tilt, rotate to one side [2] and become fixed. Radiographs of the cervical spine are difficult to assess because of the lateral flexion and rotation. Laboratory studies are normal.

Manage by immobilizing the neck with a folded towel and encourage rest. Early management is usually provided by the primary care physician. In most children, acute torticollis resolves within 24 hours. If the deformity persists longer than 24–48 hours, be more concerned and manage as rotatory displacement.

Rotatory Displacement

Persistence of acute torticollis is called *rotatory displacement* or *rotatory subluxation*. Treat rotatory displacement early to avoid permanent fixation and residual deformity.

Evaluate Image with conventional and dynamic CT scans. Look for a loss of the ability to rotate the head fully in both directions. In addition, note the relationship between C1 and C2. Fixation of this relationship throughout the arc of rotation is consistent with the diagnosis of rotatory displacement.

Manage First apply traction. If early, head-halter traction is appropriate. Later, halo traction may be required. For displacements that persist beyond a month, repositioning and fusion of C1–2 may be necessary [1 opposite page].

Category	Comments
Muscular torticollis	Most common
Acute torticollis	Acute, resolves
Occipital cervical bony defects	Hemivertebrae
Miscellaneous	Neurogenic—tumors inflammatory Traumatic Ocular: strabismus Hysterical Idiopathic: rotatory displacement



1 Causes of torticollis. The causes are many, but the vast majority of torticollis are due to disorders listed in the top three categories.

2 Acute torticollis. This form of torticollis develops suddenly in a previously normal child. Usually the deformity resolves spontaneously in a day or two.

Chronic Nonmuscular Torticollis

About 20% of children with chronic torticollis are due to nonmuscular causes. Radiographs may show conditions such as Klippel-Feil anomaly or hemivertebrae. If radiographs are negative and the sternocleidomastoid muscle not contracted, consider an ocular etiology. Refer to an ophthalmologist for evaluation. Consider the other conditions that may cause torticollis, such as neonatal brachial plexus palsies and spinal cord tumors.

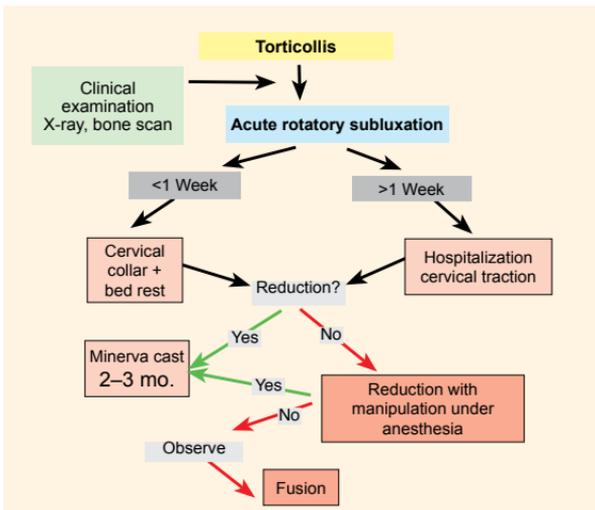
Muscular Torticollis

Muscular torticollis is relatively common and presents in two age groups.

Infantile muscular torticollis The infant [1 next page] is first seen because of a head tilt. Sometimes a history of a breech delivery is given and a firm tumor of the sternocleidomastoid muscle is palpated. Usually only a head tilt and limited neck motion due to a contracture of the muscle are found. Plagiocephaly (asymmetrical head) may be present [2 this page and 2 next page].

Be certain to rule out developmental hip dysplasia. Even if the hip examination is negative, evaluate the hip by either ultrasound if the infant is seen in the neonatal period or by a single AP radiograph of the pelvis if the infant is older than about 10 weeks of age.

Infantile torticollis resolves spontaneously in about 90% of cases. The value of physical therapy by stretching is uncertain [1 next page]. Of those that persist, operative correction is may be necessary. Delay correction until about 3 years of age. Rarely plagiocephaly persists and is a cosmetic problem (Fig. 9.28).



1 Management of rotatory subluxation. Based on Phillips and Hensinger (1989).



2 Muscular torticollis and plagiocephaly. The mass (red arrow) develops in early infancy and disappears spontaneously over a period of several months. The plagiocephaly (blue arrows) may persist longer.

Juvenile muscular torticollis Sometimes muscular torticollis appears to develop during childhood [3]. In this juvenile type, usually both heads of the muscles are contracted causing the head tilt and limiting neck motion. Usually this type of torticollis is permanent and often requires operative correction.

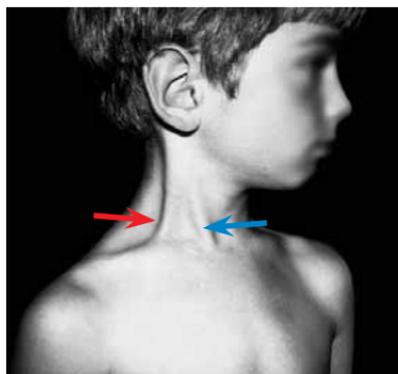
Operative correction Bipolar release [4] is the most effective procedure for correction of both infantile and juvenile forms of muscular torticollis.



1 Muscular Torticollis. This is the most common neck problem in childhood. Torticollis is usually seen first in the infant (left). Some advocate treatment by stretching (right), but its value is uncertain.



2 Plagiocephaly torticollis. Cranial deformity is readily shown by CT scans. 3D reconstructions provide graphic documentation of the extent of the deformity.



3 Sternocleidomastoid contracture. Both the clavicular origin (red arrow) and sternal origin (blue arrow) are contracted.



4 Torticollis release. This 9-year-old with torticollis (left) had a bipolar release (arrows). Note the improvement in head tilt and neck motion followed surgery (right).

Sprengel Deformity

Sprengel deformity is a congenital elevation of the scapula [1]. The deformity results from a failure of migration of mesenchyme during the second fetal month.

Clinical Features

The deformity varies in severity [1 next page], is usually unilateral, and is associated with other abnormalities in 70% of cases. These associated abnormalities include absent or hypoplastic parascapular musculature, abnormalities in the cervicothoracic vertebrae or thoracic rib cage, presence of an omovertebral bone, limited shoulder abduction and multidirectional shoulder instability. Because of restricted scapulothoracic motion, most shoulder motion occurs through the glenohumeral joint.

Management

When the deformity is mild, correction is not appropriate because the operative scar is often more unsightly than the deformity. For moderate deformity, excise the superior pole of the scapula. For severe deformity repositioning of the scapula is necessary. This repositioning requires an extensive soft tissue release, caudad repositioning of the scapula and sometimes excision of the superior portion of the scapula. Perform the correction in early childhood when the scapula is most mobile. This mobility allows maximum correction with the least risk of complications. For correction, several procedures have been described. The Woodward procedure is most widely used.



1 Sprengel deformity. Congenital elevation of the scapula causes a shoulder deformity (red arrows) that cannot be hidden by clothing. Some loss of abduction is a common mild disability (yellow arrow).

350 Upper Limb / Sprengel Deformity

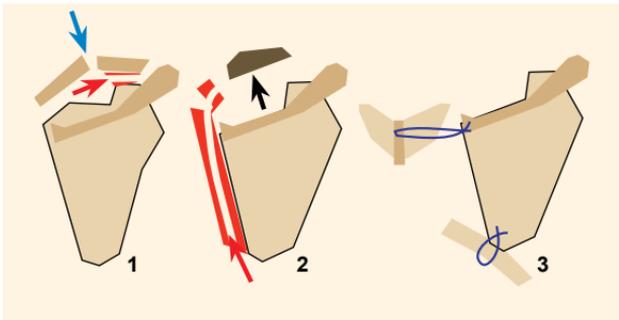
Green procedure All muscular attachments to the scapula are freed, the omovertebral band is divided, and the scapula is rotated, moved caudad to a more normal position and sutured into a pocket of the latissimus dorsi. In the original description, traction was applied by a wire attached to the scapula to hold it in the corrected position.

Kliscic procedure includes performing an osteotomy of the clavicle, extensive muscle releases, excision of the superior scapular margin, and securing the repositioned scapula with sutures to a vertebral spinous process and rib with absorbable sutures [2].

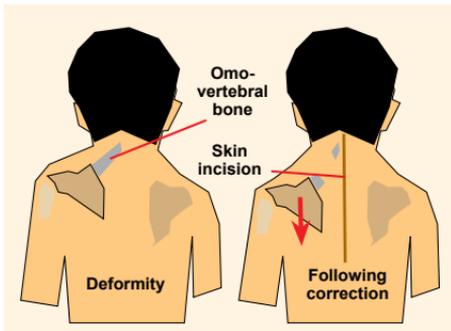
Woodward procedure exposure is made through a midline incision, the origins of the trapezius and rhomboid muscles are released, the omovertebral bone is excised, and the scapula repositioned [3]. Modifications include excising the superior and medial margins of the scapula.



1 Spectrum of severity. Disability is proportional to the deformity. Shoulder elevation may be severe (red arrow), moderate (yellow arrow), or mild (green arrow).



2 Kliscic procedure. 1. The clavicle is divided (blue arrow). 2. The superior pole of the scapula is removed (black arrow) and the muscles are released (red arrows). 3. The repositioned scapula is sutured to the rib and transverse processes (blue sutures). Based on Kliscic (1981).



3 Woodward procedure. Through a midline incision, excise the omovertebral bone, release the soft tissue attachments, and reposition the scapula at a more distal location.

Neonatal Brachial Plexus Palsy

Neonatal palsy [1] is a traction injury to the brachial plexus that usually occurs during delivery. Risk factors include shoulder or fetal dystocia, obesity, and prolonged labor. The incidence of these injuries has declined due to improved obstetrical practices.

Classification

Severity is determined by the nature and extent of the lesion [2]. Mild lesions are stretch injuries of C5–C6. Severe injuries involve avulsion of nerve roots over multiple levels down to T1. Classically three types of injuries have been described [1 next page].

Natural History

Recovery depends upon severity. Overall, up to 90% spontaneously completely resolve during the first year. Most improvement occurring in the first 3 months. Failure of recovery of elbow flexion by 3 months or more accurately, a failure of recovery of elbow flexion, wrist, and digital extension by 4 months correlates with a poorer prognosis. Common residual disabilities include loss of external rotation and abduction [2 next page].

Management

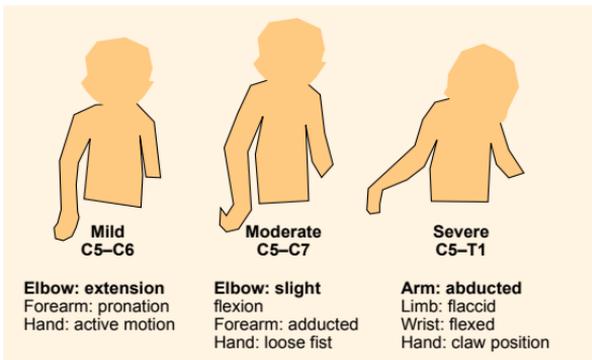
Management may be divided into 4 types.

Range of motion may be helpful to maintain joint mobility. Instruct parents how to gently range joints with each diaper change.

Brachial plexus repair is controversial. Consider repair for infants with early evidence of severe injuries and those without recovery of elbow flexion by 3 months. Evaluate by EMG, nerve conduction, myelography, and CT scans. Root avulsions cannot be repaired. More distal lesions may be repaired or grafted using microsurgical techniques. Results of repairs are unpredictable and should not compromise later orthopedic reconstructive procedures.



1 Neonatal palsies. These infants show moderate (yellow arrow) and severe (red arrow) palsies.



2 Neonatal palsy classification of severity. Early classification is based on physical position and function. Based on Lidell-Iwan, et al. (1996).

352 Upper Limb / Neonatal Brachial Plexus Palsy

Muscle procedures are indicated for children with disabling adduction and internal rotation contractures. The most common procedure is the Sever-L'Episcopo transfer. This procedure includes release of the pectoralis major, subscapularis, and joint capsule if contracted. The teres major and latissimus dorsi tendons are transferred from the anteromedial to the posterolateral aspect of the humerus. Axillary nerve palsy is a potential complication. This procedure is usually performed in early childhood.

Rotational humeral osteotomy is indicated for an internal rotation deformity that limits function. Delay the procedure until mid or late childhood. Rotate the humerus to provide about equal internal and external rotation. Results are predictable, correction is usually permanent, and complications are infrequent.

Should Dislocation

Dislocation of the glenohumeral joint may develop in children with brachial plexus birth palsy as early as 3 months of age. Manage by release of the insertions of the pectoralis major, latissimus dorsi, and teres major followed by a closed reduction of the glenohumeral joint. Transfer the latissimus dorsi and the teres major to the rotator cuff.

Level	Type	Name
C4, C5, C6	Type I	Erb's palsy
Entire plexus	Type II	Erb-Duchenne-Klumpke
C7, T1	Type III	Klumpke palsy

1 Classical typing. These palsies are classified by level.



2 Residual deformities in neonatal palsy. Medial rotation (red arrow) and limited abduction (yellow arrow) are typical deformities that limit function.

Shoulder

Congenital Pseudarthrosis of the Clavicle

Congenital pseudarthrosis of the clavicle is a rare defect of uncertain cause. The defect may be secondary to a failure of coalescence of the two ossification centers of the clavicle or to erosion of the clavicle from pulsation of the subclavian artery. The lesion practically always occurs on the right side.

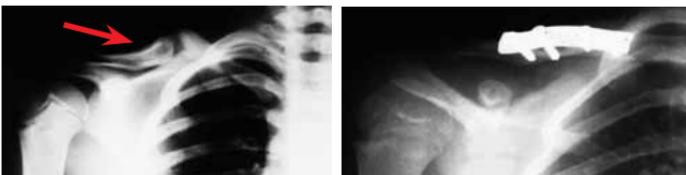
Clinical features The pseudarthrosis produces a prominence over the clavicle [1], and narrowing and slight weakness of the shoulder. Radiographs show a midclavicular defect. Rarely thoracic outlet syndrome is an associated problem. Long-term studies show little functional but some cosmetic disability.

Management Reasonable management options include either accepting or repairing the deformity [2]. Operative repair eliminates the prominence and improves shoulder symmetry but does leave a surgical scar. This scar may be mimized by positioning the incision below the clavicle, limiting its length, and utilizing a subcuticular closure technique.

- *Early operative correction* may be performed in infancy or early childhood by resecting the sclerotic bone ends, careful dissection and preservation of the periosteal sleeve to maintain continuity, and approximating the bone ends using heavy absorbable sutures. No internal fixation or grafting is necessary. Remodeling corrects bony irregularity.
- *Late operative correction* in mid or late childhood usually requires plate fixation and autogenous bone grafting to promote union.



1 Congenital pseudarthrosis of the clavicle. This pseudarthrosis produced an unsightly lump on the shoulder (red arrow). Note the gap between the ends of the pseudarthrosis (yellow arrows). The defect was operatively corrected.



2 Operative repair of congenital pseudarthrosis of the clavicle. The repair involved excision, plating, and grafting.

Shoulder Dislocation or Subluxation

Congenital dislocations are very rare. Developmental dislocations may occur with neonatal brachial plexus palsies, or develop spontaneously during childhood. Most dislocations are traumatic.

Traumatic anterior dislocation in children usually recurs regardless of the initial treatment. Prepare the patient and parent for the probability that operative repair will be required.

Recurrent posterior dislocation may occur with minimal trauma or develop spontaneously. If the deformity causes a significant disability, operative stabilization with a bone block or glenoplasty and capsulorrhaphy may be necessary.

Habitual dislocation occurs in loose-jointed older children or adolescents. One or both shoulders can be voluntarily subluxate or dislocate [2]. Management is difficult. Shoulder exercise, avoidance of voluntary displacement, and counseling maybe helpful. The child should be helped to find a more appropriate method of getting attention. Resolution usually occurs with time. Operative procedures may be necessary for persisting deformity but recurrence poses a significant problem. The condition causes little long-term disability.

Poland Syndrome

This syndrome includes absence of the pectoralis major [1] and usually finger or forearm abnormalities. The disability is cosmetic, and chest-wall and breast reconstructions is often appropriate.

Cleidocranial Dysostosis

This rare congenital defect is transmitted as a dominant trait. The clavicles are so mobile that they may be approximated [1 opposite page]. In others the clavicles are simply dysplastic. Associated findings include a large head with a small face, drooping shoulders, coxa vara, narrow chest, and sometimes recurrent shoulder or elbow dislocations. Disability is minimal.



1 Poland syndrome. Note the deficiency in the pectoralis major muscle (arrow).



2 Habitual posterior dislocation of the shoulder. This girl is able to voluntarily dislocate her right shoulder (red arrows). Voluntary reduction occurs easily (yellow arrow).

Elbow

Panner Disease

This is an osteochondritis of the capitellum that develops spontaneously during late childhood [2]. Clinical features include elbow pain, limitation of motion, and tenderness over the capitellum. Over a period of months, the capitellum fragments and then spontaneously reossifies. The process is usually benign and complete recovery occurs with time. Treatment is seldom necessary.

Adolescent Capitellar Osteochondritis Dissecans

This avascular necrosis of the capitellum is often secondary to repetitive trauma and causes articular damage and often residual long-term disability [3].

Clinical findings include a history of stiffness, pain, and catching or locking. Examination usually demonstrates a decreased elbow motion and lateral tenderness. Radiography often shows loose articular fragments, flattening of the humeral capitellum, and subchondral cysts. MRI and arthroscopic examination may be helpful. Additional lesions of the radial head may be present.

Management depends upon the clinical findings. Remove loose fragments. The value of debridement and drilling is uncertain. Limit activities until healing is complete.

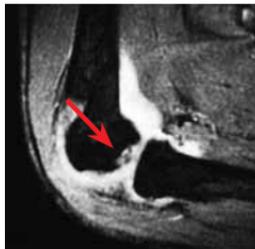
Prognosis Disability is common in adult life with about half showing joint stiffness and degenerative changes, and enlargement of the radial head.



1 Cleidocranial dysostosis. Absent clavicles allow this boy's shoulders to narrow.



2 Panner disease. This 8-year-old has pain and tenderness over the capitellum.



3 Osteochondritis of the capitellum. When this occurs in the adolescent, the problem is more serious and may cause limitation of motion. Removal of loose bodies is commonly necessary.

Recurrent Elbow Dislocation

Recurrent dislocations may be secondary to congenital hyperlaxity as occurs in Ehlers Danlos syndrome, a sequela from nonunion of a medial epicondylar fracture or due to residual instability from a previous dislocation. Evaluate with radiography, MRI, and possibly arthroscopy. Tailor operative repair based on the pathology.

Elbow Flexion Contracture

Contractures may be congenital as occurs with some forms of arthrogryposis, or acquired from burn contractures or elbow trauma with articular damage. Individualize management. Posttraumatic contractures may be improved by operative release. Release the anterior and posterior capsules and remove obstacles to motion, and provide a postoperative range of motion and splinting program.

Cubitus Varus Deformity

This deformity is usually due to a malunited supracondylar fracture. When severe, correct with a valgus osteotomy of the distal humerus [1].



1 Cubitus varus with operative correction. This girl has cubitus varus (red arrows) secondary to malunion from a supracondylar fracture. This was corrected with a valgus osteotomy (yellow arrow).



2 Congenital dislocation of the radial head. This 15-year-old boy has disability from this prominent radial head. The radial head was excised.

Forearm

Nontraumatic Radial Head Dislocation or Subluxation

The radial head dislocations may be congenital or develop gradually during infancy and childhood. Congenital dislocations are often associated with other defects.

Subluxation or dislocation limits forearm rotation and produces a palpable prominence over the displaced radial head [2 opposite page]. Once dislocated the radial head becomes progressively more prominent with growth. The radial head dislocation causes shortening of the radial side of the forearm, making the ulna more prominent at the wrist. Differentiate congenital and traumatic dislocations [1] as the management is different. Posterior dislocations are nearly always congenital. Congenital anterior dislocations are usually associated with other congenital defects.

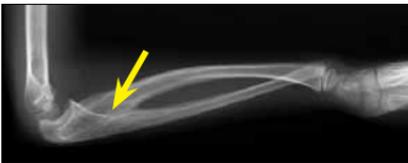
Reduction of the nontraumatic radial head displacement has not been successful. If the radial head becomes unacceptably prominent or painful, excision may be necessary. When possible delay excision until the end of growth. Excision may improve motion and reduce discomfort.

Radioulnar Synostosis

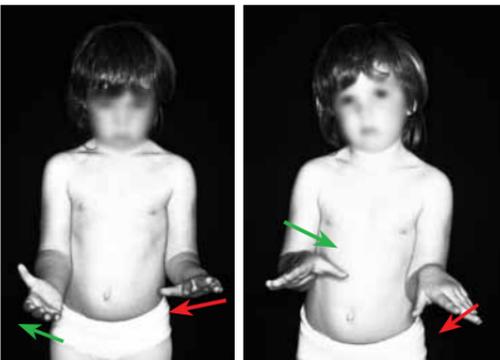
Radioulnar synostosis is usually congenital and occurs in the proximal forearm [2]. Synostoses may be unilateral or bilateral, complete or incomplete, and are usually an isolated defect. Rarely synostoses are familial. Sometimes synostoses develop after fractures of the proximal forearm.

Feature	Traumatic	Congenital
Trauma history	Yes	No
Associated defects	No	Often
Direction	Anterior	Posterior
Radial head	Concave end	Round end
Capitellum	Normal	Hypoplastic
Ulna	Normal	Convex

1 Differentiating congenital and traumatic radial head dislocation. The differentiation can usually be made by the radiographic appearance of the elbow.



2 Radioulnar synostosis limiting forearm rotation. The left forearm is fixed in pronation (red arrows). The right forearm rotates freely (green arrows). The synostosis is proximal in location (yellow arrow).



Evaluation The defect may be found during infancy if a screening examination is performed. More often, the defect becomes apparent during early childhood when the loss of forearm rotation is recognized. The position of forearm rotation is variable and determines the degree of disability.

Management is determined by the position of fixation. If rotation is fixed in a relatively neutral position, no treatment is required.

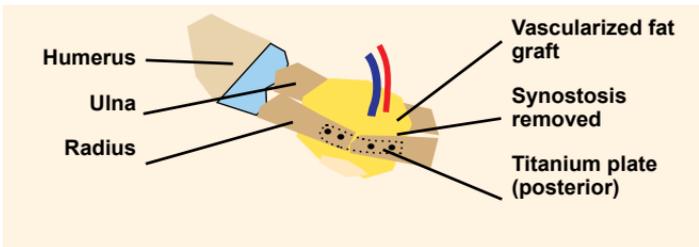
- *Rotational osteotomy* is indicated if the forearm is fixed in more than about 45° of pronation or supination. Correct by a distal osteoclastis or subperiosteal osteotomy and immobilize in a cast with the forearm positioned in neutral or slight pronation.
- *Vascularized fat graft* has been reported to be successful as an interposition tissue following resection of the synostoses in preventing recurrence [1]. Most other techniques of repair have been unsuccessful.

Osteochondromata

Multiple osteochondromata often involve the forearm and often occur at the wrist [2].

Clinical features Distal lesions of the ulna cause progressive shortening, bowing of the radius and/or ulna, increased ulnar tilt of the distal radial epiphysis, ulnar deviation of the hand, progressive ulnarward translocation of the carpus, and subluxation/dislocation of the proximal radial head.

Management is controversial. Surveys of adults suggest that the deformity causes little disability and is well accepted. Others recommend early excision of the lesions and ulnar lengthening. Be aware that operative gain in motion is usually minimal, recurrence is common, and repeated procedures are often necessary.



1 Mobilization of radioulnar synostosis. This technique utilizes a free vascularized fascio-fat graft. Based on Kanaya and Ibaraki (1998).



2 Osteochondromata of the forearm. These are the typical deformities of the typical distal ulnar lesions.

Wrist

Wrist pain

Wrist pain may be due to a number of conditions that are unique to the wrist. These include nonunion of the radial styloid, unrecognized fractures or idiopathic avascular necrosis of the navicular, and overuse syndromes. Most gymnasts experience wrist pain due to overuse. Gymnasts most at risk are older children who are new and participate many hours per week.

Madelung Deformity

Madelung deformity is a defect in the volar, ulnar portion of the distal radial physis, producing a progressive deformity.

Clinical features The deformity when associated with short stature is often inherited as an autosomal dominant defect. Most cases are idiopathic and is most common in girls, usually first noticed during mid to late childhood [1]. The physal defect causes radial shortening and a tilt of the epiphysis. The deformity is often bilateral but asymmetrical in severity.

Management If the deformity is mild, no treatment is necessary. For more severe deformities, operative correction is necessary to prevent progressive deformity characterized by a decreased radioulnar angle, lunate subluxation, and various degrees of dorsal subluxation of the distal ulnar.

- *During growth* consider closure of the distal ulnar growth plate and resection and fat interposition of the radial physal bridge.
- *End of growth* consider a corrective osteotomy of the radius and shortening of the ulna. This often improves grip strength, increase range of motion, and reduces pain.



1 Madelung deformity. This is more common in girls and produces limited wrist motion and prominence of the distal ulna (red arrow) and radial shortening (yellow arrow).

2 Kienboeck disease. This 7-year-old gymnast developed wrist pain. The sclerosis of the lunate is obvious (arrow). Usually this disease occurs in adolescence, producing pain and stiffness.

Kienböck Disease

Osteochondritis of the lunate is rare in children. It is thought to be due to repeated minor trauma together with negative ulnar variance (short ulna). The condition sometimes occurs in children with tension athetosis type of cerebral palsy, which combines increased tone and excessive motion. Clinical findings include localized pain and tenderness over the lunate, and typical radiographic features [2 previous page]. Manage with rest, NSAIDs, and time. Rarely, symptoms persist making necessary radial shortening to reduce stress on the lunate.

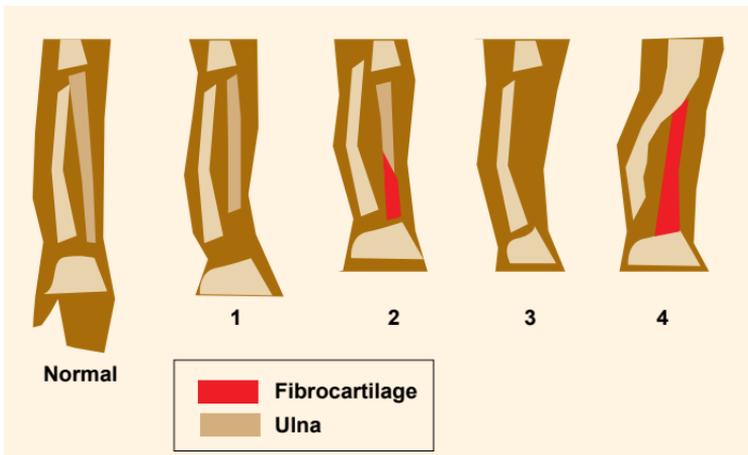
Wrist Ganglia

These cystic lesions occur in adjacency to the joints or tendon sheaths. They are most common on the dorsum of the wrist [1]. Ganglia may cause discomfort and an annoying prominence.

Manage First confirm the diagnosis by translumination or ultrasonography. One alternative is to aspirate the cyst. This confirms the diagnosis but temporarily resolves the symptoms as the cyst usually recurs. If the family and child are patient, allow the cyst to resolve with time. Most cysts will resolve spontaneously. Excise persistent or symptomatic cysts. Excision, especially of the volar ganglia, may be complex, and involve much deeper structures than one might expect. Recurrence is common after all methods of treatment.



1 Ganglia. These are the less common volar ganglia (blue arrows).



2 Types of ulnar deficiencies.

Hand

Ulnar Dysplasia

The ulnar dysplasia includes an absence or hypoplasia of the ulna [2 opposite page]. Often the radius is shortened and bowed and the ulna may be fused with the humerus. Finger deformities are common. Look for associated problems. Often function is preserved and reconstruction is not necessary. Tailor management to address the pattern of deformity. Sometimes resection of the anlage is necessary to prevent progression of radial bowing.

Radial Club Hand

Radial club hand includes an absence or hypoplasia of the radius and associated musculature producing a lateral deviation of the hand [1]. The deficiency may be isolated or part of a generalized skeletal dysplasia. Look for other problems by performing a screening evaluation giving special attention to the hematologic, urinary, and cardiac systems.

Management depends upon the severity of the deformity and the presence of associated defects. Mild hypoplasia may not require any treatment. Treat complete radial aplasia during the first year by first splinting or casting the hand and forearm to stretch the soft tissue contracture followed by operative correction. Operative correction usually include soft tissue release or lengthening and centralization of ulna on the carpals. Plan follow-up throughout infancy and childhood.



1 Radial dysplasia. This deformity causes a serious cosmetic as well as functional disability. The hand shifts to the radial side (arrow).



2 Cleft hand deformities. The child also had deformities of the feet. The condition was familial.

Cleft Hand Deformity

The cleft hand or central deficiency is a rare inherited defect that often affects both the hands [2 opposite page] and the feet. Operative reconstruction improves both function and appearance.

Hand Tumors

Primary hand tumors in children are extremely varied [1, 2 and 3] and are managed as described in Chapter 13.

Dysplasia Epiphysialis Hemimelica

Dysplasia epiphysialis hemimelica (DEH), or Trevor disease, is a rare developmental disorder causing asymmetrical epiphyseal cartilage overgrowth with accessory epiphyseal ossification centers [4]. This overgrowth causes deformity and swelling. DEH of the hand is often confused with other tumors. Manage by excising the lesions and correct secondary deformities by osteotomies. Expect recurrence as long as the child is still growing. These lesions are not premalignant.

Primary bone tumors
Osteochondromata
Enchondromas
Osteoid osteoma
Aneurysmal bone cysts
Ewing sarcoma
Epidermoid cysts
Bone cysts
Osteoblastoma
Miscellaneous types

Soft tissue tumors
Ganglia
Digital fibromas
Blood and lymph vessels
Giant-cell tumors tendon sheath
Tumeral calcinosis
Soft tissue chondroma
Synovial osteochondromatosis
Aggressive fibromatosis
Embryonal rhabdomyosarcoma

1 Primary bone hand tumors in children. Based on Kozlowski, et al. (1988).

2 Primary soft tissue hand tumors in children. Based on data from Azouz, et al. (1989).



3 Osteochondroma. This lesion is large and interferes with function.



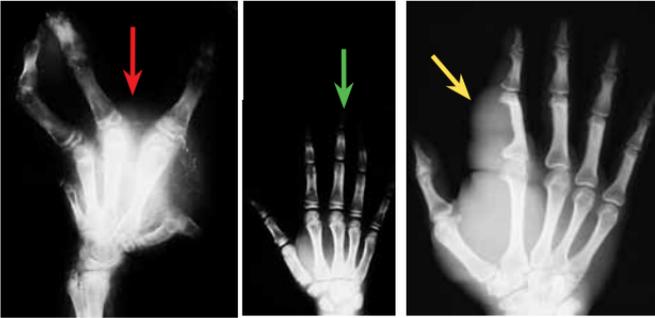
4 Dysplasia epiphysialis hemimelica. Intraarticular osteochondral tumors distort joint surfaces, causing deformity and swelling (red arrows). These lesions are often confused with other disorders.

Macroductyly

Overgrowth of the hand [1] may be secondary to a variety of disorders [2] or occur as a primary problem. In primary macroductyly, tissues are normal but growth is accelerated. This overgrowth may be greater, the same, or less than the rest of the limb. In secondary macroductyly, tissues are abnormal. In some, the tissue type is obvious as with hemangiomas. In others, MRI and biopsies may be required to establish the diagnosis. Management is often difficult. Operative procedures include soft tissue resections, epiphysiodesis, shortening osteotomies, or bone resections and sometimes amputation of digits. Recurrence is distressingly frequent.

Arthritis

Juvenile arthritis [3], autoimmune disorders, leukemia, sickle cell disease, child abuse, and infections are causes of joint problems in the child's hand. Juvenile arthritis is most common and often includes a loss of wrist extension and radial deviation of the fingers in the metacarpophalangeal joints. Ulnar shortening and involvement of the distal interphalangeal joints may occur late in the disease. See page 60 for a discussion of arthritis.

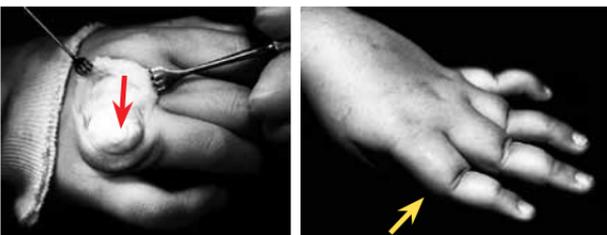


1 Hand hypertrophy. Note the massive hypertrophy of the left hand (red arrow) as compared with the normal hand (green arrow). In another child hypertrophy is associated with neural hamartoma, producing soft tissue hyperplasia (yellow arrow).

Category	Disorder
Primary	Proportionate growth pattern Accelerated growth
Secondary	Hemangiomas Lymphangioma Neurofibromatosis Fibrous dysplasia Lipoma Desmoid tumor Fibromatous hamartoma of nerve

2 Classification of macroductyly.

Secondary overgrowth may involve a variety of tissues.



3 Hand deformities in JRA.

Deformity may be due to synovitis (red arrow) or muscle imbalance (yellow arrow). Most may be prevented by systemic and local treatment.

Fingers

Separate finger deformities into categories [1]. Often finger deformities are genetic, and the genes responsible for preaxial polydactyly, cleft hand and foot malformations, synpolydactyly, and types of brachydactyly have been recently identified.

Hypoplasia of Digits

Types of hypoplasia of digits are numerous and varied. This makes it necessary to individualize management with the objective of improving function, sensibility, and mobility. Digital reconstruction by toe to finger transplants or elongation of digits are examples of effective reconstructive procedures.

Polydactyly

Polydactyly is one of the most common congenital deformities [2 and 3]. Some forms of polydactyly are inherited. For example, duplications of the middle and little fingers are often inherited as autosomal dominant defects. Duplications of the index finger, central rays, and small digit each have unique characteristic features and associations. Complex anomalies such as the mirror hand and pentadactyly are also part of the polydactyly spectrum. Duplications are classified as preaxial (thumb), central, and postaxial types. Remove simple duplications in early infancy. Delay correction of complex duplications until late in the first year.

Deformity	Description
Hypoplasia	Small digits
Polydactyly	Too many digits
Syndactyly	Fusion of digits
Finger deformities	
Camptodactyly	Flexion contracture of the IP joint
Clinodactyly	Radial or ulnar angulation
Delta phalanx	Interposed delta-shaped ossicle
Kirner deformity	Progressive palmar, radial deviation of the distal phalanx of the little finger

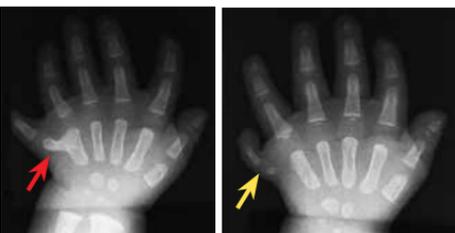
1 Classification of finger deformities.

Deformities occur in the transverse or sagittal planes.

Category	Classification
Simple	Soft tissue digit
Complex	Bony duplications
Complete	Entire digit with metacarpals



2 Classification of duplications. A simple method of classification is described. A simple duplication is shown (arrow).



3 Polydactyly. The complex polydactyly (red arrow) poses a much greater problem to repair when compared with the simple polydactyly (yellow arrow).

Syndactyly

Syndactyly [1] is a common deformity. The syndactyly may be complete, or partial, and described as simple, if only the soft tissues are involved, or complex if bones are fused [2]. Syndactyly is most common between the middle and ring fingers. Syndactyly is seen in Apert, constriction band, and Poland syndromes. Correct by operative separation and full thickness skin grafting [1 next page].

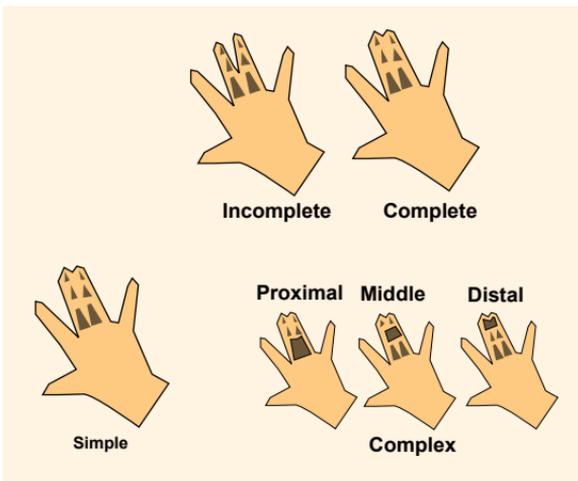
Bent Fingers

Bent or curved fingers occur in the frontal or sagittal planes. Except for camptodactyly, all have an underlying bony deformity. Many are associated with a variety of generalized disorders.

Camptodactyly is the common flexion deformity of the fingers [1 on page 367] that is divided into infantile and adolescent forms. The deformity is often progressive. Disability is usually mild. Treatment includes splinting, and rarely, operative correction.

Clinodactyly is a radial deviation of fingers, is often bilateral, and most commonly affects the little fingers [2 on page 367]. The deformity is usually considered as a variation of normal, causes no disability, and seldom requires treatment. Rarely the deformity is severe enough to require correction. Correct by performing a wedge osteotomy of the phalanx. Delay correction until late childhood or early adolescence to reduce the risk of recurrent deformity.

Delta phalanx is an abnormal interposed triangular ossicle [3 on page 367] in the finger producing an angulatory deformity. Correct by osteotomy, or resection of the bridging physis. Following physal resection, fill the defect with autogenous fat to prevent recurrence.



1 Classification of syndactyly.

Syndactyly may be complete or incomplete, simple or complex.



2 Syndactyly. Syndactyly is readily identified. Radiographs are useful in determining the degree of bony involvement.

Kirner deformity This is a rare, progressive curving of the terminal phalanx of the little finger of unknown etiology. The deformity is characteristic in appearance, usually causes little disability, and seldom requires treatment. If the lesion is painful, immobilize the finger with a splint. Rarely the deformity is severe enough to require correction by a phalangeal osteotomy [3 opposite page].

Brachydactyly

Shortening of the metacarpal or fingers is often inherited as an autosomal trait or may be associated with a variety of conditions such as Poland, Holt-Oram, Cornelia de Lange, or Silver syndromes. Rarely finger lengthening procedures are appropriate.

Symphalangism

Fusions may involve the proximal or distal interphalangeal joints. The deformities are often congenital, inherited, and varied in pattern. Sometimes an osteotomy is necessary to reposition the finger in a more functional position.

Reconstructive Procedures

Finger osteotomies are often appropriate to correct deformity and position the finger in a more functional position. Fix most finger osteotomies with K wires. These procedures are commonly performed.

Toe-to-finger transfers Toe transfers are the most effective means of improving grip function of the hand in children with absent digits. Second toe transfer is usually made. Operative indications are rare.

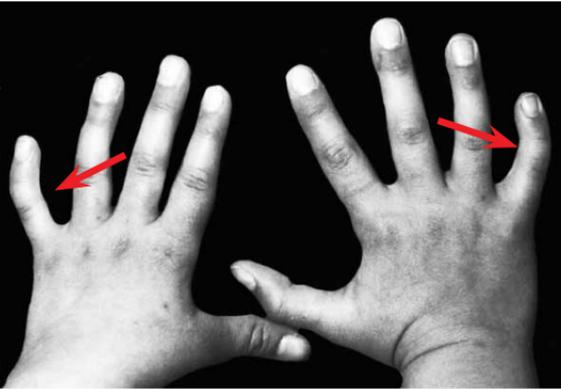
Finger lengthening of up to 10 mm in single-stage and 30 mm by gradual distraction may be achieved. Metacarpal lengthening improves pinch function in children with either transverse deficiency or constriction band syndromes. Finger lengthening improves appearance in children with brachydactyly. These lengthening procedures are rarely indicated.



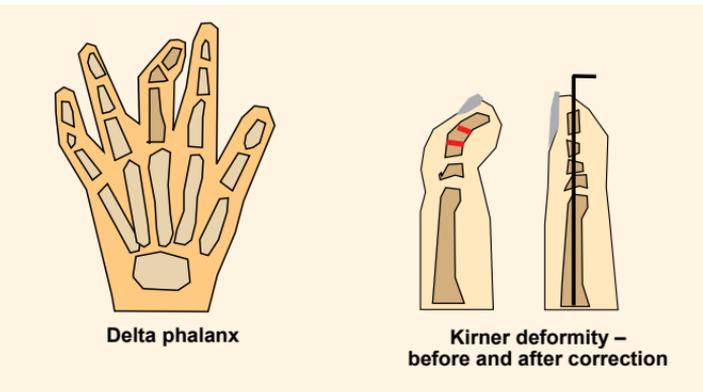
1 Operative repair of syndactyly. Repair is usually effective with good functional outcomes.



1 Camptodactyly. Note the flexion deformity of the proximal interphalangeal joint (arrow).



2 Clinodactyly. This classic deformity involves both little fingers.



3 Delta phalanx and Kirner deformity. The delta phalanx and Kirner deformity are caused by bony deformity that sometimes require operative correction.

Thumb

Hypoplasia

Thumb hypoplasia accounts for about 5% of congenital hand anomalies. Management is determined by the type of dysplasia [1].

Aplasia is treated by index pollicization.

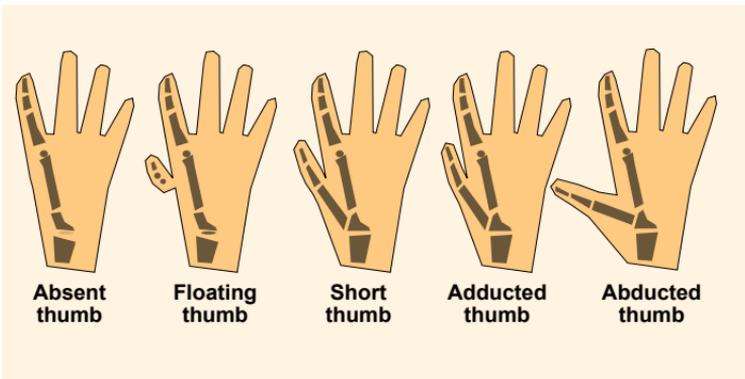
Floating thumb, or pouce flottant is usually managed by amputation, and index pollicization late in the first year.

Short thumb may be associated with a variety of syndromes. If the shortening is excessive, correct by performing a lengthening osteotomy, or by deepening the web space. Tailor correction to facilitate function.

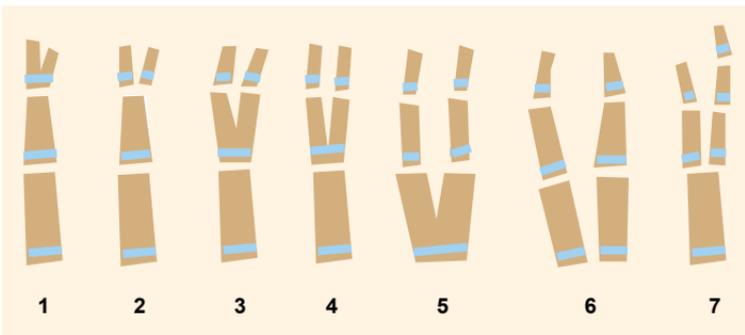
Adducted-abducted thumb deformities with shortening require tailored reconstruction that includes both soft tissue and bony reconstruction.

Polydactyly

Thumb polydactyly or *duplication*, a failure in segmentation, accounts for 5–10% of all hand deformities. Classify duplications into 7 types [2]. Type 4 is most common [1 opposite page]. Types 1–6 are usually unilateral, sporadic, and most common in boys. Type 7 may be inherited, is often bilateral, and may be associated with other abnormalities. Correct late in the first year utilizing tissues from the duplicated digits to create the best possible new thumb.



1 Thumb deformities.



2 Thumb polydactyly. Wassel classification. From Gallant and Bora (1996).

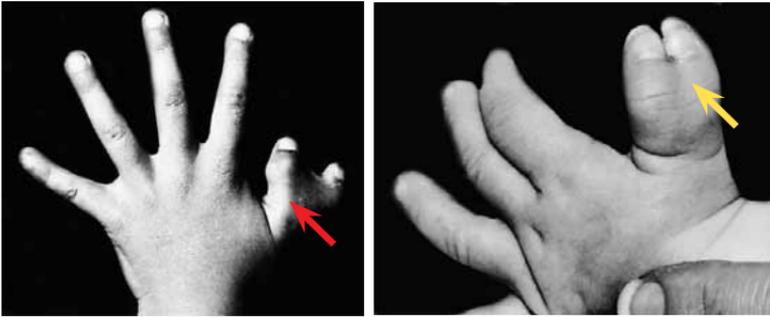
Congenital Clasped Thumb

This deformity often includes a congenital absence of the flexor tendon combined with thumb hypoplasia. Correct between 3 and 6 months with serial casting. If this fails, operative reconstruction is necessary.

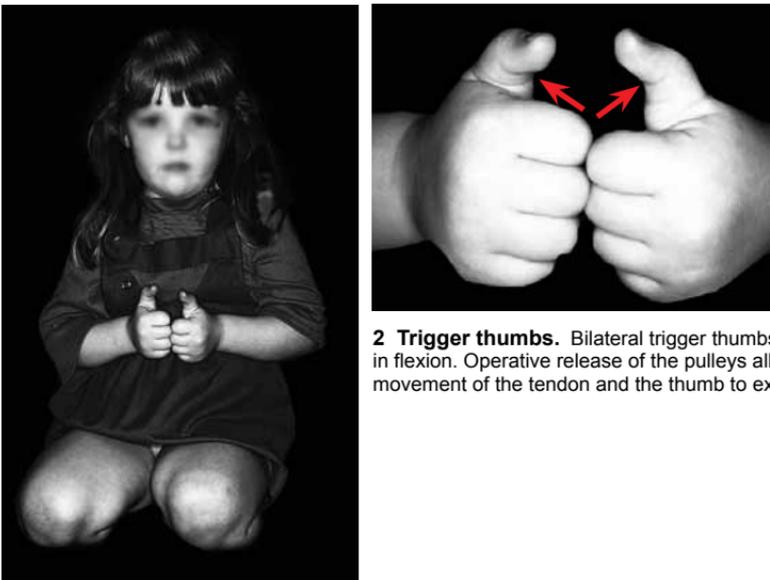
Trigger Digits

Trigger thumbs are secondary to an acquired nodular enlargement of a segment of the flexor tendon [2]. Large nodules usually become wedged at the pulley, causing the digit to lock in flexion. Smaller nodules pass through the pulley, producing a snapping sensation. Initially manage by observation. If the nodule and disability persist, consider operative release of the flexor pulley. The release frees the nodule and allows the thumb to flex and extend freely.

Trigger fingers are usually due to congenital abnormalities of the flexor mechanism. Simple release of the pulley is often ineffective, and exploration and repair of the flexor mechanism is often necessary.



1 **Thumb duplication.** Type 4 (red arrow) and type 3 (yellow arrow) are shown.



2 **Trigger thumbs.** Bilateral trigger thumbs locked in flexion. Operative release of the pulleys allowed free movement of the tendon and the thumb to extend.

Hand Infections

Hand infections [1] can be serious problems in children as they are varied, often difficult to assess, and sometimes cause long-term disability.

Penetrating Injuries

Penetrating injuries may cause infections of the soft tissues, bone, or joints of the hand. The organism is usually staphylococcus aureus.

Animal Bites

Evaluate the injury by considering the animal, nature of the wound, circumstances of the attack, interval between injury and treatment, and location of the bite. Give rabies prophylaxis for bites from carnivorous wild animals, bats, and unvaccinated domestic animals. Update the child's tetanus immunizations. Administer a broad-spectrum antibiotic early. Leave deep contaminated wounds open and close secondarily.

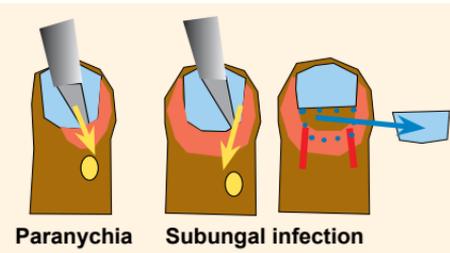
Nail Infections

Paronychia is a localized infection of the nail base. Manage with soaks and antibiotics, or drain if suppuration has occurred [2].

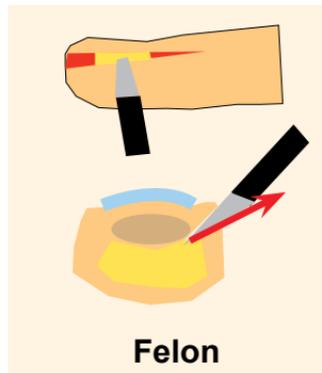
Subungual infections is a more extensive infection that often requires elevation and excision of the involved portion of the nail.



1 Bacterial infections. These include cellulitis from foreign body penetration (yellow arrow), osteomyelitis (orange arrow) and rarely residual growth arrest secondary to meningococemia (red arrow).



2 Nail infections. These may be treated with antibiotics but if suppuration occurs surgical drainage is required.



3 Felon. Drain pulp abscesses through a dorsolateral incision.

Felons

Finger tip infections may be difficult to differentiate from injury. Make this differentiation by the history, examination, systemic manifestations, and laboratory studies. Operative drainage is necessary if suppuration has occurred [3 opposite page].

Herpetic Hand Infections

Most herpetic hand infections occur in infants and young children who have oral lesions. Establish the diagnosis by clinical features, viral cultures, or Tzanck smears. Resolution occurs in 3–4 weeks. Antibiotic treatment is indicated only for superinfections. Cover the lesions to prevent spread.

Tenosynovitis

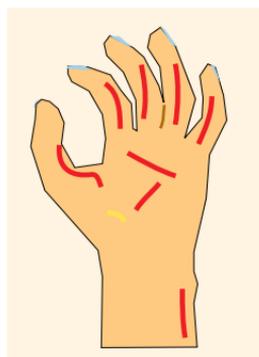
Inflammations or infections of tendon sheaths are not rare in children, and evaluation is more difficult because of lack of cooperation during examination. The pattern of bursa and tendon sheaths of the hand are the same in children and adults [1]. Ultrasound imaging may be useful in establishing the level and extent of inflammation and purulence. Manage most cases first with elevation, splinting, and antibiotics for 24 hours, then reassess. If not substantially improved consider operative drainage [2].

Dactylitis

The causes of dactylitis are numerous and include tuberculosis, sickle cell disease, congenital syphilis, psoriatic arthritis, and juvenile spondyloarthropathies. Most finger infections are due to osteomyelitis [3] or septic arthritis.

Hand Abscesses and Otopharyngeal Infections

Abscesses of the hand may be associated with inner ear or pharyngeal infections.



1 Deep infections. These may involve the tendon sheaths of the hand and cause diffuse swelling (red arrow).

2 Surgical drainage. Follows the same principles as for adults.



3 Dactylitis. Inflammation of the the digits may occur from many causes. Metacarpal osteomyelitis (red arrow) or dactylitis associated with juvenile rheumatoid arthritis (yellow arrow) are common causes.

Chapter 10 – Infection

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Infections involving the musculoskeletal system are common [1] and can cause severe disability. With optimum management, practically all infections may be cured, and deformity and disability prevented.

The prevalence of osteomyelitis is declining and changing character. Long-bone infections caused by staphylococcus aureus and septic arthritis caused by *h. influenza* have declined most. Osteomyelitis has changed forms with more complex and unusual patterns [2]. Infection with resistant strains is increasing.

Infections are still important and often challenging, but improved management makes poor outcomes less common and less acceptable.

Pathogenesis

An understanding of the pathogenesis of musculoskeletal infections facilitates management.



1 Infection causes systemic illness. This infant has septic arthritis of the elbow and is systemically ill.



2 Local spread. Infection from metaphyseal osteomyelitis may spread into adjacent joints in the infant.

Portals of Entry

Most infections are hematogenous with the primary site of entry in the in the ear, oropharynx, respiratory, GI or GU tracts [1]. Skin infections as occur after chicken pox and penetrating injuries such as nails in the sole of the foot or infections from surgical procedures are less common [2]. Extension of contiguous infections are least common, although adjacent joint infections are relatively commonly associated with adjacent osteomyelitis [2 previous page].

Initial Deposition

Bacteremia is a common event that rarely causes bone and joint infections. Why bone and joints are more vulnerable than other tissues is unknown.

Bone is usually infected in the metaphysis. Bacteria are deposited in capillary loops adjacent to the physal plate. Nearly always these bacteria are quickly destroyed by phagocytosis. Trauma is a factor that reduces resistance by causing the formation of a hematoma [3]. Bacterial proliferation is enhanced by the elaboration of biofilm, which enhances bacterial adhesion to bone and provides protection from phagocytosis or antibiotics.

Joints may be infected by hematogenous spread via the synovium, pen-

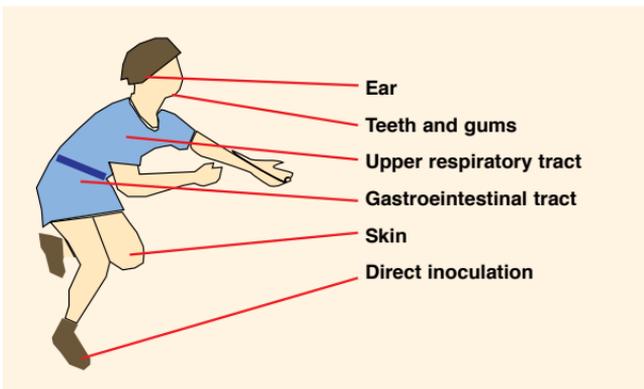
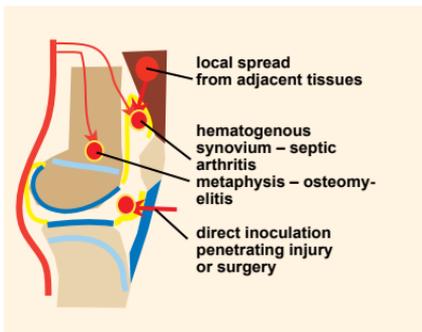
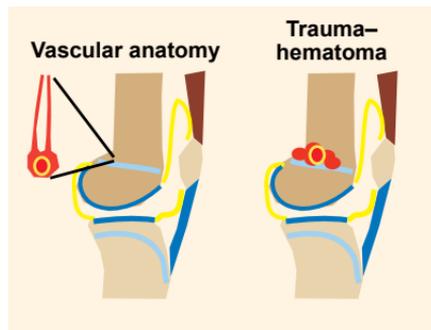


Fig. 12.3 Portals of bacterial entry.



2 Spread to bone and joints. Infection is commonly hematogenous but may be by local spread or from direct inoculation.



3 Vascular anatomy of the metaphysis. Bacteria may accumulate in vascular loops in the metaphysis. Trauma with hematoma formation provide good environment for bacterial proliferation.

etrating injury of joints, by direct spread from a contiguous infection, or by bacterial transport by way of transphyseal vessels. Transphyseal vessels are present in early infancy before the formation of the growth plate [1]. This may account for the frequency of septic arthritis of the hip in the neonate [2]. In children about a third of long-bone osteomyelitis is associated with septic arthritis of the adjacent joint.

Natural History of Infection

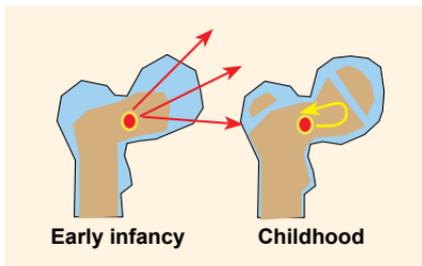
It is probable that the vast majority of bacterial colonies are destroyed by systemic and local mechanisms. The likelihood of progression is based on the balance between organism virulence and host resistance [1 next page].

Spontaneous resolution is common. Host resistance exceeds the virulence of the organism.

Subacute osteomyelitis is less common. Host resistance and virulence are about equal. A bone abscess forms reactive sclerotic bone walls off the abscess. No equivalent of this subacute form exists for septic arthritis.

Classic acute osteomyelitis or septic arthritis results from a virulent organism and a normal host. The patient becomes systemically ill, and untreated may develop septicemia and die. In others, extensive local bone necrosis occurs and chronic osteomyelitis follows.

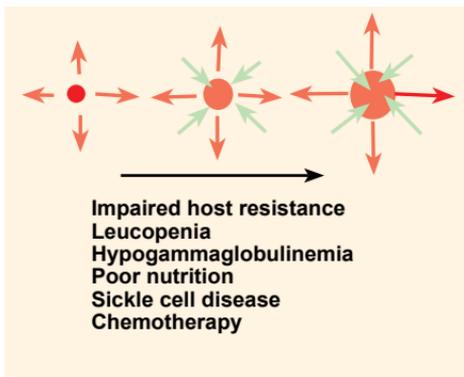
Impaired host may allow development of a bone or joint infection from organisms of relatively low virulence as seen in conditions such as sickle cell disease [3].



1 Transphyseal spread. Transepiphyseal vessels allow spread of infection from the metaphysis into the joint (red arrows). The presence of a physis blocks this spread (yellow arrow).



2 Neonatal septic arthritis of the hip. Note the widening of the joint space. This is a late finding.



3 Impaired host resistance. Organisms of low virulence may cause infections when host resistance is impaired.

Organisms

The organisms that infect the musculoskeletal system are numerous, varied, continually changing, and have predilection for site, tissue, and age of host [1 opposite page].

Staphylococci

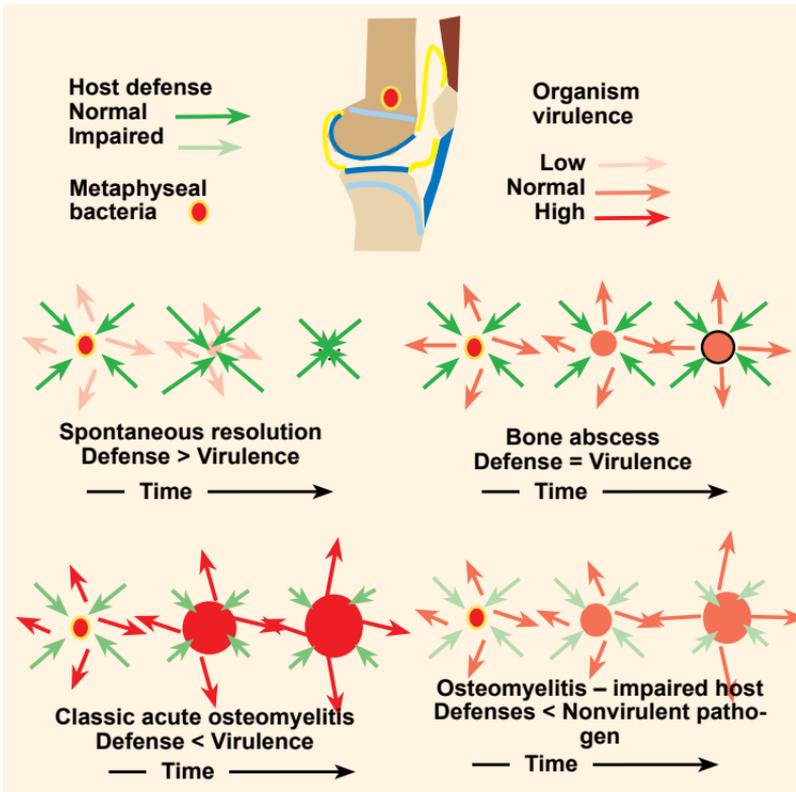
Gram positive organisms are the most common cause of infections. The incidence is declining. *Staphylococcus aureus* includes a variety of pathogenic strains. New strains are methicillin resistant. *Staphylococcus epidermidis* may cause infection in impaired hosts.

Streptococci

Streptococcal infections cause soft tissue and bony infections. β -hemolytic streptococci are common pathogens. *Streptococcus pneumoniae* may cause septic arthritis. *Streptococcus agalactiae* is a common cause of neonatal osteomyelitis. *Streptococcus pyogenes* is a less common pathogen.

Neisseria Meningitidis

Meningococcal septicemia causes acute and chronic orthopedic problems. The disseminated intravascular coagulation and focal infections acutely cause necrotizing fasciitis and damage physeal circulation, causing physeal arrest and limb deformities [2 opposite page].



1 Natural history of osteomyelitis determined by host resistance and organism virulence.

Pseudomonas Aeruginosa

This is a gram negative rod, chondrophilic, common cause of joint infections of the foot from penetrating injuries.

Escherichia Coli

This is a gram negative rod and a rare cause of musculoskeletal infections.

Salmonella

This is a gram negative rod most likely to be encountered in sickle cell osteomyelitis.

Mycobacteria Tuberculosis

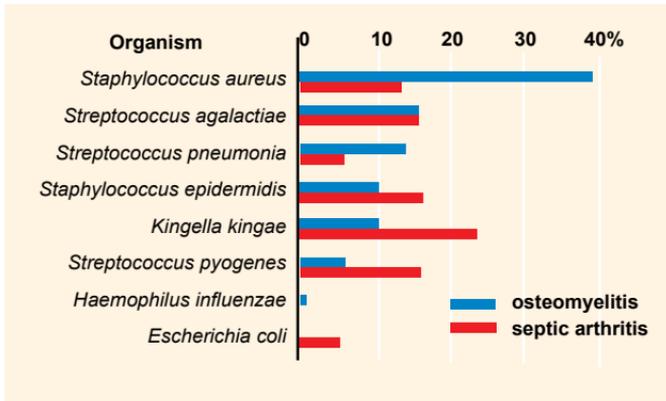
This is an acidfast organism with resurgence of worrisome drug resistant strains. It causes bone and joint infections in children. Tuberculous spondylitis with kyphosis is a common and serious deformity [3].

Kingella kingae

This is a gram negative coccobacillus common in the respiratory system, slow growing, aerobic, and fastidious. It is difficult to culture. Only recently has it been found to cause musculoskeletal infections. It remains susceptible to most antibiotics.

Haemophilus influenzae

This was previously a common cause of septic arthritis in infants. Now it is rare because of immunization programs.



1 Infecting organism in bone and joint infections in children under 36 months of age. The percentage of 30 cases of osteomyelitis (blue bars) and 30 cases of septic arthritis (red bars) caused by different organisms. From data of Lundy and Kehl (1998).



2 Septic physeal fusion. This child with meningococemia developed a fusion of the distal tibial growth plate (arrows).



3 Tuberculous spondylitis with kyphosis. This was a common condition in the past (red arrow) in North America, and it is still prevalent in developing countries (yellow arrow).

Evaluation

The child's medical history is important in the assessment of any previous injury or medical problem in the course of the current illness. The duration of symptoms in septic arthritis is of prognostic significance. Infections that are present for more than 3 days may cause residual joint damage, especially in the newborn. It is important to inquire about previous antibiotic treatment.

Physical Examination

Perform a screening examination first. Does the child appear ill [1]? The presence of systemic signs distinguishes septic arthritis from toxic synovitis. Is spontaneous movement present? The most reliable sign of septic arthritis of the hip in the newborn is a reduction of spontaneous movement of the limb. The reduction of movement from infection is described as *pseudoparalysis*. Swelling, erythema, and increased temperature are signs of inflammation, often due to infection. Note the position of the limb. Most infected limbs are positioned with the joints in slight flexion to reduce the intraarticular pressure. The hip is usually positioned in slight flexion, lateral rotation, and abduction [2].

Note the extent of soft tissue swelling and joint effusions. Try to localize the area of tenderness about the knee, ankle, wrist, or elbow to determine whether the primary problem is in the joint or the adjacent metaphysis. This is helpful in differentiating septic arthritis from osteomyelitis.

Move the joint through a gentle range of motion to assess guarding or limitation of the arc of motion. Medial rotation is limited by inflammation about the hip.



1 Ill Child. This child is systemically ill from septic arthritis. Note the lethargy and dehydration.



2 Cellulitis involving the hip. This child has a cellulitis about the left hip (arrow). The limb is held in a position of flexion and abduction that reduces soft tissue pressure and discomfort.



3 Soft tissue swelling. Soft tissue swelling is present in this infant with septic arthritis of the elbow.



4 Delay in radiographic appearance of osteomyelitis. This original film at the onset of the disease was normal (left). At 2 weeks (right), the lytic lesion is seen in the metaphysis (red arrow).

Imaging

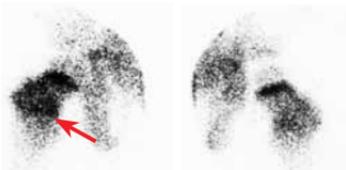
Conventional radiographs may show soft tissue swelling [3 opposite page] and obliteration of the soft tissue planes, but little else during the early course of an infection. A reduction of bone density of about 30% is necessary before radiographic changes are present. This usually requires 10–14 days [4 opposite page].

Bone scans are useful in evaluating infection in the early stages of the illness. Technesium scans in septic arthritis are usually “warm.” Scans in osteomyelitis are usually warm or hot but may be cold early in the disease. In the early phase of the disease, uptake may be reduced and a cold segment of bone may indicate the presence of a severe infection. In early osteomyelitis the phasic scan may be useful. The early phase includes vascular perfusion that parallels the physical findings of swelling and inflammation. In the second or osseous phase, uptake is greater over the site of involvement. Bone scans are not necessary if radiographic changes are already present. Often the bone scan is helpful in localizing the site of involvement [1]. Order a “pinhole collimated” scan for increased resolution. The bone scan is unaffected by bone or joint aspiration.

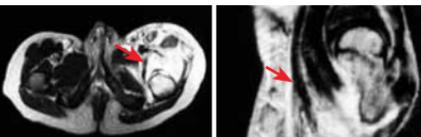
Ultrasound evaluation for hip joint effusions [3] may be helpful if the ultrasonologist is experienced. A negative study should not delay a diagnostic aspiration if the clinical signs suggest the possibility of an infected joint. Ultrasound is also useful in localizing abscess formations around long bones, and its use is underutilized.

MRI studies of infection may be useful in localizing an abscess [2]. MRI studies of discitis may be alarming and can lead to overtreatment. Use newer imaging techniques only as adjuncts to conventional well-understood techniques.

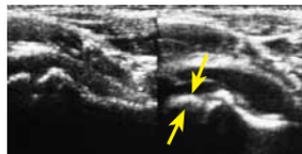
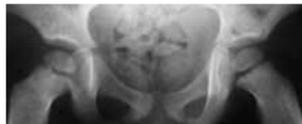
CT studies are sometimes useful in evaluating deep infections such as those about the pelvis. CT and MRI studies may be helpful in localizing abscess and planning the surgical approach for drainage.



1 Osteomyelitis of proximal femur. Bone scans aid in localizing the site of infection. The bone scan may be useful in determining the site for surgical drainage.



2 MRI showing a thigh abscess. This study shows the massive upper thigh abscess (arrows) secondary to femoral osteomyelitis.



3 Imaging in septic arthritis. This child developed hip pain and guarding. The initial radiograph (top left) was negative. The family refused aspiration. When seen the next day, the radiograph showed widening of the joint space (red arrow) and the ultrasound showed a joint effusion (yellow arrow).

Laboratory Studies

The erythrocyte sedimentation rate (ESR), C reactive protein (CRP), and cultures are the most valuable laboratory tests. Serial measures are useful in following the course of infection. Often the WBC is normal.

ESR is still valuable. Following the onset of infection the ESR slowly rises to peak at 3–5 days and remains elevated for about 3 weeks if treatment is successful [1].

CRP peaks in 2 days and follows most closely the clinical course of the infection. If treatment is successful, the values return to normal in about a week.

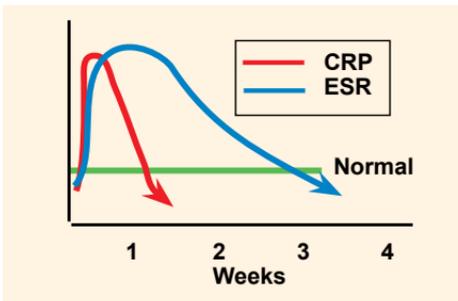
Cultures are essential and usually include blood, joint fluid, wound and biopsy samples. Blood cultures are positive in 30–50% of patients. Be aware that negative cultures are common in both osteomyelitis and septic arthritis.

Differentiation from Neoplasm

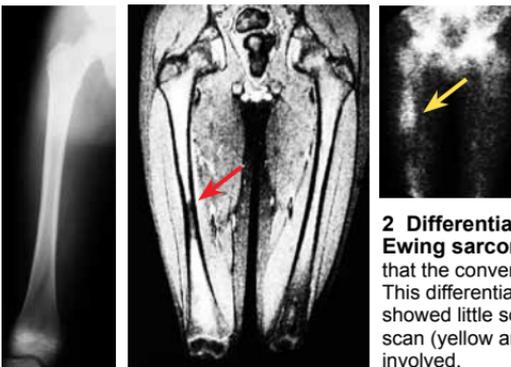
The differentiation of infection from neoplasm is sometimes difficult. Infections are more common, especially in the younger subjects, and often show signs of inflammation. Subacute osteomyelitis may be confused with osteoid osteoma, osteosarcoma, chondroblastoma, Ewing sarcoma, fibrosarcoma, or eosinophilic granuloma. If necessary, establish the diagnosis with biopsy, curattage, and cultures. If the lesion is well demarcated, making a malignant tumor less likely, consider prescribing a course of oral antibiotics. If the lesion is due to an infection, the treatment is both diagnostic and therapeutic.

Eosinophilic granuloma may show inflammatory features.

Ewing sarcoma differentiation may pose a major problem. MRI and bone scans may be helpful [2]. Sometimes biopsy and cultures are necessary.



1 CRP and ESR changes with time. Following a musculoskeletal infection, the CRP declines more rapidly than the ESR. Based on Unkila-Kallio (1993).



2 Differentiating diaphyseal osteomyelitis from Ewing sarcoma. In this child with osteomyelitis, note that the conventional radiograph was negative (left). This differentiation was aided by the MRI study, which showed little soft tissue involvement (arrow). The bone scan (yellow arrow) showed that only the femur was involved.

Management Principles

Management of infections in children is guided by a number of principles that often differ from those that apply to adults.

Greater Healing Potential

The potential for healing infection is remarkable in children. For example, discitis usually resolves with time with or without treatment. Bone damaged by osteomyelitis heals. Infection of bone may be contained and localized to only a residual abscess or resolve completely without treatment. Chronic osteomyelitis can nearly always be cured in the child. Operative wound infections are uncommon in children.

Antibiotics

The selection of the antibiotic agent is complex. Consider the disease, organism, and special features of the child. These features include the age, concurrent illness, and the family situation. The route of administration and duration of treatment are other factors to consider. Initial therapy should be intravenous or, if access is difficult, intramuscular. Certain antibiotics are most commonly used for musculoskeletal infections [1 this page and 2 next page].

Oral antibiotic therapy is justified if the infection is minor, the agent is well absorbed, and the family is reliable. In most serious infections, treatment may begin with parenteral antibiotics and switch to oral agents when the disease is under control. Before switching to the oral route of administration, be certain that adequate blood levels are documented following oral administration and that the family is reliable.

Duration of antibiotic treatment is controversial. Several factors should be considered in determining the duration. Consider the severity and potential for disability that the infection poses, the rapidity of response to treatment [2], serial determinations of the ESR and CRP, and the results of published studies. There are, however, some generalizations that can be made [1 next page]. These can be modified according to the situation. Joint suppuration in septic arthritis reduces the effectiveness of the antibiotic treatment.

Operative Drainage

Drainage may be accomplished by needle aspiration, arthroscopic decompression, or open procedures.

Agent	Dose
Oxacillin	150–200 mg/kg/day
Nafcillin	150–200 mg/kg/day
Dicloxacillin	75–100 mg/kg/day
Cephalexin	100–150 mg/kg/day
Cefazolin	100–150 mg/kg/day
Cefotaxime	100–150 mg/kg/day
Cefuroxime	150–200 mg/kg/day
Gentamicin	5–7.5 mg/kg/day
Clindamycin	30–40 mg/kg/day

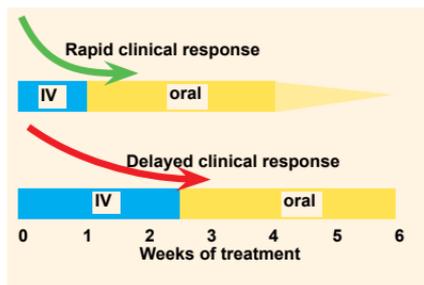


Fig. 12.22 Daily dosage of antibiotic treatment. These are some broad generalizations for infants over 1 month and for children.

2 Duration of IV antibiotic treatment. Base duration of parenteral antibiotics on clinical response.

Indications Drainage is necessary whenever antibiotic penetration into the infected site is impaired [3]. This penetration is most often due to the presence of an abscess or an accumulation of pus within a joint [1 opposite page]. Impaired penetration may also be due to a loss of vascularity as occurs in chronic osteomyelitis with sequestration or in soft tissue with poor vascularization due to thrombosis of vessels and acute inflammation. The presence of an abscess may be demonstrated by clinical examination, imaging such as ultrasound or MRI, needle aspiration, or suggested by a failure of clinical response to antibiotic treatment. This failure of response to antibiotics [2 opposite page] is the failure of reduction in fever, pain, local inflammatory signs, and CRP during the first 48–72 hours after instituting antibiotic treatment. Keep in mind that this failed response may also be due to an ineffective antibiotic agent or to an immunocompromised child.

Technique may be simply needle aspiration [3 opposite page] as is feasible for most joints, arthroscopic or open drainage. Open drainage of abscess due to acute infections requires simply draining the abscess through a small window in the cortex. If the abscess is near a growth plate, take care to avoid injuring the physis [4 and 5 opposite page]. Monitor the position of the curette with fluoroscopy.

Disease	Comment
Septic arthritis	7 days IV, 3–4 weeks total
Osteomyelitis	7 days IV, 4–6 weeks total or until ESR normal
Cellulitis	10–14 days
Surgical prophylaxis	Single dose prior to incision

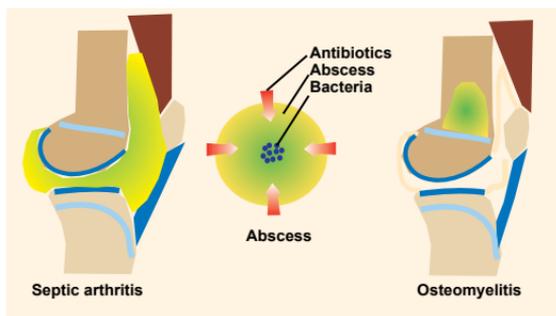
1 Duration of antibiotic treatment. These are some broad generalizations, which serve as an average duration of treatment.

Condition	Organism	Agent
Sepsis		
Neonate	Group A and B strep, coliforms	Nafcillin + gentamicin
Infant	H. flu, pneumococcus, meningococcus	Ceftriaxone or cefotaxime
Septic Arthritis		
Neonate	Group B strep, staph, coliforms	Nafcillin + gentamicin
Infant	H. flu, staph A, group A and B strep,	Cefotaxime
Child	Staph. aureus	Nafcillin
Osteomyelitis		
Neonate	Group B strep, staph A, coliforms,	Nafcillin + gentamicin
Infant/Child	staph A	Nafcillin
Nail Puncture		
Through Shoes	Pseudomonas	Ceftazidime or Ticarcillin
Barefoot	S. aureus	Nafcillin
Discitis	S. aureus	Nafcillin
Open Fractures	S. aureus	Nafcillin
Operative Prophylaxis	S. aureus	Nafcillin

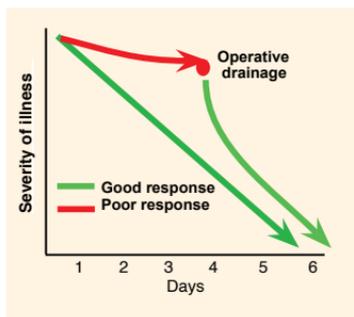
2 Commonly used antibiotics for musculoskeletal infections.

Indication	Comment
Abscess	Open drainage
Hip Joint	Open drainage
Other Joints	Drain by aspiration
Osteomyelitis	Drain abscess open
Brodie Abscess	Drain open if necessary
Sequestrum	Excise

3 Indications for drainage. These are the common indications for operative drainage.



1 Abscess protects bacteria from antibiotics. The abscess prevents antibiotic penetration, protecting bacteria.

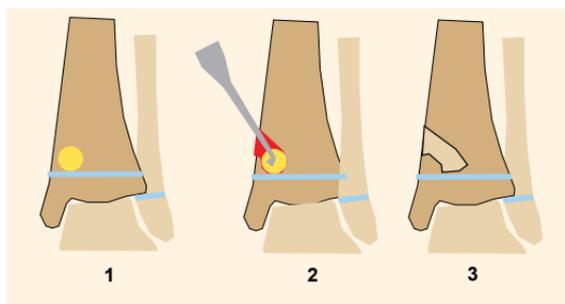


2 Indication for operative drainage. The failure of response to antibiotic treatment is often an indication for drainage.

3 Needle aspiration. This drainage is usually adequate for most joints with septic arthritis.



4 Drainage in difficult locations. Monitor position of curette with fluoroscopy and avoid the physis (red arrow). Usually a drain is placed (yellow arrow).



5 Surgical drainage of acute or subacute osteomyelitis. The infection is drained locally with care taken to avoid injury to the growth plate. Bone fill is in the defect with time.

Osteomyelitis

Osteomyelitis is an infection of bone [1]. The infection may be acute, sub-acute, or chronic. The infection may involve any bone [2]. Osteomyelitis in the preantibiotic era often caused death or severe disability. Currently, osteomyelitis remains a relatively common problem but with a much better prognosis.

Natural History

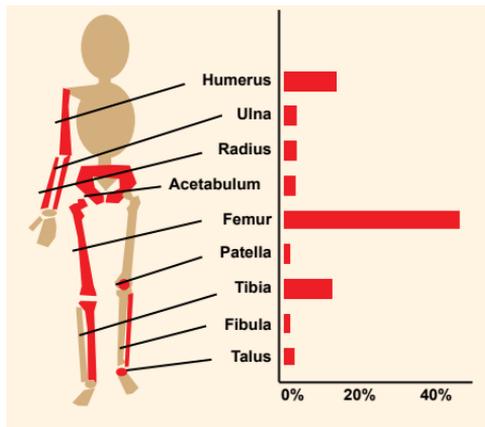
The natural history of osteomyelitis depends upon the virulence of the organism, the resistance of the host, and the age of onset [3]. Virulent organisms may cause death of the child due to overwhelming sepsis or if localized progress to chronic osteomyelitis. Chronic osteomyelitis develops through stages, which include bone and soft tissue abscesses causing sequestrum formation [1 opposite page], intermittent drainage, and a life-long disability. Chronic drainage may lead to development of squamous cell carcinoma of the sinus tracts during adult life.

Acute Osteomyelitis

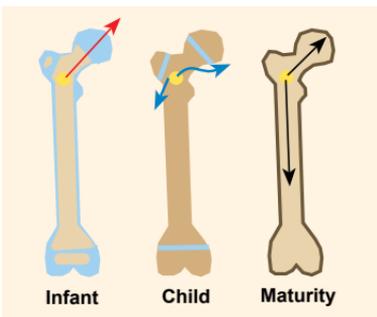
Acute osteomyelitis produces local pain, swelling, warmth, erythema, tenderness, and systemic manifestations of fever and malaise. Laboratory findings usually include a leukocytosis and elevated CRP and ESR. The CRP and ESR elevations are the most consistent laboratory findings.



1 Osteomyelitis. This boy has osteomyelitis of the upper tibia with an associated soft tissue abscess (arrow).



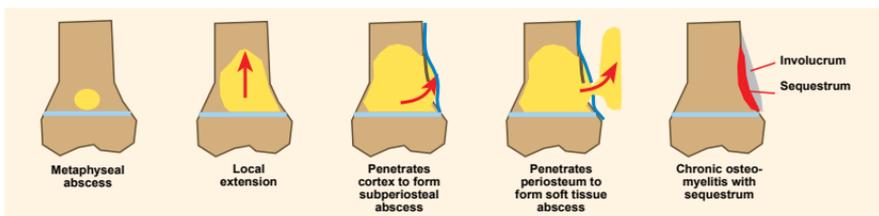
2 Distribution of osteomyelitis. From a series of 66 patients reported by Perlman (2000).



3 Spread of osteomyelitis by age. Bone structure affects the spread of osteomyelitis. In the infant the absence of an epiphyseal plate may allow spread into a joint (red arrow). In the child the path of least resistance is through the adjacent cortex to an extramedullary abscess (blue arrow). In the mature adolescent the thick cortex and absence of growth plates allows extension throughout the medullary cavity (black arrows).

Image with conventional radiographs to provide a baseline and assess for soft tissue swelling. A bone scan may be useful in localizing the site of involvement. Ultrasound and MRI studies may be helpful in localizing any abscess. To isolate the organism, culture the blood and consider aspirating the site of infection. Aspiration is most successful if a subperiosteal abscess is present.

Management When planning management, estimate the stage of the disease [1]. Antibiotic treatment is usually successful without the need for drainage if the osteomyelitis is discovered early before suppuration has occurred. Start antibiotic treatment while awaiting the results of the cultures. The selection of agent is done empirically, taking into consideration the age of the patient and the presence of any special features. Antibiotics are first given parenterally to ensure effective blood levels. The clinical course is monitored. If the antibiotic is effective against the organism and no suppuration is present, clinical improvement will occur with reduction in local signs of inflammation and systemic manifestations. If such improvement fails to occur over a period of 24–48 hours, the most likely cause is the formation of an abscess. An abscess requires operative drainage [2].



1 Natural history of osteomyelitis. The infection starts in the metaphysis. Contained by the growth plate, the infection spreads through the metaphysis, then penetrates the cortex, creating a subperiosteal abscess. This may penetrate the periosteum to produce a soft tissue abscess. During healing, new bone (involucrum) forms around the devitalized cortical bone. This dead bone is called a "sequestrum."



2 Drainage of osteomyelitis. Drain by windowing the cortex and exploring adjacent bone with a curette (yellow arrow).



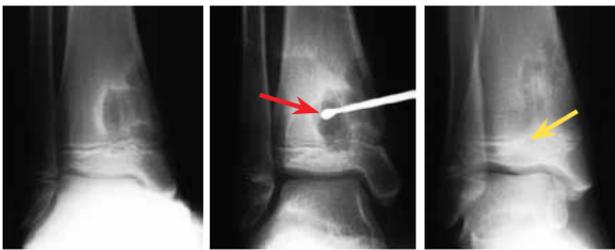
3 Subacute osteomyelitis. Osteomyelitis is contained in a bone abscess (arrows).

Subacute Osteomyelitis

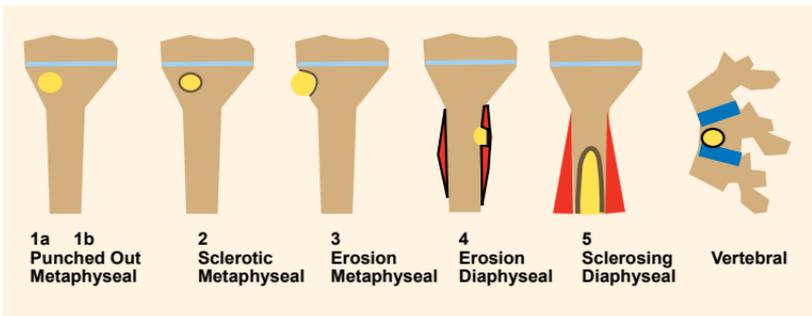
Subacute osteomyelitis is an infection with a duration longer than 2–3 weeks. Often this type of osteomyelitis is the residual of acute osteomyelitis that has been contained but not eradicated [3 previous page]. The child may show no or little systemic response but with local swelling, warmth, and tenderness. Sometimes the complaint is a limp.

Evaluation Radiographs will show the lesion. The appearance will be variable [2] and may be confused with a primary bone tumor, especially when diaphyseal and showing periosteal elevation. The differentiation between infection and Ewing sarcoma or leukemia is usually not difficult.

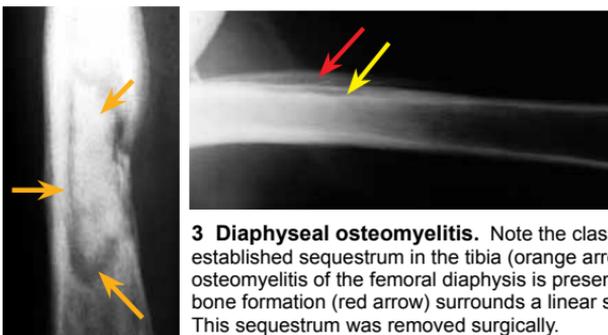
Management Manage classic metaphyseal lesions by antibiotic treatment without drainage. Drain and culture if lesions are atypical, if concern exists about a neoplastic etiology, if the child is immunologically impaired or the lesion or symptoms persist following antibiotic treatment [1].



1 Drainage of persistent subacute distal tibial osteomyelitis. Tenderness and inflammation and radiographic changes were indications for operative drainage. Avoid placing the curette across the physis (red arrow). Defect is healing 4 weeks later (yellow arrow).



2 Types of subacute osteomyelitis. Redrawn from Roberts (1982)

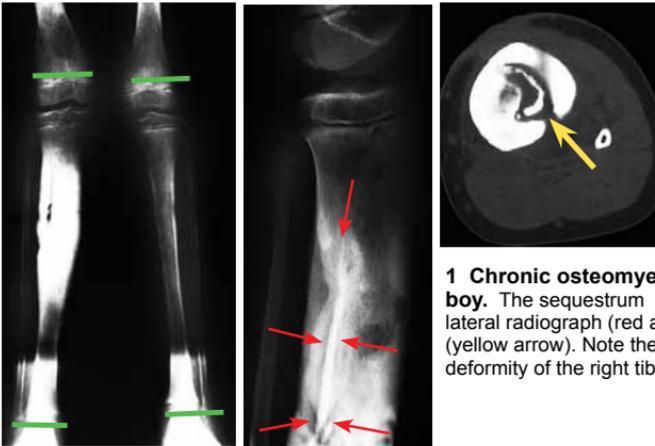


3 Diaphyseal osteomyelitis. Note the classic appearance of a well-established sequestrum in the tibia (orange arrows). Uncommon pattern of osteomyelitis of the femoral diaphysis is present in an 8-year-old girl. New bone formation (red arrow) surrounds a linear sequestrum (yellow arrow). This sequestrum was removed surgically.

Chronic Osteomyelitis

Untreated acute osteomyelitis usually becomes chronic with the disease localized to a segment of bone. Long bones are most likely to develop chronic osteomyelitis as a segment of cortex may be devascularized to form a sequestrum [3 opposite page]. Flat bones, such as those of the pelvis, are primarily cancellous with better blood supply and less likely to develop chronic disease. The patterns of chronic osteomyelitis are numerous [1 and 2]

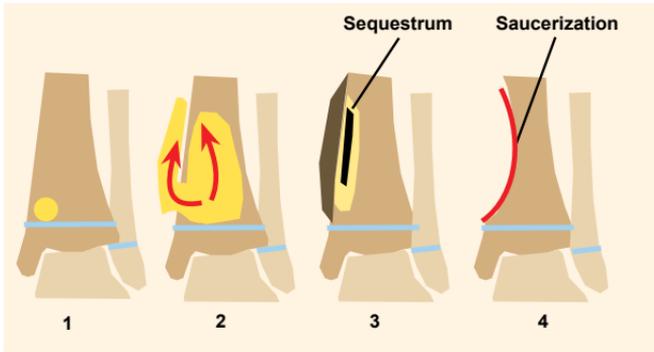
Management requires operative sequestrectomy and resection of infected tissue by saucerization to allow filling of the dead space with viable tissue [1 next page]. In very long-standing infections complex sinus tracts may develop. Assess the condition preoperatively with MRI, CT scans, and possibly by contrast injection of the sinus tract to determine its location, path, and depth of the sinus. Before resection, consider injecting dye into the sinus to stain the infected tissue [2 next page]. Plan the operative approach that will allow excision of all infected tissue. Provide antibiotic coverage based on preoperative sinus cultures. If the periosteum is viable, new bone will fill in the surgically created bony defect.



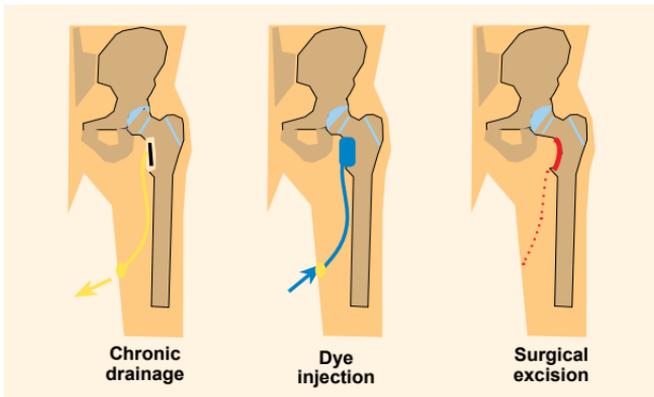
1 Chronic osteomyelitis in a 12-year-old boy. The sequestrum is clearly shown on the lateral radiograph (red arrows) and CT scan (yellow arrow). Note the overgrowth and valgus deformity of the right tibia (green lines).



2 Sclerosing osteomyelitis. The entire shaft of the femur was converted into an abscess cavity in this adolescent boy.



1 Saucerization of chronic osteomyelitis. If the infection spreads and devascularizes a segment of bone, this dead bone becomes a sequestrum (black) under the involucrum (dark brown). Manage by *saucerization* to remove the sequestrum and infected tissue. The healthy overlying soft tissue fills in the saucer.



2 Operative debridement. The site of drainage of chronic osteomyelitis (proximal medial femur) may be distant from the sequestrum (left). Define the sinus tract and infected tissue with a sinogram with methylene blue dye. Excise the sequestrum and all infected tissue (red).



3 Pathological fracture complicating osteomyelitis. This girl was treated only with antibiotics for a metaphyseal osteomyelitis. Her discharge radiographs showed no deossification. She returned 3 weeks later with a pathological fracture (red arrow) through the deossified metaphysis (yellow arrow).



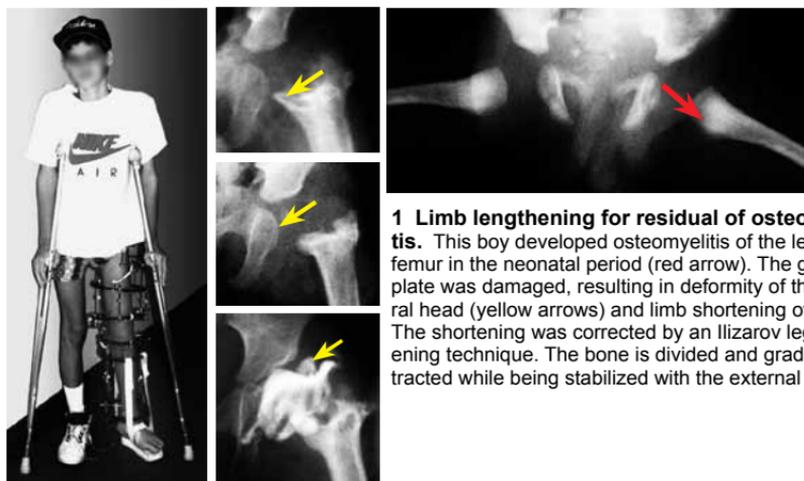
4 Severe genu valgum due to infection. This child lost the lateral half of the distal femoral growth plate due to osteomyelitis in early infancy. The deformity is progressive and difficult to correct.

Complications of Osteomyelitis

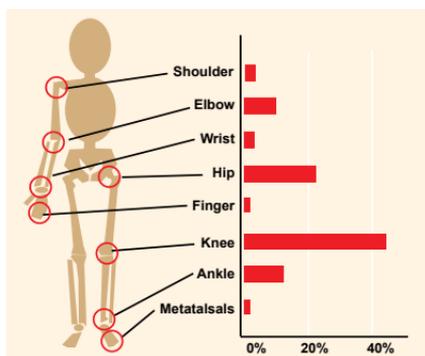
Systemic complications Untreated osteomyelitis may lead to systemic infections such as bronchopneumonia and septic pericarditis with life-threatening consequences.

Local complications are uncommon with current treatment. Complications due to deformity of bone can usually be reconstructed with a satisfactory outcome. This is in contrast to the complications of septic arthritis, which often damage joints with no satisfactory reconstruction usually possible.

- **Pathologic fracture** is a serious complication of osteomyelitis [3 opposite page]. Often the extent of the deossification is not appreciated and the child is discharged with the affected limb unprotected. Pathological fractures are slow to heal and may heal in a deformed position. The deossification resulting from osteomyelitis lags the activity of the infection by 2–3 weeks. The risk of pathological fracture should be anticipated and a protective cast applied before the deossification occurs.
- **Sequestrum formation** is usually due to delay in diagnosis. Sequestrectomy is usually effective and curative for chronic disease.
- **Growth disturbance** may be due to initial damage from the infection or operative drainage. Infections that destroy the growth plate or epiphysis may cause significant deformity [4 opposite page and 1 this page].



1 Limb lengthening for residual of osteomyelitis. This boy developed osteomyelitis of the left upper femur in the neonatal period (red arrow). The growth plate was damaged, resulting in deformity of the femoral head (yellow arrows) and limb shortening of 8 cm. The shortening was corrected by an Ilizarov leg lengthening technique. The bone is divided and gradually distracted while being stabilized with the external fixator.



2 Distribution of septic arthritis. From data of Jackson and Nelson (1982).

Septic Arthritis

Septic arthritis is a joint inflammation due to an infection usually involving synovial joints [2 previous page]. Many agents may cause septic arthritis [1], but the vast majority are due to various strains of *Staphylococcus* and *Streptococcus* and *Kingella kingae* [1 on page 377]. Septic arthritis can cause severe deformity and disability, especially when involving the hip during the neonatal period. The joint is damaged by enzymes produced by the bacteria and leukocytes causing proteoglycan loss and collagen degradation. Inflammation may cause secondary vascular damage from thrombosis or direct compression of vessels.

Natural History

Unlike osteomyelitis/ which may resolve without treatment, septic arthritis causes joint damage [2]. This makes septic arthritis a more serious disease than osteomyelitis.

Diagnosis

Clinical features are age related.

Neonate with septic arthritis may show few clinical signs. The most consistent finding is a loss of spontaneous movement of the extremity and posturing of the joint at rest. The hip is positioned in flexion, abduction, and some lateral rotation. Fever is often absent and the neonate may not appear ill.

Organism	Comment
Staphylococcus	Most common
Hemophilus	Becoming rare
Streptococcus	
Meningococcus	Primary or secondary
Pneumococcus	Infant
E. Coli	Infant
Gonococcus	Adolescent
Lyme Disease	
Tuberculosis	Increasing frequency
Fungal Infections	Certain areas endemic
Viral Infections	Rare



2 Residual deformity of septic arthritis of the hip. Note the severe deformity or Choi type 4 (see Fig. 12.56).

1 Organisms in septic arthritis. These organisms are listed in categories according to relative frequency.



3 False negative radiograph. The radiograph was read as negative, no treatment given, and the hip was destroyed by the septic arthritis.



4 Hip aspiration for diagnosis. Open drainage is required for septic arthritis of the hip.

Infant and child septic arthritis produces local and systemic signs of inflammation. The joint is swollen and tender, and the child resists movement. Hip infections result in severe limitation of rotation, a useful sign in separating septic arthritis from osteomyelitis. Radiographs early in the disease may be deceptive [3 opposite page]. A negative study is not significant. Widening of the joint is significant. Ultrasound studies may show joint effusions. Bone scans show slight to moderate increased uptake over the joint.

The most useful laboratory studies are the sedimentation rate and CRP. The ESR is usually elevated above 25 mm/hour. This test is not reliable for diagnosis in the neonate.

The diagnosis of septic arthritis is established by joint aspiration [4 opposite page]. This evaluation should be performed early and not delayed to obtain a bone scan or other imaging studies. Joint fluid in septic arthritis is cloudy, with leukocyte counts above 50,000 and PMNs predominating. Perform a Gram stain and culture. Cultures will be negative in 20–30% of cases of septic arthritis and thus a negative study does not rule out a joint infection. Culture the blood before starting antibiotic treatment.

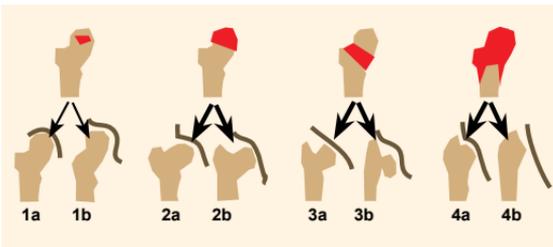
Differential diagnosis includes toxic synovitis, poststreptococcal reactive arthritis, and rheumatoid arthritis.

Age	Organism	Antibiotic	Dose
Neonate	Group B streptococcus <i>S. aureus</i> <i>E. coli</i>	Cefotaxime	150 mg/kg/day
Infant	<i>S. aureus</i> Group A streptococcus Pneumococcus	Nafcillin	150-200 mg/kg/day
Child	<i>S. aureus</i>	Nafcillin	150–200 mg/kg/day

1 Antibiotic management of septic arthritis by age group. The usual infecting organism and appropriate antibiotic are categorized by age group.



2 Arthroscopic drainage of septic arthritis of the knee. This is an acceptable method of drainage.



3 Classification of sequelae from septic arthritis of the hip. This classification demonstrates that the initial necrosis (red) determines the severity of the final deformity. From Choi (1990).

Management

Manage with antibiotics and drainage.

Antibiotic treatment Start with an agent that is statistically most likely to be effective [1 previous page]. Later, the antibiotics may be changed based on the culture reports. Parenteral treatment is continued for several days and the clinical course monitored. Failure to improve suggests that the antibiotic is ineffective or that the drainage is incomplete. The duration of parenteral antibiotics should be based on the rapidity of clinical response in reduction of fever, local inflammation, and C reactive protein response. Arbitrary rigid regimens prolong hospitalization and increase costs and patient discomfort without improving results. Most septic arthritis may be managed with parenteral antibiotics for 3 to 21 days followed by oral antibiotics for a total of about 4 weeks.

Joint drainage is necessary for all cases and should be done promptly. See page 380.

Serial needle aspiration is a traditional method of drainage. Aspirate initially and as necessary to keep the joint free of pus. Most joints should be drained several times. If response to needle aspiration is slow, consider open or arthroscopic drainage.

Open drainage is mandatory for the hip. Consider open drainage for other joints if the diagnosis is delayed or situation complicated.

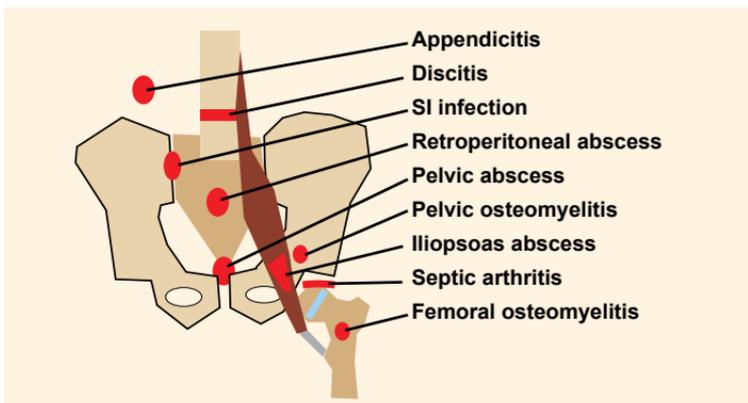
Arthroscopic drainage is an option for large joints in children [2 previous page]. Place a drain.

Immobilization in septic arthritis is unnecessary. Avoid placing the child in traction as the child will naturally hold the limb in the position of greatest comfort, which is the position in which intraarticular pressure is least.

Residual Deformity

Knee Residual deformity is most likely if the infection occurs in infancy and treatment is delayed. Usually a valgus or varus deformity develops due to displacement or loss of the physis. The deformity is usually permanent and often progressive.

Hip Ischemic changes are common and varied. Absence or delayed ossification, loss then return of ossification, or most severe complete loss or collapse. In this more severe form, increasing deformity may be present. Deformity is varied depending upon the extent of articular and physeal cartilagenous damage [3 previous page].



1 Sites of infections about the pelvis. Consider these possibilities in the differential diagnosis.

Pelvic Infections

The combination of hip or flank pain, limp, and fever suggests an infection about the pelvis [1 opposite page]. As these infections are deep in location, localization by physical examination is more difficult than for infections of the extremities. Each infection has some unique characteristics that aid in diagnosis.

Evaluation

Physical examination may localize the infection. Tenderness and pain in the back or abdomen suggests discitis or an abdominal problem. Limitation of hip rotation suggest septic arthritis of the hip. Tenderness over the SI joints or proximal femur may help localize the process to those sites. Performing a rectal examination may help localize the problem.

Imaging is usually necessary. The bone scan is most helpful in localizing the infection [1]. CT scans may show soft tissue swelling. Ultrasound evaluation may demonstrate inflammatory changes in muscles.

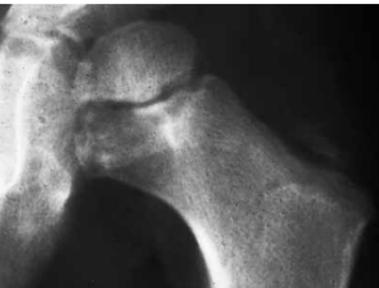
Laboratory Growth of *Staphylococcus aureus* from a pelvic abscess indicates that the process is musculoskeletal in origin. Growth of fecal flora suggests an intraabdominal cause that warrants further study.

Differential Diagnosis

Septic arthritis of the hip requires most urgent management. Pain on passive rotation of the leg suggests this diagnosis. Confirm by aspiration. Urgent open drainage is necessary.



1 Sacroiliac joint infection. Initial radiographs were negative but a bone scan demonstrated involvement of SI joint (red arrow). Radiographs 1 month later demonstrated a bone abscess (yellow arrow).



2 Osteomyelitis proximal femur. These lesions may be lytic (red arrow) or, when chronic, become more sclerotic (yellow arrow).

Iliopsoas abscess causes pain and positioning of the hip in flexion. Extension is painful. Iliopsoas abscess can be readily diagnosed by ultrasonography or computed tomography and treated by percutaneous retroperitoneal drainage.

SI infection of the joint or adjacent bone are best demonstrated by bone scan [1 previous page. Manage with antibiotic treatment. Drainage is usually not necessary.

Pelvic osteomyelitis may occur in varied sites. Localize with bone scan, demonstrate any abscess by US or CT scans. Treat with antibiotics. If unresponsive, consider aspiration with image guidance.

Femoral osteomyelitis is more serious with the potential for joint or growth damage [2 previous page]. Open drainage is often necessary.

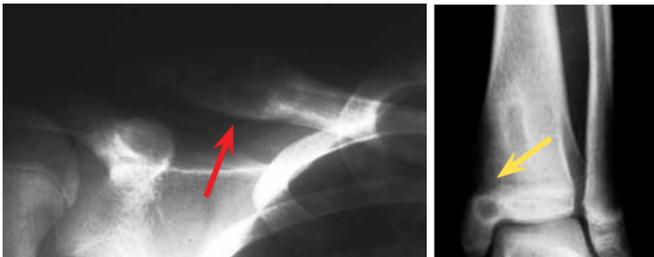
Unusual Forms of Osteomyelitis

Osteomyelitis of the Clavicle

The clavicle responds to osteomyelitis with thickening and cystic changes that give the appearance of a neoplasm [1, left]. CT scans may demonstrate a bone abscess, which may be drained. Cultures may be negative. Consider chronic recurrent multifocal osteomyelitis in the differential. Treat bacterial infections with drain and antistaphylococcal antibiotics.

Epiphyseal Osteomyelitis

Primary hematogenous osteomyelitis rarely affects the epiphysis primarily [2, right]. The infection may spread through the growth plate from a metaphyseal origin. The physeal erosion allowing this transphyseal spread



1 Unusual forms of osteomyelitis. Chronic osteomyelitis of the clavicle produces bony overgrowth with a cystic appearance often confused with a neoplasm (red arrow). Rarely, lesions occur in the epiphysis (yellow arrow).



2 Salmonella osteomyelitis in sickle cell disease. This osteomyelitis elicits an extensive subperiosteal new bone formation (red arrows) that completely surrounds the original diaphysis (yellow arrows).

usually heals without the formation of a physeal bridge. The exception is in meningococemia and severe infections with delayed treatment.

Salmonella Osteomyelitis

Salmonella and staphylococcal aureus osteomyelitis occur in children with sickle cell disease [2 opposite page] The infection is characterized by polyostotic distribution, extensive diaphyseal involvement, massive involucrum, and frequent complications due to compromised immune status and poor circulation of blood in bone. Manage by decompression and parenteral antibiotics.

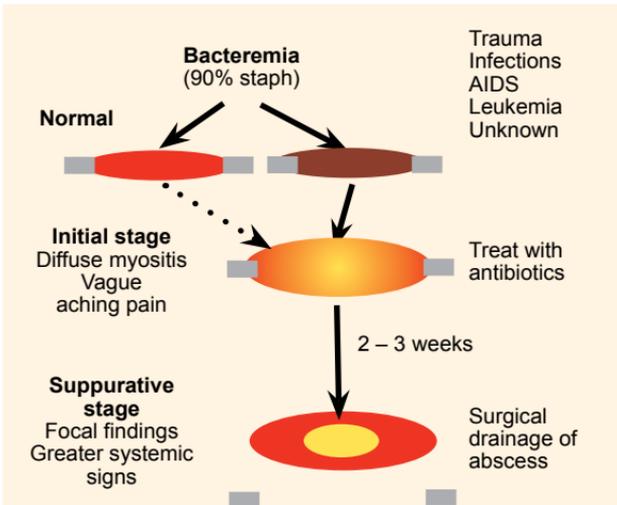
Soft Tissue Infections

Chicken Pox (Varicella)

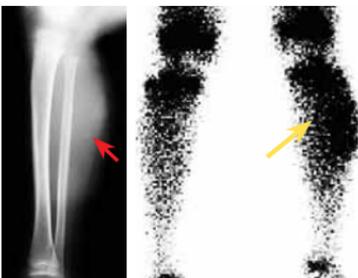
Group A streptococcal infections may cause cellulitis, abscesses, septic arthritis, or extensive necrotizing fasciitis.

Toxic Shock Syndrome (TSS)

TSS is due to a toxin elaborated by different types of *S. aureus* and streptococcus. TSS has been reported about 2 weeks following orthopedic procedures and under casts in children. About half are nonmenstrual. The characteristic features include high fever, vomiting, diarrhea, rash, hypotension, pharyngitis, headache, and myalgia. Management is directed toward controlling the effects of the toxemia.



1 Natural history of polymyositis. The natural history of this infection includes cellulitic and suppurative stages.



2 Polymyositis. Note the soft tissue swelling (red arrow) and increased muscle uptake (yellow arrow) on bone scan.

Polymyositis

Muscle abscesses are infrequent as skeletal muscle is resistant to bacterial infections. A bacteremia seeds muscle abscess [1 previous page]. In some cases some underlying condition reduces resistance. Untreated, the generalized inflammation becomes focal with abscess formation in 2–3 weeks. The child becomes progressively more ill with the potential of death. *Tropical polymyositis* often occurs in anemic and malnourished children.

Initial stage The child presents with poorly localized aching pain and fever. The most common sites are the hip and thigh. Clinical and laboratory signs of infection are present. Radiographs show soft tissue swelling, bone scan shows increased uptake [2 previous page], and the MRI is most specific and diagnostic. Treat with parenteral antistaphylococcal antibiotics.

Suppurative stage The child shows more systemic signs and focal tenderness. The MRI demonstrates a muscle abscess. Confirm the diagnosis and determine the organism by aspiration of the abscess. In some this is adequate. Most require operative drainage.

Lyme Disease

Lyme arthritis in children may mimic other pediatric arthritides. The natural history of untreated Lyme disease in children may include acute infection followed by attacks of arthritis and then by keratitis, subtle joint pain, or chronic encephalopathy. Treat with amoxicillin, doxycycline, and ceftriaxone. With treatment complete, resolution is expected within 2–12 weeks and prognosis is excellent.

Puncture Wounds

Foot infections are often due to puncture wounds. The classic example is the nail puncture wound of the foot. When these occur through shoes, the infecting organism is *Pseudomonas*. Puncture wounds in other situations are usually due to staphylococcus. Retention of foreign material such as wood may be best imaged by ultrasound [1]. Removal of foreign bodies is often more difficult than expected. See page 378.



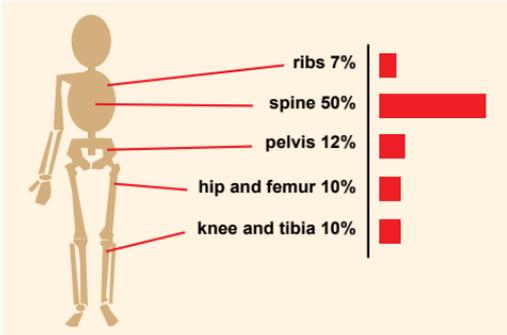
1 Foreign body infections. The foot is swollen (red arrow) but without bony change. The ultrasound shows the wood fragment (yellow arrow).

Tuberculosis

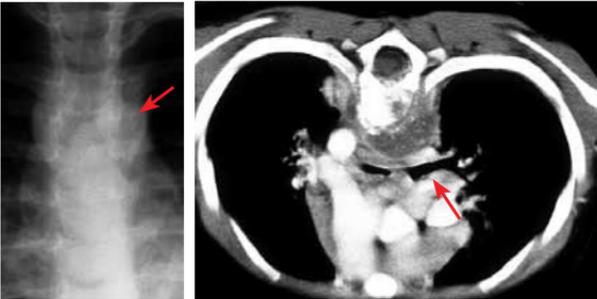
More new cases of tuberculosis are being seen throughout the world. This is due to increasing number with suppressed immune systems, drug resistant strains of *Mycobacterium*, an aging population, and more exposed health care workers. Musculoskeletal tuberculosis most often involves the spine [1].

Spinal Tuberculosis

In children the infection usually involves only bone [2] leaving the disc and cartilagenous endplates intact. This improves prognosis and allows spontaneous correction of the kyphosis with growth. Medical management is the primary treatment. Treat all patients with at least three drugs for a prolonged period.



1 Distribution of musculoskeletal tuberculosis.



2 Tuberculous paravertebral abscess. The abscess can be seen both on the chest radiograph and the CT scan (red arrows).



3 Tuberculous hip infection. Note the infection involves the proximal femur (red arrow), joint (yellow arrow), and acetabulum (orange arrow).

Operative management is controversial. Undisputed indications include a significant neurological deficit, a neurological deficit or kyphosis progressing despite adequate medical management, or compromised pulmonary function from the abscess.

Tuberculous Osteomyelitis

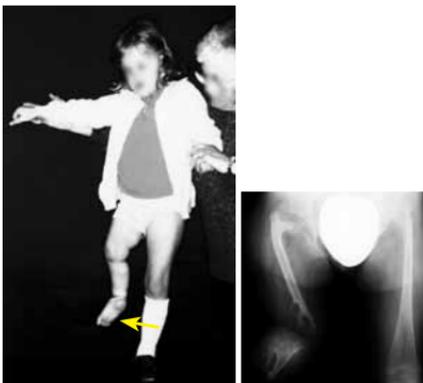
In young children this may be associated with BCG vaccination. The children are usually afebrile and show local swelling and discomfort, which may alter function. Mild leukocytosis and increased ESR are common. The CRP is usually normal. Radiographs show metaphyseal lesions with soft tissue swelling. Manage with operative drainage and long-term antituberculosis chemotherapy for about 1 year.

Tuberculous Arthritis

In contrast to pyogenic arthritis, tuberculosis causes a slow, progressive joint disintegration [3 previous page]. Management usually requires medical management, joint debridement, and stabilization such as by arthrodesis.



1 Necrotizing fasciitis. This 12-year-old boy developed meningococemia and four-extremity involvement. Despite fasciotomies (arrows), the limbs became gangrenous. The disease was fatal.



2 Meningococemia with growth plate injury. This child developed meningitis and meningococemia. Meningococcal osteomyelitis damaged the growth plate causing severe residual shortening of the right leg. A below the knee amputation was performed to make prosthetic fitting possible (yellow arrow).



3 Physeal arrest from meningococemia. Note the sclerosis and shortening of the radius.

Meningococcal Infections

Purpura Fulminans

Meningococemia causes disseminated intravascular coagulation. This results in extensive soft tissue damage with compartment syndromes and skin necrosis most evident in the extremities [1 opposite page].

Meningococcal Multifocal Osteomyelitis

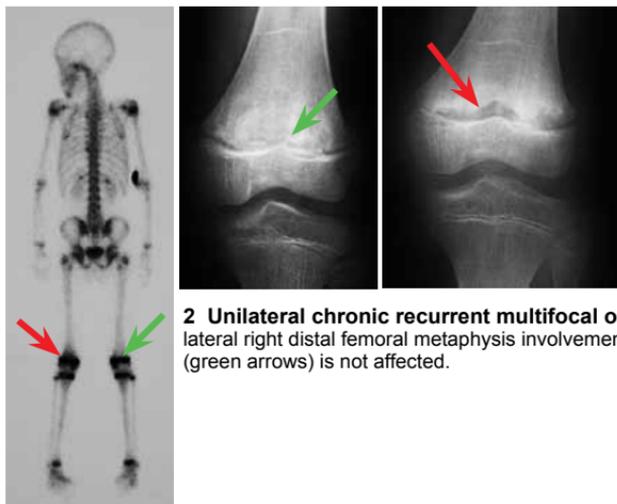
This infection is unique as it often affects the growth plate causing physeal fusions and severe deformities [2 and 3 opposite page].

Chronic Recurrent Multifocal Osteomyelitis

Chronic recurrent multifocal osteomyelitis (CRMO) is a rare form of osteomyelitis of unknown etiology characterized by symmetrical bone lesions [1]. It is occasionally associated with palmoplantar pustulosis. The lesions were most often located at the metaphyseal region of tubular bones and the clavicle, but also at the spine, ischiopubic bone, and the sacroiliac joint. Progressive sclerosis and hyperostosis occurred mostly in the clavicle and occasionally in the tibia, femur, metatarsal, and ischiopubic bone, similar to those sclerosing bacterial osteomyelitis. Less common is unilateral involvement [2] CRMO is often recurrent, but resolves slowly over an extended period. Cultures are negative and antibiotic treatment ineffective. Treat with nonsteroidal antiinflammatory drugs.



1 Chronic recurrent multifocal osteomyelitis. This atypical form of osteomyelitis produces symmetrical bone lesions (red arrows).



2 Unilateral chronic recurrent multifocal osteomyelitis. Note the unilateral right distal femoral metaphysis involvement (red arrows). The left femur (green arrows) is not affected.

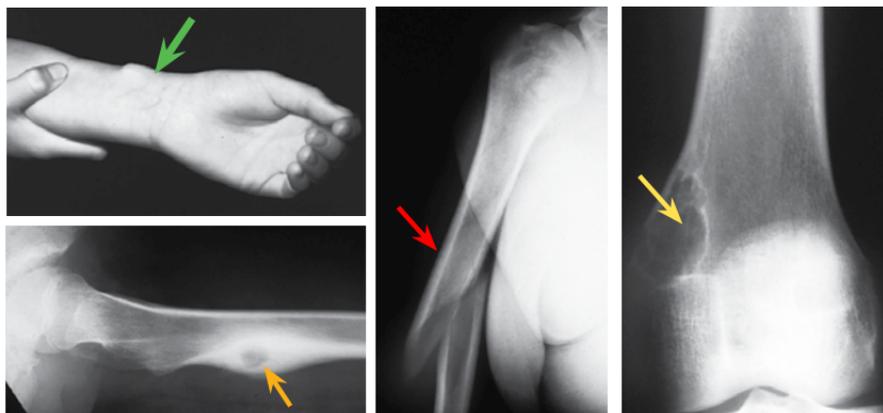
Chapter 11 – Tumors

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About 2000–3000 new cases of malignancies of the musculoskeletal system are diagnosed each year in United States. The number of benign neoplasms is estimated to be about ten times this number.

Evaluation

Evaluate tumors by taking the patient's history, performing a careful physical examination, and obtaining necessary laboratory and imaging studies. Usually the diagnosis of a tumor is made by the presence of pain, a mass, or a pathologic fracture, it may be as an incidental finding [1].



1 Presentations of tumors in children. The common modes of presentation are with a mass, as in osteochondroma (green arrow); with pain, as in osteoid osteoma (orange arrow); with a pathologic fracture, as in osteosarcoma (red arrow); or as an incidental finding, such as this small nonossifying fibroma (yellow arrow).

History

Tumors usually present as a mass, produce pain, or cause disability. How long a mass has been present is often difficult to determine from a history. Frequently, a large lesion such as a slow-growing osteochondroma is not noticed until shortly before the consultation. The family incorrectly concludes that the tumor had grown quickly.

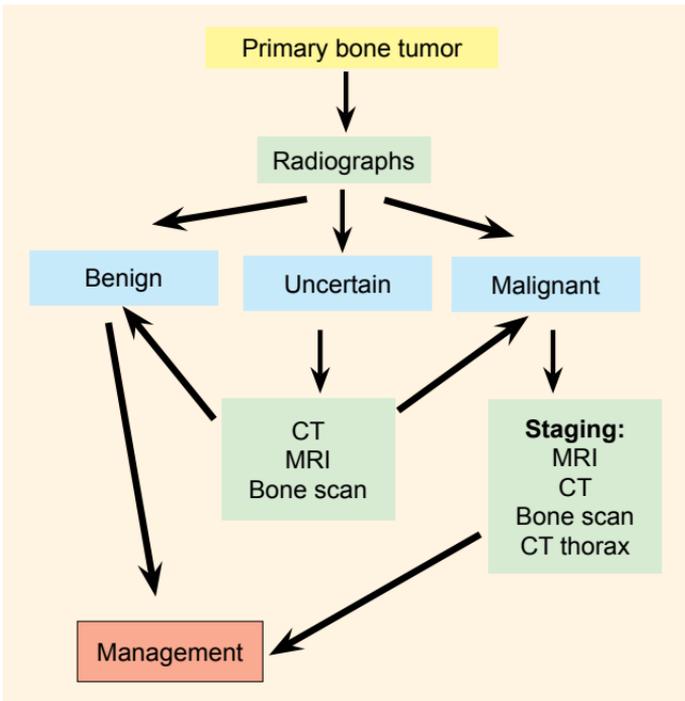
Pain is a more reliable indicator of the time of onset of a tumor. Inquire about the onset, progression, severity, and character of the pain. Night pain is characteristic of both malignant tumors and some benign lesions such as osteoid osteoma. Malignant lesions produce pain that progressively increases over a period of weeks. Night pain in the adolescent is especially worrisome and should be evaluated first with a conventional radiograph. An abrupt onset of pain is usually due to a pathologic fracture. Such fractures most commonly occur through bone cysts.

Age of the patient is helpful. Bone lesions in children under age 5 years are likely to be due to an infection or eosinophilic granuloma. Giant cell tumors and osteblastomas occur in the late teen period.

Race is notable, as blacks seldom develop Ewing sarcoma.

Examination

A screening examination should be performed first. Some lesions, such as osteochondroma, are usually multiple. Look for asymmetry, deformity, or swelling. Palpate for masses. If a mass is present, measure its size, assess for tenderness, and note any associated inflammation. Malignant tumors are typically firm, often tender, and may produce signs of inflammation.



2 Flowchart for imaging primary bone tumors.

Imaging

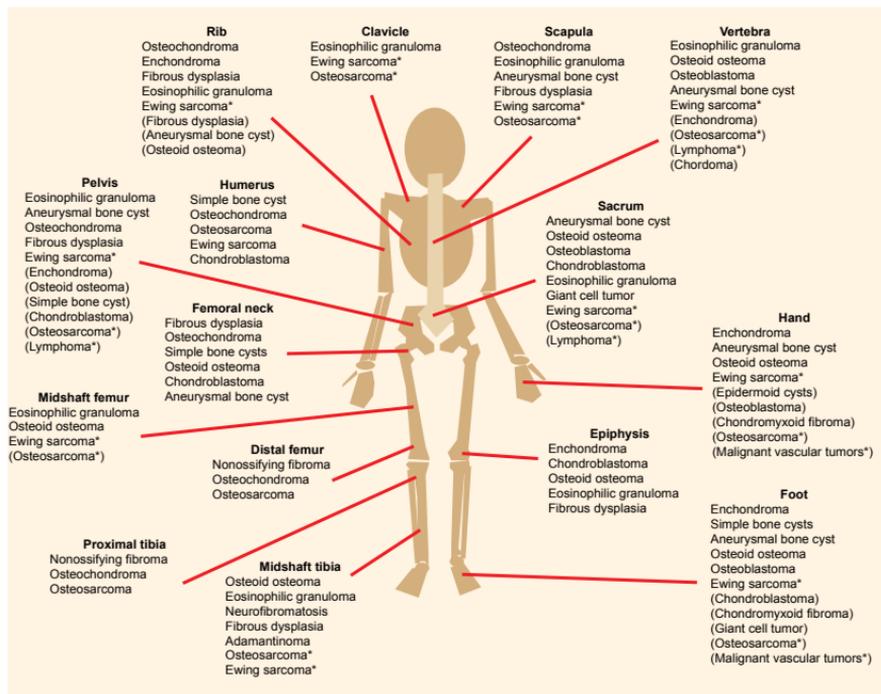
Order imaging studies with a plan in mind [2 opposite page]. Start with good- quality radiographs. Conventional radiographs remain the basic tool for diagnosis. Consider several features in assessment.

Location Lesions tend to occur in typical locations both with respect to the bone involved [1]) and the position in the bone [1 next page].

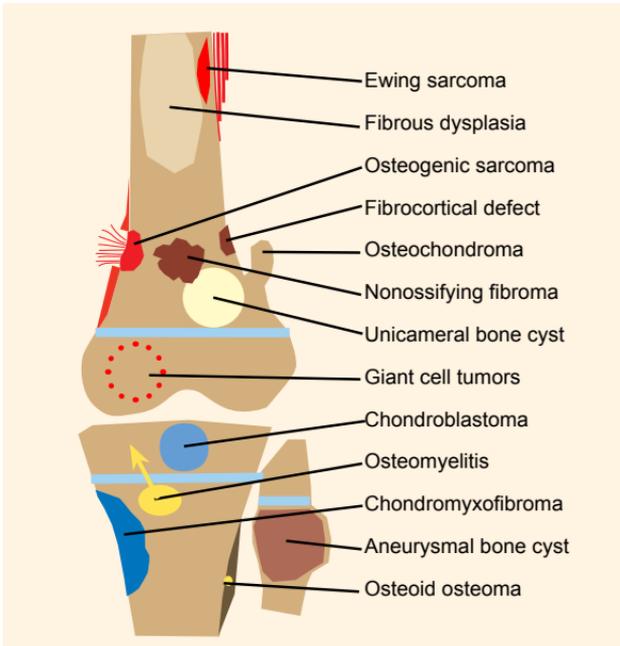
Effect of lesion Note the lesion’s effect on the surrounding tissue [2 next page].

- **Effect of lesion on bone** Sharply punched out lesions are typical of eosinophilic granuloma. Osteolytic lesions are typical of most tumors; few are osteogenic on radiographs.
- **Effect on normal adjacent bone** is useful in determining the invasiveness of the lesion. An irregular, moth-eaten appearance suggests a malignant lesion or an infection. A lesion that expands the adjacent cortex is usually benign and typical for aneurysmal bone cysts.
- **Diagnostic features** suggest the aggressiveness of the lesion. Sclerotic margination suggests that the lesion is long-standing and benign. Periosteal reaction suggests a malignant, traumatic, or infectious etiology.

Special imaging Consider special types of conventional radiographs such as for soft tissue or bone detail.



1 Tumor types per site. Less common tumors at each site are in parentheses. Asterisks (*) indicate malignant tumors. Based on Adler and Kozlowski (1993).



1 Typical locations for various tumors.

Effect of lesion on bone			Effect of lesion on normal adjacent tissue		
<p>Punched out margins Eosinophilic granuloma</p>	<p>Destructive Blurred margins Malignant tumor Infection</p>	<p>Aggressive Crosses physis Malignant tumor Infection</p>			
<p>Effect of lesion on bone. Bone destruction from eosinophilic granuloma in an infant (green), destructive lesion in osteogenic sarcoma (yellow), and aggressive osteogenic sarcoma that readily crosses the physis (red).</p>					
<p>Sclerotic margins benign Nonossifying fibroma</p>	<p>Expansile benign Aneurysmal bone cyst</p>	<p>Bone reaction varied Osteogenic sarcoma Eosinophilic granuloma Infection</p>			
<p>Effect of lesion on normal adjacent tissue. Note the sclerotic margins in the nonossifying fibroma (green), expanded cortex in an aneurysmal bone cyst (yellow), and marked periosteal reaction in an eosinophilic granuloma (red).</p>					
Diagnostic features of lesion			Diagnostic features		
<p>Ground glass Diaphyseal fibrous dysplasia</p>	<p>Speckled Cartilage tumor</p>	<p>Osteoblastic Varied tumor Infection</p>			
<p>Diagnostic features. Note the ground glass appearance of fibrous dysplasia (green), speckled calcification in a cartilage tumor (yellow), and osteoblastic features of an osteoblastic osteogenic sarcoma (red).</p>					

2 Diagnostic features by conventional radiography. Note the effect of the lesions on bone (top), the effect on normal adjacent tissues (middle), and special diagnostic features (bottom).

Special studies may be essential to establish the diagnosis [1].

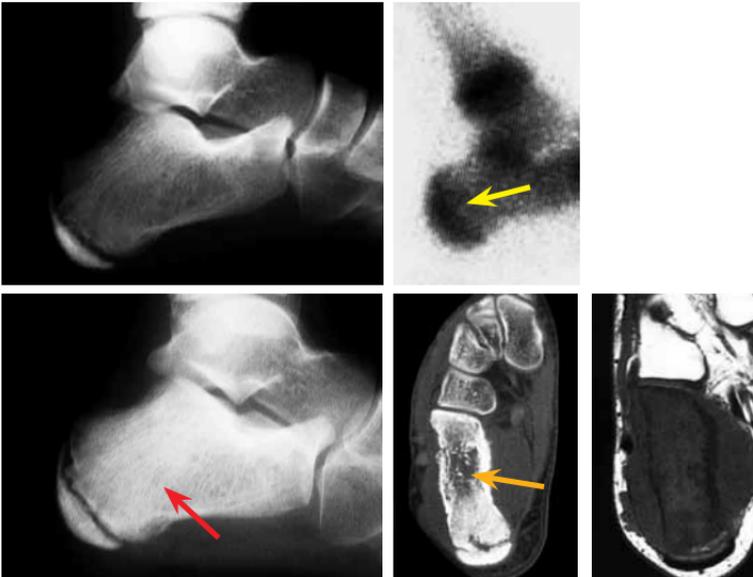
- **CT scans** are useful in assessing lesions of the spine or pelvis. Whole lung CT studies are highly sensitive for pulmonary metastases.
- **MRI** is the most expensive and is limited in young children because of the need for sedation or anesthesia. However, it is the most sensitive in making an early diagnosis and excellent for tissue characterization and staging of tumors.
- **Bone scans** are the next most useful diagnostic tool. These scans are helpful in determining if the lesion is solitary and if other lesions are present. The uptake of the lesion is important to note. A cool or cold scan suggests that the lesion is inactive, and only observation may be necessary. Warm scans are common in benign lesions. Hot scans suggest that the lesion is very active and that it may be either a malignant or benign lesion, such as an osteoid osteoma. Biopsy or excision is required.

Laboratory

CBC is useful as a general screening battery and helpful in the diagnosis of leukemia.

Erythrocyte sedimentation rate (ESR) is often elevated in Ewing sarcoma, leukemia, lymphomas, eosinophilic granuloma, and infection.

Alkaline phosphatase values may be elevated in osteosarcoma, Ewing sarcoma, lymphoma, and metastatic bone tumors. The value of the study is limited because of the natural elevation of this value during growth, especially in the adolescent.



1 Evaluation by imaging. This child had foot pain and a negative radiograph (upper left). A month later, the patient was seen again with increasing night pain. At that time, a bone scan showed increased uptake (yellow arrow), the radiograph showed increased density of the calcaneus (red arrow), a CT scan showed erosion of the calcaneus (orange arrow), and MRI showed extensive marrow involvement (white arrow). Ewing sarcoma was suspected by these findings.

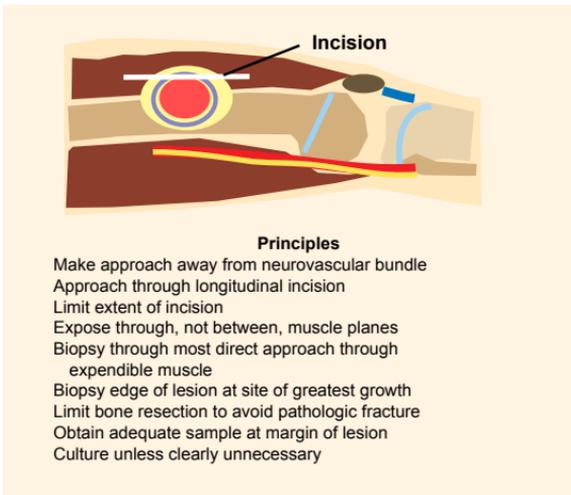
Biopsy

The biopsy is a critical step in management [1] and should be performed thoughtfully by an experienced surgeon. In most cases, an open biopsy is appropriate. Needle biopsy is indicated for lesions located at inaccessible sites and for special circumstances. The biopsy should provide an adequate sample of involved tissue, and the tissue should be cultured unless the lesion is clearly neoplastic. The biopsy procedure should not compromise subsequent reconstructive procedures.

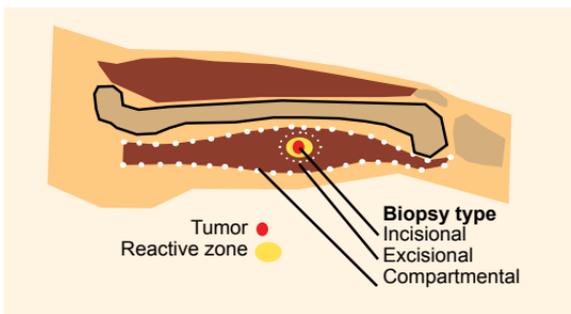
Biopsies can be incisional, excisional, or compartmental in type [2]. Excisional biopsy is appropriate for benign lesions such as osteoid osteoma [1 opposite page] or for other lesions when the diagnosis is known before the procedure and the lesion can be totally resected.

Staging

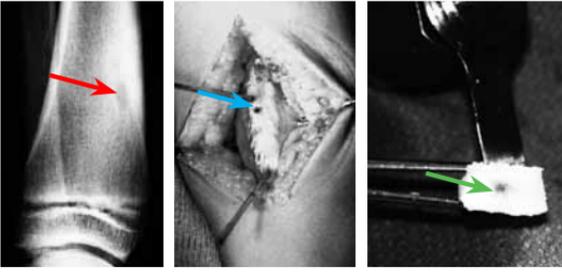
Staging of malignant tumors provides a means of establishing a prognosis. Prognosis depends on the grade of the lesion (potential for meta-stases), the extent and size of the lesion, and the response to chemotherapy. The extent of the lesion is categorized by whether the lesion is extracompartmental or intracompartmental [2 opposite page] and whether any distant metastases are present. A knowledge of the response to chemotherapy [3 opposite page] helps the surgeon to determine the appropriateness of limb salvage procedures and how wide the surgical margins must be to avoid local recurrence following resection.



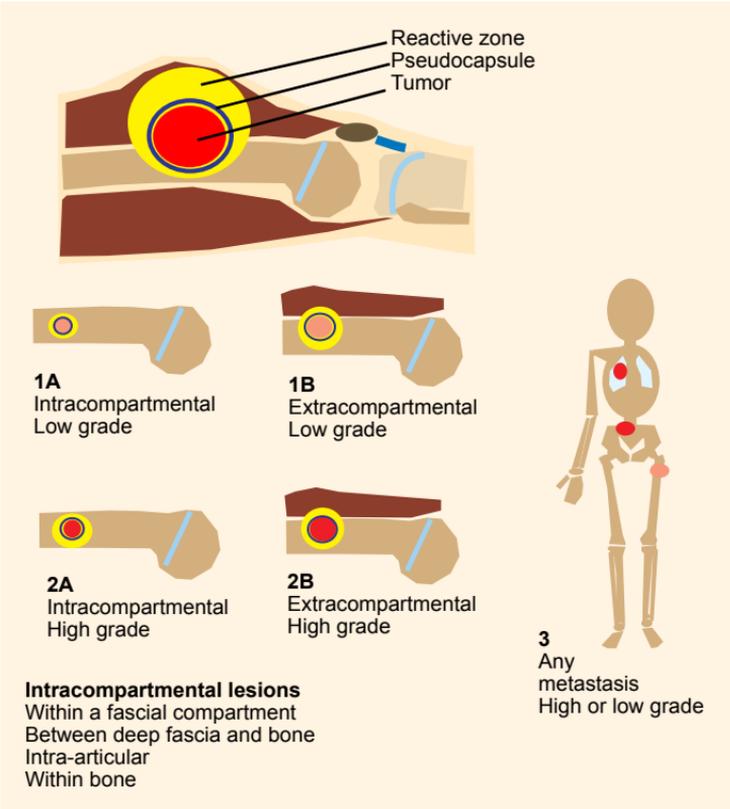
1 Biopsy principles. Follow these basic principles during biopsy procedures.



2 Biopsy types. These include incision, excision, and compartmental resection, depending upon the objective of the biopsy.



1 Excisional biopsy of osteoid osteoma. The original lesion (red and blue arrows) is excised in a block of bone. This block is divided in the operating room to make certain the entire lesion is removed. The nidus of the tumor is clearly seen (green arrow).



2 Staging of musculoskeletal tumors. Staging is determined by the grade and extent of the lesion. Based on Wolf and Enneking (1996).

Grade	Tumor response in osteosarcoma
1	Little or none
2	Extensive necrosis >10% viable tumor
3	Extensive necrosis <10% viable tumor
4	Complete necrosis

3 Response to chemotherapy. The response to chemotherapy is helpful in determining the prognosis and subsequent management.

Differential Diagnosis

Differentiating from Myositis Ossificans

Differentiating bone tumors from myositis ossificans (MO) is sometimes difficult. MO lesions have reactive bone that is most active on the margins. MRI studies are rarely necessary, but will show an inflammatory lesion with a tumor core [1].

Differentiating Neoplasms, Infection, and Trauma

Sometimes a child presents with pain [3] and tenderness over a long bone (usually the tibia or femur). The radiographs may be negative or show only slight periosteal elevation. The differential diagnosis often includes osteomyelitis, a stress fracture, or Ewing sarcoma [2]. This differentiation is usually established without a biopsy. The evaluation usually requires a careful physical examination, radiographs, a bone scan, and a determination of the ESR.



1 Myositis ossificans. Note that the lesion appears to be extracortical in origin and that the inflammatory mass does not include the bone.

2 Bone infections. Sometimes unusual bone infections may be difficult to differentiate from tumors, as a diaphyseal lesion (red arrow) or osteomyelitis that extends across the physis (yellow arrow).

Category	Examination	Imaging or laboratory	Comment
Malignant tumor	Mass, diffuse tenderness and MRI findings	Hot bone scan	Pain progressive, often occurs at night
Osteomyelitis	Inflammatory signs Metaphyseal location	Elevated ESR Warm to hot bone scan Culture positive	Systemic illness Recent onset
Stress fracture	Localized tenderness Typical locations	Normal ESR, no mass	Overuse history Pain reduces with rest

3 Differentiating tumor, infection, and traumatic lesions. Features of the lesion allow classification into diagnostic categories without the need for a biopsy.

Unicameral Bone Cysts

Unicameral (UBC), solitary, or simple bone cysts are common lesions of unknown cause that generally occur in the upper humerus or femur [1]. Theories of etiology include a defect in enchondral bone formation or altered hemodynamics with venous obstruction causing increased interosseous pressure and cyst formation. The cysts are filled with yellow fluid and lined with fibrous tissue [2].

Diagnosis

These cysts are most often first diagnosed when complicated by a pathologic fracture [1 next page]. Their radiographic appearance is usually characteristic. The lesions are usually metaphyseal, expand the bone, have well-defined margins, evoke little reaction, and appear cystic with irregular septa. Sometimes a fragment of cortical bone (called *fallen leaf sign*) can be seen in the bottom of the cavity.

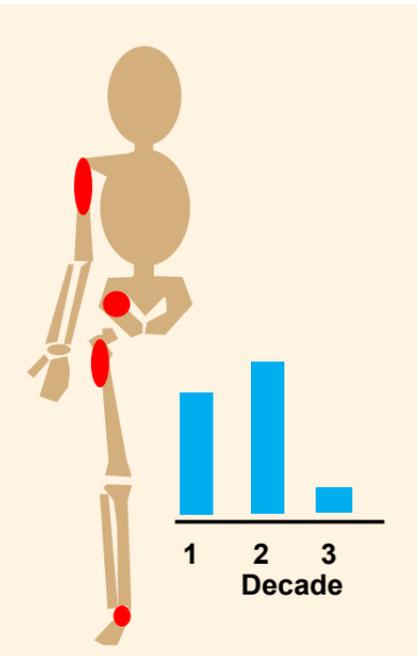
Active cysts abut the growth plate and occur in children less than 10 to 12 years of age. They are more likely to recur after treatment.

Inactive cysts are separated from the plate by normal bone and usually occur in adolescents over age 12 years.

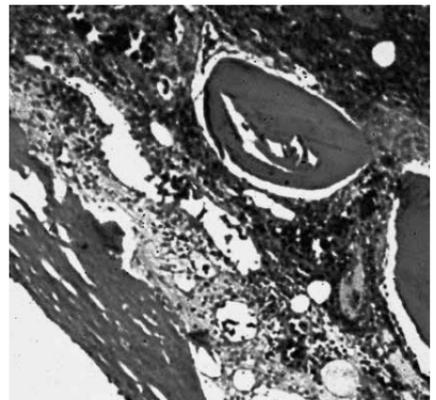
Fractures are usually the presenting complaint. Sometimes the fracture line is difficult to separate.

Management

Management is complicated by recurrence. The usual natural history of these cysts is to become asymptomatic following skeletal maturation. The objective of treatment is to minimize the disability when cysts are likely to fracture. These lesions are not precancerous.



1 Unicameral bone cysts. Common locations (red) and incidence per decade of life (blue) are shown.



2 Unicameral bone cyst lining. This shows the synovial lining of a cyst wall.

Humeral cysts Place the child in a sling to allow the fracture to heal and reestablish stability. Seldom does the effect of the trauma result in permanent healing of the cyst. Plan to manage the cyst by a series of injections [2] with steroid, bone marrow, or bone matrix. Some doctors recommend breaking up the adhesion by forceful injections or perforating the septa with a trochar. Recurrence can be managed by repeated injections or curettage and grafting with autogenous or bank bone. Opinions differ regarding how aggressively recurrence is managed.

Femoral cysts are much more difficult to manage. Plan to curette and graft the cyst and stabilize the fracture with flexible intramedullary fixation. Complications include malunion with coxa vara and avascular necrosis with displaced neck fractures. This fixation is permanent and may prevent additional fractures even if some recurrent cyst formation occurs. An alternative approach is injection followed by spica cast protection for 6 weeks.

Calcaneal cysts are best managed by curettage and bone grafting.

Complications

Recurrence and growth arrest of the upper humeral physis are most common. Growth arrest is usually due to the lesion, not the surgery. Recurrence is common and requires long-term thoughtful management.



1 Unicameral bone cysts. Note the typical active (red arrow) and inactive (yellow arrow) cysts with fractures. Flexible intramedullary rod fixation of an upper femoral cyst (orange arrow) is shown. A calcaneal cyst is an additional common site (white arrow).



2 Unicameral bone cyst. This is the classic location of this type of bone cyst. This 12-year-old boy developed pain in the right upper arm; a radiograph showed the typical cyst with a pathologic fracture (red arrow). The lesion was treated by steroid injection (yellow arrow) with satisfactory healing (orange arrow). A year later, the cyst has recurred (white arrow), but not to a degree that required additional treatment.

Aneurysmal Bone Cysts

An aneurysmal bone cyst (ABC) is considered to be a pseudotumor possibly secondary to subperiosteal or interosseous hemorrhage or a transitional lesion secondary to some primary bone tumor.

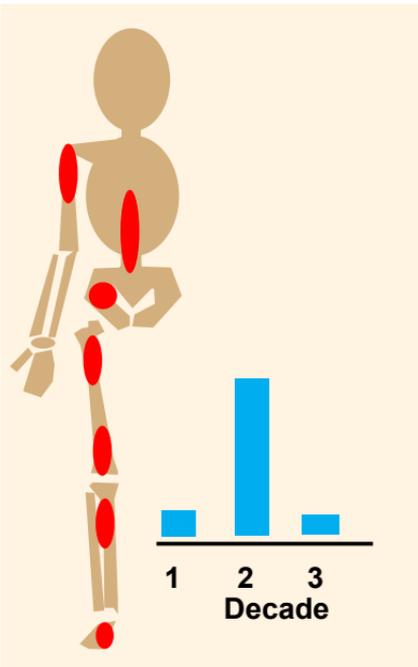
Diagnosis

The diagnosis can usually be established by a combination of the location of the lesion, the age of the patient [1], and the appearance on conventional radiographs [2]. ABCs are eccentric, expansile, cystic lesions. They often present with defined patterns [3 next page]. Sometimes the lesions are less eccentric and difficult to differentiate from simple bone cysts [1 next page].

Activity of the lesion The activity level can also be assessed by the appearance of the lesion's margins.

- *Inactive cysts* have intact, well-defined margins.
- *Active cysts* have incomplete margins but the lesion is well defined.
- *Aggressive cysts* show little reactive bone formation and poorly defined margins.

Other imaging is often necessary, especially in aggressive cysts. Fluid levels are common and can be seen on CT scans and MRI studies [2 next page].



1 Aneurysmal bone cysts.

Shown here are the common locations (red) and incidence per decade of growth (blue).



2 Aneurysmal bone cyst of the vertebral column.

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Management

Manage ABCs on the basis of the patient's age, the site, and the size of the lesion.

Age ABCs in early childhood are very likely to recur. Plan for this probability in discussing the procedure with the family.

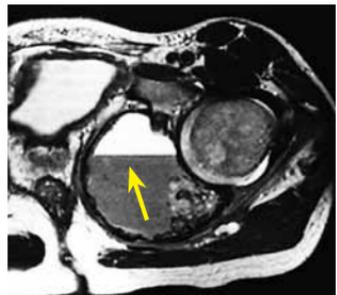
Spine About 10–30% of these lesions are in the spine. Study with CT and MRI preoperatively. The possible need for a combined approach, complete excision, and stabilization, as well as the risk of recurrence, complicates management.

Long bones Options include complete excision or saucerization leaving a cortical segment intact, or curettage with cryotherapy or with a mechanical burr. Consider prophylactic intramedullary rodding to prevent refracture.

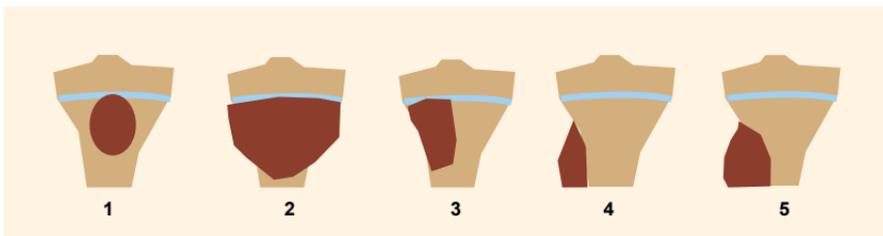
Pelvis Manage most lesions by curettage and bone grafting. Some recommend selective embolization. Be prepared for extensive blood loss.



1 Aneurysmal bone cyst of the upper femur.



2 Aneurysmal bone cyst of the pelvis. Note the extensive lesion (red arrow) and fluid level (yellow arrow) on the MRI.



3 Classification of aneurysmal bone cysts. Types 1–5 are various common patterns of lesions of long bones. Based on Capanna et al. (1985).

Benign Cartilagenous Tumors

Osteochondroma

Osteochondromas (osteocartilagenous exostoses) include solitary [1] and multiple lesions [2]. The multiple form is inherited but thought to be due to a loss or mutation of two tumor suppressor genes. Lesions sometimes develop after chemotherapy and radiation therapy. Most tumors develop by enchondral ossification under a cartilage cap.

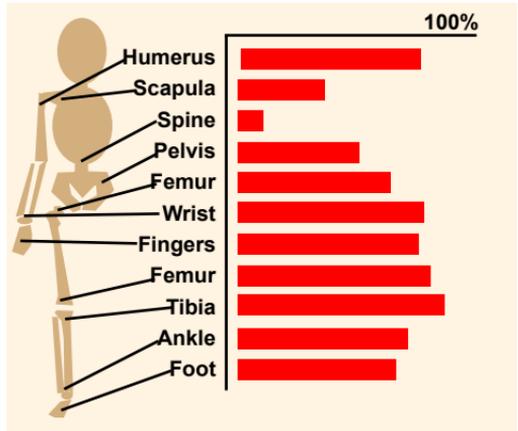
Diagnosis Osteochondromas are usually first noted as masses that are painful when injured during play [3]. These lesions are usually pedunculated but may be sessile. They may grow to a large size. Osteochondromas are so characteristic in appearance that the diagnosis is made by conventional radiographs.

Solitary Osteochondromas These lesions are most common in the metaphyses of long bones. They occur sporadically and present as a mass, often about the knee. Presentations in the spine may be associated with neurologic dysfunction.

Multiple Osteochondromas The common multiple form [1 next page] is inherited in an autosomal dominant pattern and is more common in boys. Multiple lesions about the wrist and ankle often cause progressive deformity [2 next page]. Others may cause valgus deformities about the knee.



1 Common locations of solitary osteochondromas.



2 Multiple familial osteochondromas. Note the widespread involvement. Based on data by Jesus-Garcia (1996).



3 Typical location for symptomatic lesions. Lesions about the knee are frequently irritated and painful (red arrows).

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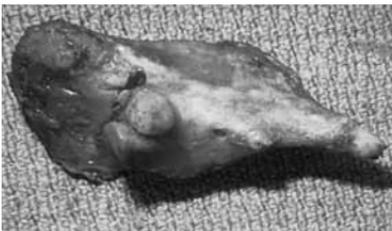
Management depends on the location and size of the tumor.

- *Painful mass* is the most common indication for removal [3]. Often several lesions are removed in one operative setting. Complications of excision include peroneal neuropraxia, arterial lacerations, compartment syndromes, and pathologic fracture.
- *Valgus knee* can be managed by medial femoral or tibial hemistapling in late childhood.
- *Limb length inequality* may require correction by an epiphysiodesis.
- *Ankle and wrist deformities* result from growth retardation of the distal ulna or fibula. Management of these deformities is complex and controversial.

Prognosis Very rarely malignant transformation to chondrosarcoma occurs during adult life. Because the transformation is very rare, prophylactic removal is not appropriate.



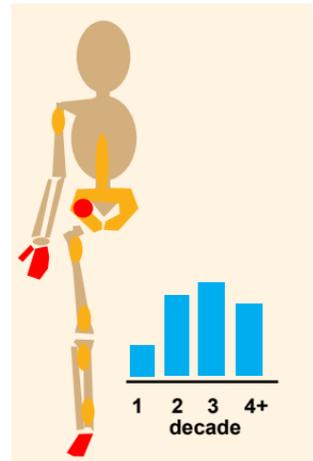
1 Multiple osteochondroma.
This child has multiple lesions (arrows).



3 Removed osteochondroma.
This resected lesion is large and irregular.



2 Common deformities that cause disturbed growth. These are common about the wrist (red arrow) and ankle (yellow arrow).



4 Enchondromas.
Common (red) and less common (orange) locations are shown. Age pattern of involvement is shown in blue.

Enchondroma

These cartilage tumors within bone are common in the phalanges and long bones and increase in frequency during childhood [4 opposite page]. These lesions may expand bone and produce the classic characteristic of cartilage tumors: speckled calcification within the lesion [1].

Types There are several different types of enchondromas.

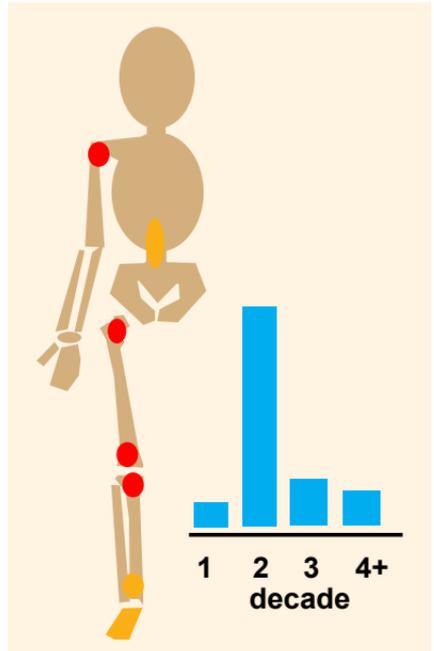
- *Solitary lesions* occur most commonly in the hands and feet. Removal and grafting is indicated if the lesions cause disability.
- *Ollier disease* is a generalized disorder with cartilagenous enchondromas as one feature. These children often have limb shortening and varus deformities [1]. About one-fourth develop chondrosarcoma in adult life.
- *Maffucci syndrome* is a rare disorder with subcutaneous hemangiomas and multiple enchondromas. Malignant transformation is usual in adult life.

Chondroblastoma

These uncommon tumors occur in the epiphysis of long bones often during adolescence [2 this page and 1 next page and cause an inflammatory reaction. They can be confused with infection or arthritis. They are aggressive and prone to recur. Treat by thorough curettage and possibly cryotherapy or phenolization and bone grafting. Avoid operative injury to the growth plate or articular cartilage. Expect a recurrence of about 20%.



1 Ollier disease. Note the extensive lesions of the distal femur and tibia (red arrows) with shortening and varus deformity.



2 Chondroblastoma. Common (red) and less common (orange) locations are shown. Age pattern of involvement is shown in blue. Based on Schuppers (1998).

Chondromyxoid Fibroma

This is a rare primary bone tumor that occurs most commonly about the knee during the second decade. The radiographic appearance is often characteristic with an eccentric position, a sclerotic rim with lobulated margins, and prominent septa. Manage with local resection and grafting.

Dysplasia Epiphysialis Hemimelica

Dysplasia epiphysialis hemimelica (Trevor disease) is a rare cartilagenous tumor that arises from the growth plate or articular cartilage [2]. The most common sites of involvement are the distal tibia and the distal femur. Lesions often involve one side of the epiphysis and may show multilevel involvement in the same limb. The diagnosis is often difficult early on, as the lesion is primarily cartilagenous and poorly imaged with conventional radiographs. MRI is helpful in showing the extent of the tumor and separating the lesion from the normal epiphysis or joint cartilage. Excise extraarticular lesions. Remove intraarticular lesions and correct secondary deformity with an osteotomy as necessary. Recurrence of the tumor is common, and multiple resections throughout childhood may be necessary.



1 Chondroblastoma. This is a greater trochanter lesion (arrow).



2 Dysplasia epiphysialis hemimelica. Note the swelling of the knee (red arrows) and ankle involvement (yellow arrow).

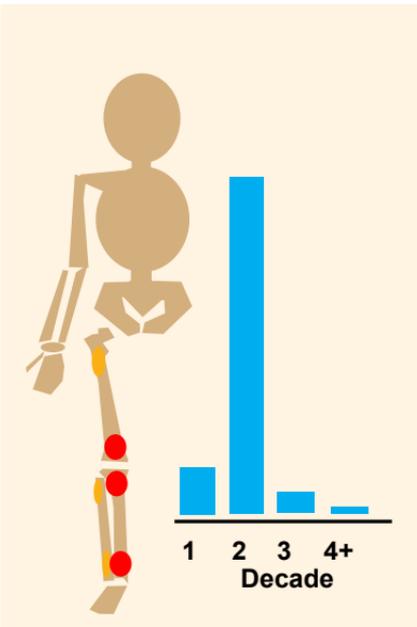
Fibrous Tumors

Fibrocartilagenous Defects

Fibrocartilagenous defects (or *fibrous metaphyseal defects*) are fibrous lesions of bone that occur in normal children, produce no symptoms, resolve spontaneously, and are found incidentally. They occur at insertion of a tendon or ligament near the epiphyseal growth plate, which may be related to the etiology. They have a characteristic appearance that is eccentric and metaphyseal, with scalloped sclerotic margins. These lesions often cause concern that sometimes leads to inappropriate treatment. Fortunately, the lesions have a characteristic radiographic appearance that is usually diagnostic. They are small in size, cortical in location, and well-delineated by sclerotic margins. They usually resolve spontaneously over a period of 1 to 2 years.

Nonossifying Fibroma

A larger version of the fibrocartilagenous defect is called a *nonossifying fibroma*. These lesions are present in classic locations and are usually diagnosed during adolescence [1]. They are metaphyseal, eccentric with scalloped sclerotic margins [2 and 3], and may fracture when large or if present in certain locations. Manage most by cast immobilization. Resolution of the lesion occurs with time. Rarely, curettage and bone grafting are indicated if the lesion is unusually large or if a fracture through the lesion occurs with minimal trauma.



1 Nonossifying fibroma. Common (red) and less common (orange) locations are shown. Age pattern of involvement is shown in blue.



2 Nonossifying fibroma of the distal femur.



3 Typical nonossifying fibroma.

Fibrous Dysplasia

Fibrous dysplasia includes a spectrum of disorders characterized by a common bony lesion. The neoplastic fibrosis replaces and weakens bone, causing fractures and often a progressive deformity. Ribs and the proximal femur are common sites, and the lesions are most common in adolescents [1].

Fibrous dysplasia can be monostotic or polystotic. The polystotic form is more severe and is more likely to cause deformity. This deformity is often most pronounced in the femur where a "shepherd's crook" deformity is sometimes seen [2] and may show extensive involvement of the femoral diaphysis. Rarely, fibrous dysplasia is associated with café au lait skin lesions and precocious puberty, as described with Albright syndrome.

Management of fibrous dysplasia is surgical. Weakened bone can be strengthened by flexible intramedullary rods. Leave these rods in place indefinitely to prevent fractures and progressive deformity [2].



1 Fibrous dysplasia. Common (red) and less common (orange) locations are shown. Age pattern of involvement is shown in blue.



2 Fibrous dysplasia of proximal femur. These patients show a femur at risk for deformity (red arrow), varus deformity (yellow arrow), and intramedullary fixation to prevent deformity (orange arrow).

Osseous Tumors

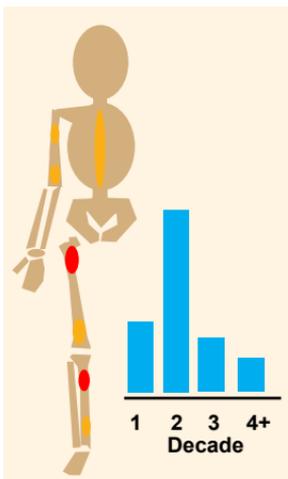
Osteoid Osteoma

This benign, bone-producing, highly vascular tumor induces an intense bony reaction and a characteristic pain pattern. These tumors occur most commonly in long bones during the second decade [1]

Diagnosis The pain occurs at night, is well-localized, and is often relieved by aspirin. Spine lesions occur in the posterior elements of the spine and often cause secondary scoliosis. Lesions are tender and, if close to a joint, cause joint inflammation that may be confused with primary arthritis. Lesions may cause hemideossification due to chronic pain and a limp [2]. The radiographic appearance is often characteristic for well-established lesions. A radiolucent nidus is surrounded by reactive bone [1 next page]. The bone scan is diagnostic, with intense localized uptake at the nidus.

Management New options for management supplement the traditional approach of open excision.

- *Antiinflammatory* Eventually, lesions resolve over many years. This option is rarely acceptable to families.
- *Percutaneous ablation* using CT is accomplished by removal, alcohol injection, thermocoagulation, or interstitial laser photocoagulation.
- *Open excision* is a standard practice.



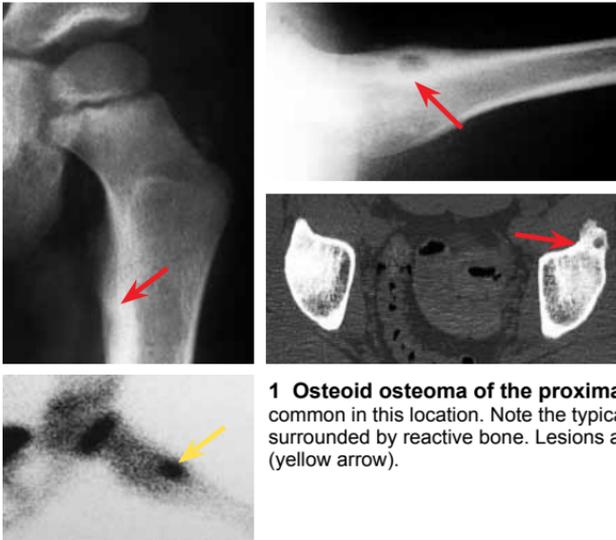
1 Osteoid osteoma. Common (red) and less common (orange) locations are shown. Age pattern of involvement is shown in blue.



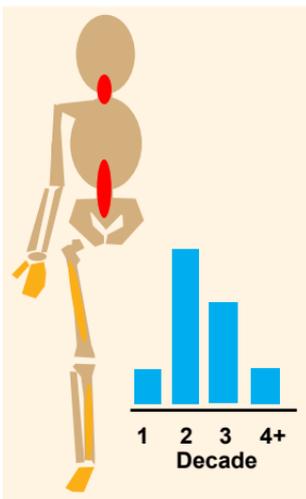
2 Osteoid osteoma. Note the hemideossification of the left pelvis and femur. This is due to pain and limp over a period of months from a proximal femoral lesion.

Osteoblastoma

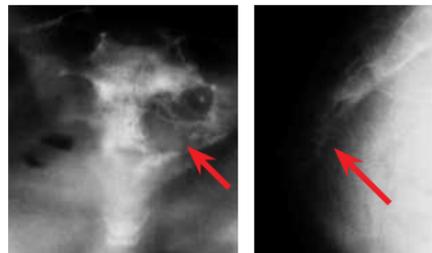
This benign bone-producing tumor is similar to the osteoid osteoma but larger. These lesions occur in the spine and long bones most frequently during the second decade [2]. One-third of these lesions occur in the spine [3], causing back pain and often scoliosis. They are sometimes difficult to differentiate from osteosarcoma. Spinal lesions are most difficult to manage because of the adjacent vertebral artery in a cervical spine lesion. Manage by complete resection. Expect a recurrence rate of about 10–20%.



1 Osteoid osteoma of the proximal femur. Lesions are common in this location. Note the typical nidus (red arrow) surrounded by reactive bone. Lesions are very “hot” on bone scan (yellow arrow).



2 Osteo-blastomas. Common (red) and less common (orange) locations are shown. Age pattern of involvement is shown in blue.



3 Osteoblastoma of the sacrum.

Miscellaneous Bone Tumors

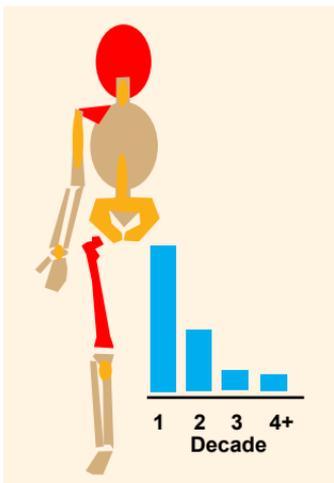
Eosinophilic Granuloma

Eosinophilic granuloma is the localized form of Langerhans' cell histiocytosis or histiocytosis X.

Diagnosis The peak age of onset is between 1 and 3 years of age [1]. Lesions are painful and are most often confused with osteomyelitis or sometimes Ewing sarcoma. Lesions often appear "punched out" on conventional radiographs [2 this page and 1 next page], but sometimes elicit periosteal reactions, suggesting a sarcoma. The child may have a low-grade fever and elevated ESR and CRP, making the differentiation from an infection difficult. Consider ordering skull films to aid in assessing a generalized disorder. Sometimes the diagnosis must be established by biopsy.

Management The natural history is of spontaneous resolution over a period of many months. Management options include simple observation, immobilization to improve comfort and reduce the risk of pathologic fracture, injection with steroid, limited curettage, or radiation treatment.

- *Spine lesions* cause collapse (*vertebra plana*) and sometimes neurologic involvement. Manage by observation or brace immobilization. Rarely, curettage is necessary to hasten resolution.
- *Lower limb long-bone lesions*, if large enough, may pose a risk of pathologic fracture. Curettage and cast protection may be appropriate.



1 Eosinophilic granuloma. Common (red) and less common (orange) locations are shown. Age pattern of involvement is shown in blue.



2 Eosinophilic granuloma of the scapula. This lesion is shown by CT scan (red arrow) and conventional radiograph (yellow arrow).

Giant Cell Tumors

Giant cell tumors (GCT) are aggressive tumors that fall between the usual classification of benign and malignant lesions. They occasionally occur in adolescents. Lesions are usually metaphyseal or epiphyseal, eccentric, expansive, and show little sclerosis or periosteal reaction [3]. These tumors are locally invasive and often recur. Manage by curettage and grafting. Provide careful follow-up because recurrence is common.

Neurofibroma

Neurofibromatosis causes widespread pathology, including scoliosis, pseudoarthrosis of long bones, thoracic lordoscoliosis, protrusio acetabuli, and abnormal bone growth (see details in Chapter 15).

Osseous Hemangioma

This is often present in the vertebrae or skull but may appear in the extremities. Lesions are diffuse and suggest a malignant tumor [2]. Wide resection is necessary and recurrence is common.



1 Eosinophilic granuloma in varied locations.



2 Hemangioma of bone. These lesions (arrow) are often difficult to differentiate from malignant lesions.



3 Giant cell tumor of bone. These lesions (arrows) occurred shortly after the end of growth. Note the lack of periosteal reaction.

Benign Soft Tissue Tumors

Hemangioma

Hemangiomas are common during childhood. They may be part of a systemic condition [1] or an isolated lesion [2].

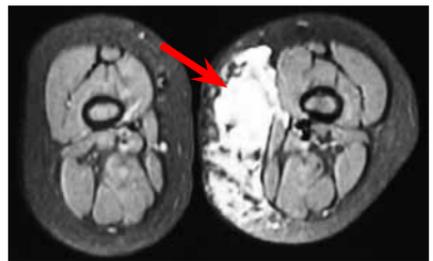
Diagnosis The clinical features depend on the location and size of the lesions. Subcutaneous lesions are usually locally tender. Intramuscular lesions cause pain and fullness, and very large or multiple lesions may cause overgrowth or bony deformity.

Imaging Punctate calcification in lesion is diagnostic. CT and MRI are most useful for diagnosis and preoperative planning.

Management Many patients are diagnosed clinically and treated symptomatically. Large and very painful lesions may require resection. Resection is often difficult, as the lesions are poorly defined and may be extensive. Recurrence is common.



1 Hemangioma. This boy has Klippel-Weber-Trenaunay syndrome with extensive hemangioma and limb hypertrophy (red arrow).



2 Extensive thigh hemangioma. This large lesion involves much of the medial thigh muscles (red arrows).

Synovial Hemangioma

Hemangioma of the knee is a cause of pain and recurrent hemarthroses in the pediatric age group [1]. The diagnosis may be delayed and the condition misdiagnosed as an internal derangement of the knee. Historically, long delays in diagnosis have occurred. Conventional radiographs show soft tissue swelling. MRI is usually diagnostic. Diffuse lesions are difficult to excise arthroscopically, and open wide excision is often required. Recurrence is common.

Plantar Fibroma

Fibromas may occur in infants with a lump on the anteromedial portion of the heel pad. Most remain small and asymptomatic, some disappear, and rarely do they persist and require excision.

In the child, plantar fibroma usually occurs as nodular thickening of the plantar fascia [3]. Observe to determine the potential for enlargement. Resect enlarging lesions. Be aware that mitotic figures are common in the specimen. Recurrence is frequent; overtreatment is common. Sometimes differentiating from fibrosarcoma or desmoid tumors is difficult.

Other Tumors

A variety of other tumors occur in childhood, including lipomas [2], lymphangiomas, and benign fibrous tumors.



1 Synovial hemangioma. This child had a swollen knee with frequent bloody effusions. Repeated resections over a period of many years were required.



2 Lipoma. Note the large lipoma (arrow) of the distal leg in a 16-month-old infant.



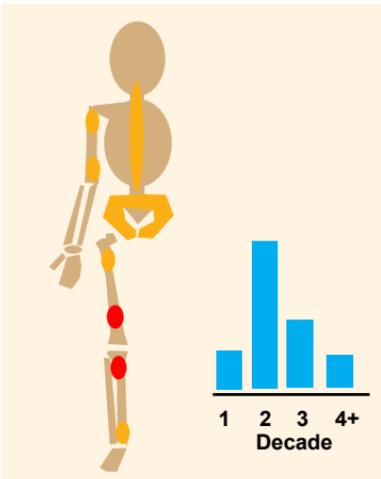
3 Plantar fibromatosis. Note the plantar thickening with overlying thickening of the skin.

Malignant Bone Tumors

Osteosarcoma

Osteogenic sarcoma is the most common malignant tumor of bone. Primary osteosarcoma occurs in children.

Diagnosis It commonly occurs during the second decade of life and often occurs about the knee [1]. Aching, nocturnal pain is often the initial complaint. Tenderness is often present, and a mass is sometimes palpable. Sometimes the patient presents with a pathologic fracture. Radiographs may identify either an osteolytic or osteogenic lesion of metaphyseal bone [1 next page]. Bone scans are helpful in identifying other affected sites. CT and MRI are helpful in assessing the osseous and soft tissue components of the lesion and in staging the tumor [2 next page]. The histology [2] shows tumor cells with primitive bone matrix formation.



1 Osteosarcoma. Common (red) and less common (orange) locations are shown. The age of onset in decades is shown in blue.



2 Pathology of osteosarcoma. This photomicrograph (left) shows tumor cell and primitive bone matrix formation. The gross specimen of the proximal humerus from an adolescent shows the intramedullary tumor (red arrow) and periosteal new bone formation (yellow arrow).

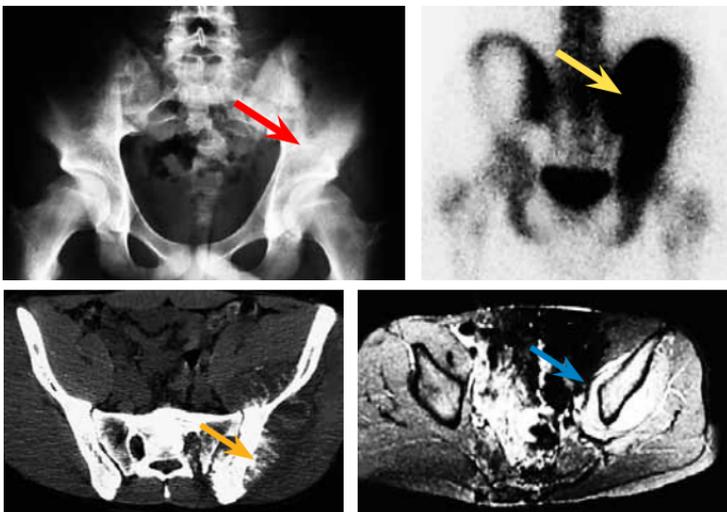
Management Chemotherapy followed by surgical eradication of the tumor is necessary. Manage the tumor by amputation or local resection. Limb salvage can be done by replacing the resected bone with an allograft or metallic implant. Rotationplasty includes lesion resection and rotational reconstruction, substituting the ankle for the knee. Limb salvage during growth results in significant limb shortening. This can be managed by procedures designed to spare the growth plate, expandable prosthetic implants, or contralateral epiphysiodesis. At present, the overall 5-year survival rate is about 75% following aggressive chemotherapy and resection.

Variants Osteosarcoma has several types.

- *Parosteal osteosarcoma* These well-differentiated lesions develop on the surface of the bone, such as the posterior femoral metaphysis, with little or no medullary involvement. Manage by wide local resection.
- *Periosteal osteosarcoma* develops on long tubular bones, especially the tibia and femur. In contrast to parosteal osteosarcoma, periosteal osteosarcoma is less differentiated, resulting in a poorer prognosis.



1 Varied radiographic appearance of osteosarcoma. Lesions may be destructive (red arrow), osteogenic (yellow arrow), cause a moth-eaten appearance (orange arrow), or show combined osteoblastic and lytic features (white arrow).



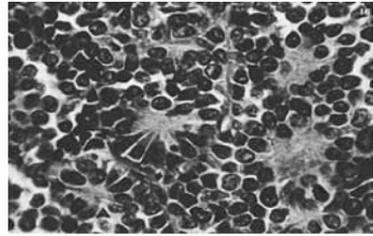
2 Osteogenic sarcoma of the pelvis. Note that the lesion is not readily identified on conventional radiographs (red arrow), but it is well imaged by bone scan (yellow arrow), CT (orange arrow), and MRI (blue arrow).

Ewing Sarcoma

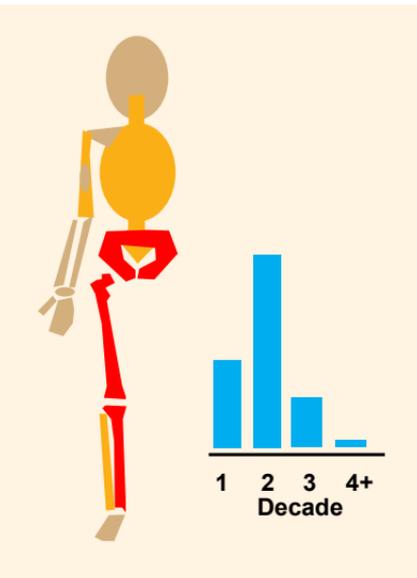
Ewing sarcoma is the second most common childhood malignant bone tumor. The tumor is most common in the second decade and occurs most commonly in the pelvis, femur, and tibia [1]. This is a very malignant round cell tumor [2].

Diagnosis These tumors cause pain and often present with a large soft tissue mass. The lesion is usually diaphyseal [3] and osteolytic or permeative in character. Bone scans and MRI are useful [1 next page]. Because the tumor may cause fever, leukocytosis, anemia, and an elevated sedimentation rate, it can be confused with osteomyelitis. Confirm the diagnosis by biopsy.

Management Treatment by chemotherapy and resection has increased the 5-year survival rate into the 70% range. This malignancy differs from osteogenic sarcoma by a greater likelihood of metastasizing to bone and a lower survival rate.



2 Pathology of Ewing sarcoma. Note the cortical destruction and extracortical extension in the proximal femur. This photomicrograph shows small round cell tumor cells.



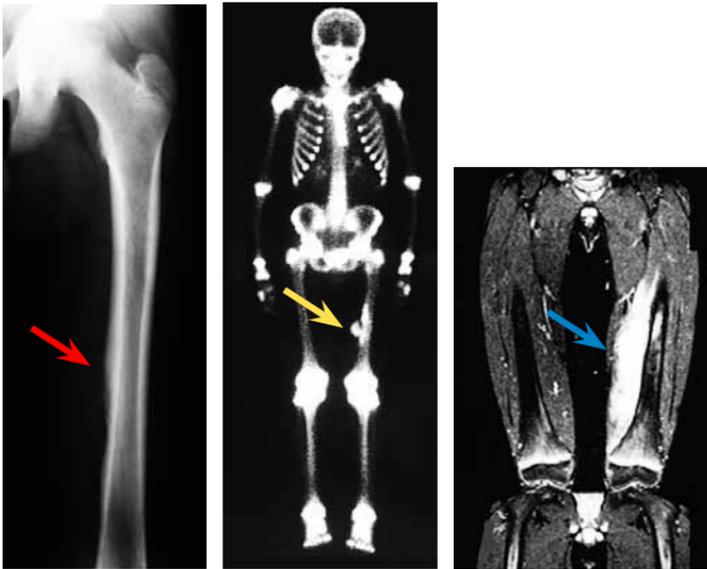
1 Ewing sarcoma. Common (red) and less common (orange) locations are shown. The age of onset in decades is shown in blue.



3 Typical radiographic features of Ewing sarcoma. Note the diaphyseal location with periosteal reaction.

Leukemia

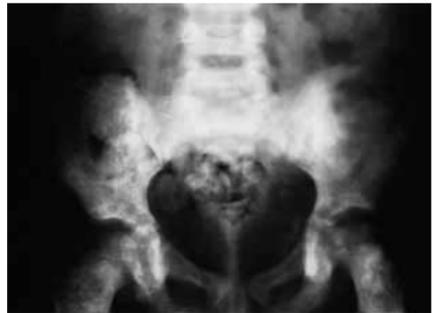
About 20% of children with leukemia present with bone pain and may first be seen by an orthopedist or rheumatologist. Common findings include bone pain, joint pain and swelling, antalgic gait, mild lymphadenopathy and hepatosplenomegaly, and a moderate fever. Radiographic findings include diffuse osteopenia, metaphyseal bands, periosteal new bone formation [2], sclerosis, and a combination of sclerosis and lytic features. Usual laboratory findings include an elevated ESR, thrombocytopenia, anemia, decreased neutrophils, increased lymphocytes, and blast cells on the peripheral blood smear. Confirm the diagnosis with a bone marrow biopsy.



1 Ewing sarcoma. Note the diaphyseal location (red arrow), the positive bone scan (yellow arrow), and extensive soft tissue involvement (blue arrow).



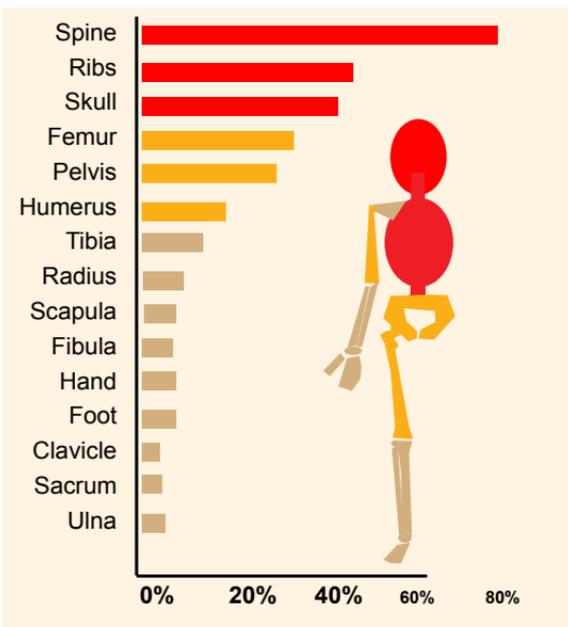
2 Leukemia. Note the periosteal bone of the proximal ulna (arrow).



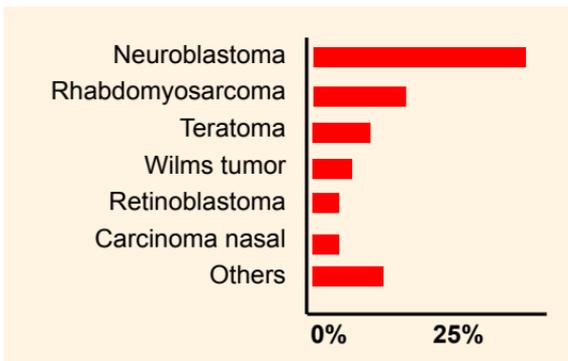
[3] Metastatic neuroblastoma. Note the extensive metastases in the pelvis and proximal femora.

Metastatic Bone Tumors

Metastatic tumors to bone are most likely to involve the axial skeleton [1]. The most common primary tumors are neuroblastoma followed by rhabdomyosarcoma [2]. Vertebrae metastases are most common in the lumbar spine, while thoracic and cervical involvement are less common. The primary site of tumors with spinal involvement are neuroblastoma and astrocytoma. Complications of spinal metastasis include paralysis, pathologic fractures, and kyphoscoliosis. Assess children with neuroblastoma for bony metastatic disease by CT, MRI, scintigraphy, or bone marrow biopsy. Extensive bony involvement is a relatively late finding [3 opposite page].



1 Site and frequency of skeletal metastasis. From data of Leeson et al. (1985).



2 Tumors producing skeletal metastasis. From data of Leeson et al. (1985).

Malignant Soft Tissue Tumors

These tumors account for about 7% of malignant tumors of childhood. About half are rhabdomyosarcomas. These soft tissue malignancies are divided into five general categories [1].

Rhabdomyosarcoma

This is a sarcoma of the skeletal muscle. It is the most common pediatric soft tissue sarcoma. Extremity tumors account for 20% and carry a poorer prognosis.

Lesions are firm, nontender, and within the muscle compartment. Tumors occur in childhood, and metastasize to lymph nodes and later to bone [2]. Manage by total excision and chemotherapy. Expect the 5-year survival in the 65–75% range.

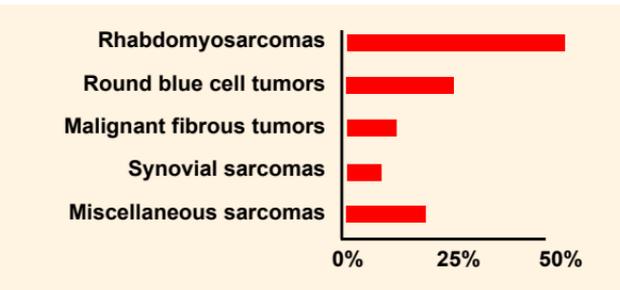
Round Blue Cell Tumors

These tumors include primitive neuroectodermal tumors, soft tissue Ewing sarcoma, and Askin tumors. Askin tumors are round-cell tumors involving the central axis and chest wall.

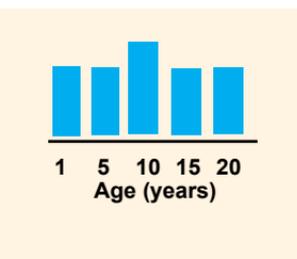
Malignant Fibrous Tumors

Desmoid tumors, or fibromatosis, are sometimes considered to be benign. However, because of their tendency to recur, they are sometimes considered as low-grade fibrosarcomas. Most occur in the extremities, creating a soft tissue mass and sometimes erosion or deformity of the adjacent bone [1 opposite page]. The natural history of fibromatosis is variable; lesions often recur and undergo spontaneous remission. Fibromatosis seldom metastasize or cause death.

Manage by total resection when possible. If surgical margins cannot be achieved without sacrificing the limb or its function, excisional resection is an acceptable alternative. The role of adjuvant chemotherapy or radiation is controversial.



1 Soft tissue sarcomas. From data of Conrad et al. (1996).



2 Rhabdomyosarcoma age distribution. These tumors are common in infancy and early childhood. They can extend to bone (arrow).

Synovial Sarcoma

These tumors occur most commonly in adolescents [3] and adults. Most occur in the lower extremities. Primary metastases are usually to regional lymph nodes or the lungs. Manage by chemotherapy, nonmutilating resection, and radiation. Expect 70–80% survival.

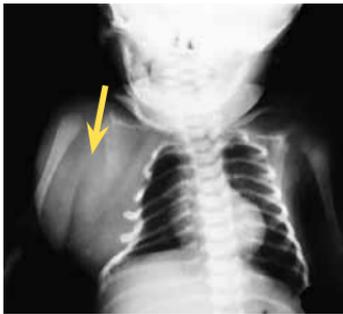
Miscellaneous Sarcomas

Peripheral nerve sheath sarcomas Malignant degeneration occurs in 5–10% of patients with neurofibromatosis (NF1). Enlarging lesions in these patients should be documented by MRI and excised or biopsied.

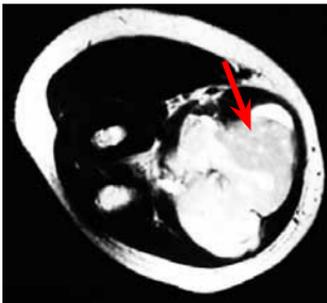
Other sarcomas These include a variety of tumors [2]: leiomyosarcoma, liposarcoma, angiosarcoma, and many others.



1 Desmoid. Note the large soft tissue mass (red arrow) and deformation of the second proximal phalanx (yellow arrow).



2 Soft tissue malignancies in infants. This liposarcoma (red arrow) and fibrosarcoma (yellow arrow) have become massive lesions.



3 Synovial sarcoma. This lesion in a 16-year-old boy involving the elbow joint is poorly seen on conventional radiographs but readily imaged by MRI (red arrow).

Chapter 12 – Neuromuscular Disorders

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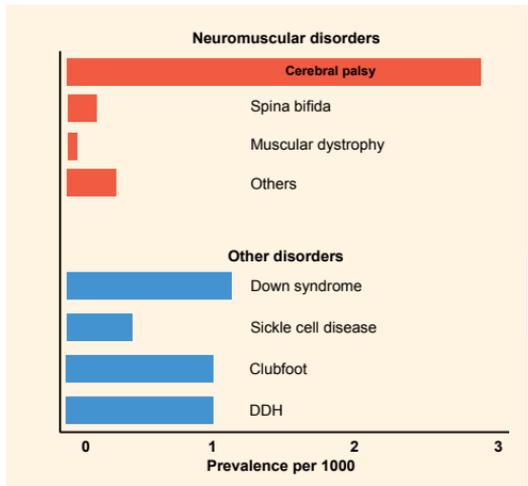
Neuromuscular disorders [1] are the most common cause of chronic disability in children. Because motor dysfunction is often an early manifestation of these diseases the orthopedist may be the first to see the child. The child with in-toeing may have mild spastic hemiparesis or the boy whose walking deteriorates in early childhood may have muscular dystrophy.

Statistics

Prevalence of cerebral palsy far exceeds other disorders [2] with an estimated 750,000 affected individuals in United States alone. Poliomyelitis is still a problem in developing countries [1 on page 435].



1 Neuromuscular disorders. Children with neuromuscular disorders such as cerebral palsy (left) and myelodysplasia (right) are common.



2 Prevalence of neuromuscular disorders in North America. This graph shows the relative frequency of each disorder and a comparison with other conditions causing disability in children.

Evaluation

An accurate diagnosis is essential for effective management. Classify the condition first by anatomical level and then match the clinical and laboratory findings with the various diagnostic possibilities to establish the diagnosis [1 this page and 2 opposite page].

Family History

A family history is often positive in peripheral neuropathies and muscular dystrophy.

Medical History

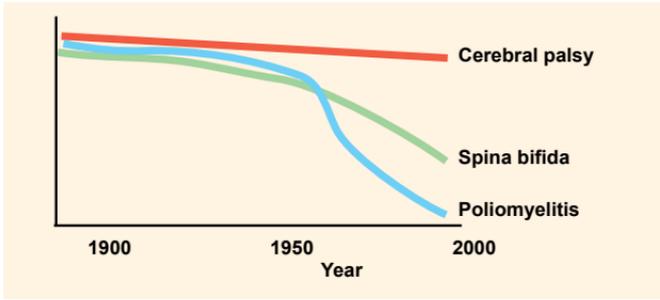
The time of onset is helpful. Mothers of children with neurological problems often note something unusual about the pregnancy or early infancy. The mother may note less than expected or late fetal movement during pregnancy. The pregnancy may be abnormal, delivery prolonged, birth weight low. The Apgar score may be low. Feeding problems, respiratory difficulties, and delay in acquiring motor skills are common during infancy. Mothers often sense that something is wrong during pregnancy or during early infancy. The mother’s intuition is surprisingly accurate.

Acquisition of Motor Skills

Infants with neurological impairment usually show a variety of abnormal findings including abnormal reactions, retention of primitive reflexes, and delays in acquiring motor skills. The most useful and consistent finding is a delay in motor development. The normal infant shows head control by about 3 months, sitting by 6 months, standing with support by 12 months, and walking independently by about 14 months [3 opposite page]. Although considerable individual variability exists, gross delay in developing these skills is worrisome.

<p>Upper motor neuron</p> 	<p>Spastic paralysis Increased tone Hyperreflexia Persisting primitive reflexes MRI abnormalities Normal laboratory values Sensation perception altered</p>
<p>Anterior horn cells</p> 	<p>Decreased tone Hyporeflexia Flaccid paralysis Variable EMG Nerve conduction normal</p>
<p>Peripheral nerves</p> 	<p>Distal weakness Hypotonicity – hyporeflexia Family history sometimes abnormal Nerve conduction slowed Sensation often altered</p>
<p>Muscle</p> 	<p>Family history often abnormal Proximal weakness Hypotonicity CPK high in some EMG abnormal Nerve conduction normal</p>

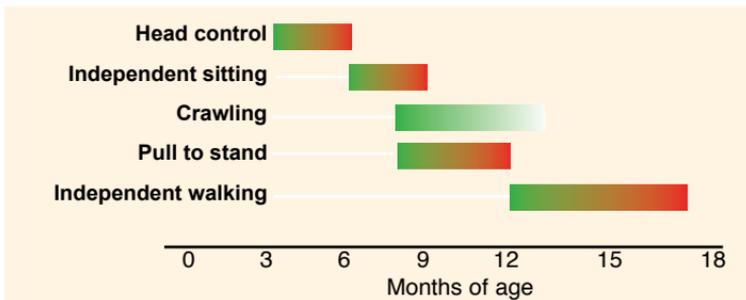
1 Physical and laboratory features by level. Each level of pathology has unique features that are useful in assessment.



1 Incidence of neuromuscular disorders with time in North America. Dramatic declines in incidence of poliomyelitis and spina bifida are shown.

				
Cause	Upper Motor Neuron	Anterior Horn Cell	Peripheral Nerves	Muscle
Congenital	Cerebral palsy Spina bifida Mental retardation	Diastematomyelia	Insensitivity pain	Congenital myopathy Arthrogyposis Absent muscles
Degenerative-Inherited		Friedreich ataxia Syringomyelia Spinal muscle atrophy	CMT Herniated disc	Muscular dystrophy Myotonia
Infectious, toxic inflammatory	Meningitis Encephalitis	Poliomyelitis Transverse myelitis Guillain-Barré syn.		Myositis Collagen disorders Dermatomyosities
Tumors	Brain and cord	Cord	Neurofibromatosis	
Vascular	Aneurysms	Ant. spinal artery syn		
Traumatic	Head injury		Obstetrical palsy	Torticollis

2 Disorders by level of pathology.



3 Normal range of motor development. Motor milestones are reached on an average (green) and then 95% at end of bar (red).

Physical Examination

Give special attention to the neurological examination of infants suspected of have a neuromuscular problem. Perform a screening examination. See page 20. Observe the child running. Gait disturbances that are minimal during walking become pronounced when the child runs.

Assess strength Does the infant show signs of weakness? If possible watch the child walk up and down stairs. Observe the child arising from a position on the floor. The Gower maneuver is a classic test for weakness seen in muscular dystrophy [1]. In the older child assess strength of specific muscle groups. For example, assess foot eversion strength if Charcot Marie Tooth disease is suspected. Grade muscle strength by standard criteria. See page 39.

Assess tone Tone is often altered in these children.

- *Hypotonus* Infants with reduced tone are often described as *floppy infants*. There are many causes of hypotonia [2]. Head control is poor, motor development is delayed, feeding and respiratory problems are frequent. Often infants with cerebral palsy show hypotonia in early infancy and spasticity later.
- *Hypertonus* Increased tone may be due to spasticity or rigidity. Spasticity is most common and characterized by hyperreflexia, increased resting tone, an exaggerated response to suddenly applied stretch, and tone that increases when the child is stressed or upright positioned.

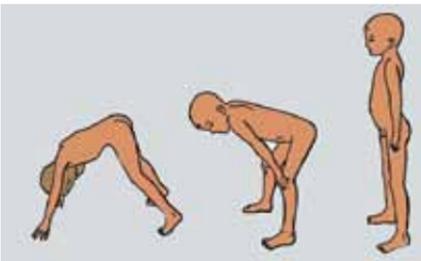
Sensation

Sensory abnormalities are classic in myelodysplasia and in the sensory neuropathies. Less appreciated are the sensory alterations that occur in cerebral palsy where the pathways are intact but appreciation and integration of the sensory input is impaired.

Deformity

Deformity may be dynamic, fixed, or a combination.

Dynamic deformities causes abnormal position or function without structural muscle shortening. For example, scissoring or a crossing of the legs may occur before fixed adduction contractures develop. An equinus position may precede the development of fixed triceps contractures. Assess the severity of dynamic deformity by noting the posture of the child while held in the upright position. The upright position increases tone that accentuates dynamic deformity.



1 Gower sign. Note the characteristic maneuver to stand from a position on the floor.



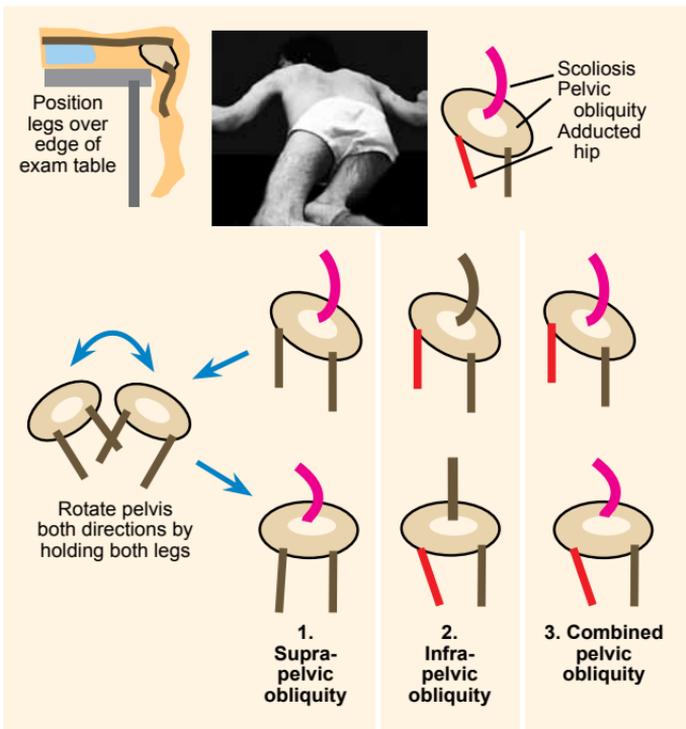
2 Floppy infant. Note the lack of head control.

Fixed contractures are due to shortening of the muscle-tendon complex. This shortening occurs over a period of months following the onset of dynamic contractures due to spasticity as seen in cerebral palsy. Shortening occurs more slowly, over a period of years, from chronic positioning of children with flaccid paralysis. An example is the development of hip flexion contractures in children with myelodysplasia who sit all day. Finally contractures of the capsular or ligaments about joints develop most slowly over a period of several years.

Combined contractures Most contractures have elements of both dynamic and fixed types. The proportion of fixed to dynamic contracture gradually increases with time.

Progression Fixed contractures eventually cause joint deformity. The articular surfaces of the joint first become deformed. Later joint subluxation and sometimes dislocation develop. Scoliosis, hip subluxation or dislocation, or flattening of the femoral condyles from lack of normal motion are examples of the late effect on joints of fixed contractures.

Pelvic obliquity Assess pelvic obliquity with the child positioned over the edge of the examination table [1]. Assess the effect of rotating the pelvis in each direction to determine whether the deformity causing the obliquity has its origin in the spine (suprapelvic), in the pelvis (pelvic) or from hip deformity (intrapelvic). Sometimes deformities are combined and complex.



1 Assessing cause of pelvic obliquity. Position the child with the legs over the end of the examining table. Rotate the legs as a unit from one side to the other. 1. Suprapelvic obliquity is present if the spine remains deformed and the pelvic-thigh position becomes neutral. 2. Intrapelvic obliquity is present if the spine straightens but the thigh remains adducted. 3. Combined deformity is present if deformities of the spine and adduction of the thigh persist when the pelvis is rotated in either direction.

Management Principles

Management of neuromuscular problems is often complex. This complexity is due to the great variety of disorders, varied individual response to disease, and often the lack of effective treatment methods. Application of general principles of management often simplifies and improves management.

Natural History of Deformity

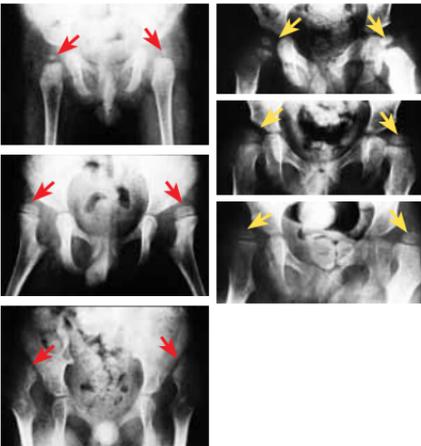
Neuromuscular disorders cause disability not only from weakness, altered sensation, and incoordination but also from acquired deformity. These acquired deformities develop in a definite sequence [1]. For example, the child with cerebral palsy is first hypotonic and then becomes spastic. This spasticity causes dynamic deformities that becomes a fixed contractures with time. These fixed contractures cause altered loading of joint cartilage, disturbed growth, and finally bony deformity. These acquired deformities limited function and mobility further increasing disability. Understanding this sequence of progressive deformity and disability is important in planning management.

Effectiveness of Orthopedic Treatment

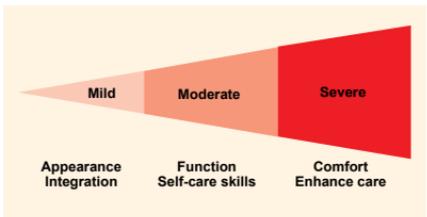
Musculoskeletal procedures in neuromuscular disorders focus on the secondary problems of spasticity, contracture, and bony deformity. As our management can only alter the *affects* rather than the *cause* of the disease, treatment is not definitive.

Focus Management on Comfort, Function, and Appearance—Not Deformity

The objective of management is to improve comfort, function, and appearance, not to correct deformity. Individualize management based on the severity of the disability [2]. For example, in the severely disabled child the first priorities are to enhance comfort and to facilitate care. The objective is to correct only those deformities that interfere with these priorities.



1 Natural history of hip deformity in cerebral palsy. With adduction contractures, hips gradually dislocate with time. Note the sequence from ages 1, 4, and 10 years (red arrows) in a child. Note less symmetrical progression in another child at 1, 2, and 3 years (yellow arrows).



2 Progression of disability. Increasing severity causes disability to increase from appearance to function and finally causes pain.

Appreciate the Significance of Sensation and Perceptive Disabilities

Loss of sensation or perceptive impairments are often underrated in importance. The child with cerebral palsy is described as having *spastic paralysis*. Often the focus is on the *deformity* alone. Equally important but less obvious are sensory and perceptive disabilities. For example, the child with arthrogryposis who has more deformity but intact sensation functions at a higher level than a child with cerebral palsy with less deformity but impaired sensation. Recurrent ulcers of an insensate foot often cause more disability than the motor weakness for the child with myelodysplasia.

Establish Appropriate Priorities

Adults with disability rank communication and socialization above mobility in importance. Frequently the family's major concern is whether or not their child will walk. Help the family understand that the long-term objectives are independence and social integration. During each visit attempt to keep the focus on the essential needs of the child such as self-care capabilities and communication and away from trivial problems. For example, efforts to overcome minor variations in gait may waste energy and resources needed to help the child develop independence in self care, communication skills, and effective mobility.

Provide Effective Mobility

Establish effective, age-appropriate, independent mobility [1]. The 2-year-old child with normal mental development should have a means of mobility controlled and initiated by the child. This mobility is often a combination of methods, which may include rolling, crawling, and walking using splints, a walker, or a wheelchair—whatever works for the child. Mobility must be practical, effective, and energy efficient. Mobility is the goal; the method is much less important. Children with effective mobility develop intellectual and social abilities most rapidly. Provide a wheelchair early if necessary to enhance mobility. Children do not become *addicted* to wheelchairs.

Focus on Self-Care

During early and midchildhood, focus on self-care. Assess the child's self-care abilities – eating, dressing, and toileting. Order occupational therapy with specific objectives. Avoid simple stretching programs that waste time and accomplish little.

Focus on Assets

Often children with disability have talents that need to be identified. Time spent developing the child's assets may be more productive than attempting to overcome the child's disability.



1 Effective mobility. Various mobility aids allow the child to be independent.

Shift Priorities with Age

Focus on mobility early, self-care in early childhood, school, and socialization [1] in midchildhood and early teens and on vocational training [2] in late teen years. Make these age priorities.

Maintain Family Health

Avoid exhausting the family with nonessential or unproven treatments. Be aware that the strength of the marriage and the well-being of the siblings are important [1 opposite page] for the handicapped child. Recognize that all treatments have a price for the child and family. Ineffective treatments deprive the child of important play time and energy and waste the family's limited resources.

Treat the Whole Child

The objective is a child who meets its potential both emotionally and physically. Play is the occupation of the child. The child with a disability has need of play just as other children. Preserve time and energy for this experience. The individual is a child only once.

Avoid Management Fads

Attempt to steer the family away from interventions that are unproven or unrealistic. Such treatments sap the resources of the family and lead to eventual disappointment for the child. History of cerebral palsy management includes a vast number of treatments that were either harmful or ineffective. Extensive bracing, misguided operations, and exhaustive therapies are examples of treatments once in vogue but later abandoned. Often the outcome of treatment is studied and if the treatment is found ineffective, it is replaced by a new treatment, which is applied until it too is found unsatisfactory. Results are often poor because *cause* of the neurological problem cannot be corrected with current management techniques.

See Child Several Times before Deciding on Surgery

Delay major management decisions such as surgical procedures until the child is seen several times. This delay provides an opportunity to better understand the family and allows evaluation of the child in different situations. If after each assessment the surgeon's decision is the same, the odds for a successful outcome are improved.



1 Socialization skills. Integration and play with other children are essential childhood experiences and of special importance to those with disability.



2 Vocational planning. During the second decade the need for vocational training becomes a major priority.

Consider Regression

Motor regression following surgery is greatest following major procedures that require prolonged immobility in the older child. Be very cautious about performing procedures in late childhood or adolescence in marginal walkers as the child may never regain their preoperative walking ability.

Prevent Complications

Take active measures to prevent skin ulcers, pathological fractures, and motor regression.

Gait Lab Comparison with Normals

Be cautious about using laboratory data [2] that compares data based on normal children as controls. The objective of management is not to create normal laboratory values but to enhance function.

Develop Appropriate Recreational Programs

Tailor recreational programs to the child. Often these children do better in individual rather than team sports. Special olympics and wheelchair basketball are examples of appropriate team sports.

Help Family Find Support Groups

Support groups provide information, perspective, and friendship, which can be invaluable for the family.



1 Family health. A healthy family is especially important for a child with a disability.



2 Gait laboratory evaluation. Be aware that these evaluations should be viewed as only one part of a comprehensive assessment.

Cerebral Palsy

Cerebral palsy (CP), or static encephalopathy, is a nonprogressive central nervous system disorder that causes perceptual and neuromotor disability with an onset in infancy or early childhood. Most CNS insults are pre- or perinatal in origin [1]. CNS injury may cause a variety of clinical problems [2].

Etiology

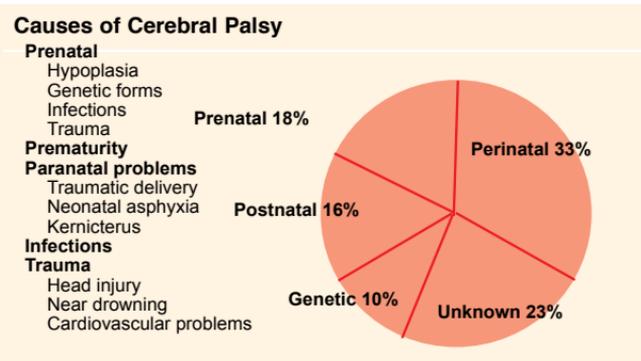
CP is an inclusive diagnosis with an extensive list of causes [1]. Common causes include trauma, infection, and toxins. Common associations include prematurity, low Apgar score, difficult delivery, and neonatal illnesses.

Pathology

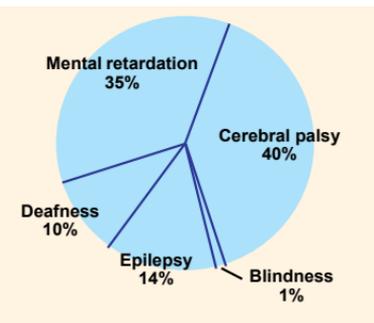
Most infants with CP show pathologic changes in the brain that correlate poorly with the clinical features. Focal and generalized involvement of the cortex, basal ganglia, and brain stem result from ischemic damage, atrophy, agenesis, gliosis, and degenerative changes. MR imaging shows abnormalities, which include periventricular leukomalacia in preterm infants, a variety of abnormalities in term infants, and extrapyramidal cerebral palsy shows lesions in the putamen and thalamus.

Evaluation

Consider conditions that may be confused with cerebral palsy. Progressive diseases such as spinal cord or brain tumors are sometimes slow growing and can be confused with CP. Demyelinating, degenerative or familial (spastic paraplegia) disorders must be ruled out. If any question exists, refer to a neurologist to confirm the diagnosis.



1 Causes and timing of onset of cerebral palsy.



2 CNS insult outcomes.



3 Spastic cerebral palsy. Note the adductor spasticity with the scissoring.

Tone

Spasticity is increased tone with passive stretch. This response is greatest if the stretch is applied quickly. This is the common form of cerebral palsy [3 opposite page].

Rigidity is an increased resistance to passive stretch that is independent of the speed of application. Rigidity may be a uniform (lead pipe) or intermittent (cog wheel) type. This is uncommon in cerebral palsy.

Athetosis is characterized by involuntary movement. The combination of athetosis and spasticity is referred to as *tension athetosis*.

Ataxia is a loss of muscular coordination and balance.

Dystonia is an intermittent distorted posturing.

Ballismus is an uncontrolled involuntary motion.

Distribution

Cerebral palsy is usually classified by distribution of involvement [1] Be aware that on careful examination the so-called uninvolved extremity often shows subtle abnormalities. Slight upper limb involvement is seen in diplegia and opposite limb abnormalities in hemiplegia. Side-to-side asymmetry is also common in spastic quadriplegia and diplegia.

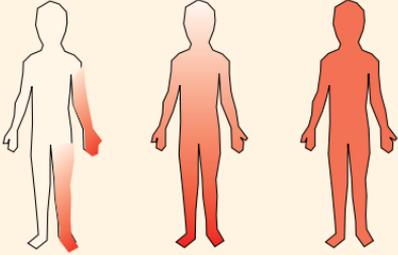
Monoplegia is uncommon and affects a single limb.

Hemiplegia is common and affects the limbs on one side. The upper limb are often more involved. Hemiparesis is the mild form.

Diplegia is common, and affects the lower limbs more than the upper limbs. This pattern is sometimes called *paraplegia*.

Triplesia is uncommon and involves three limbs.

Quadraplegia, or total body involvement, is common and most severe.



Features	Hemiplegia	Diplegia	Quadraplegia
Frequency	Common	Common	Common
Disability	Mild to moderate	Moderate	Severe
Feet	Equinovarus	Equinovalgus	Equinovalgus
Knees	Mild flexion	Moderate flexion	Severe flexion
Hips	OK	May dislocate	Early dislocation
Spine	OK	Usually OK	Scoliosis
Upper Limbs	Distal	Little involvement	Flexion deformity
Hands	Flexion deformity	Little	Flexion deformity
Seizures	Common	Rare	Common

1 Classification of cerebral palsy based on distribution of involvement.

Deformity

Deformity changes during childhood as dynamic deformity becomes fixed with time [1]. Examine the child in both the supine and upright position. Dynamic deformity changes with positioning and stress. The physical examination increases stress, increasing tone and dynamic deformity. Separate dynamic deformity from fixed contracture. Assess fixed deformity by gentle and prolonged stretch with the patient as relaxed and comfortable as possible [2]. Record severity in degrees. Assess the torsional profile, pelvic obliquity, and spinal deformity.

Supine measurements are made first.

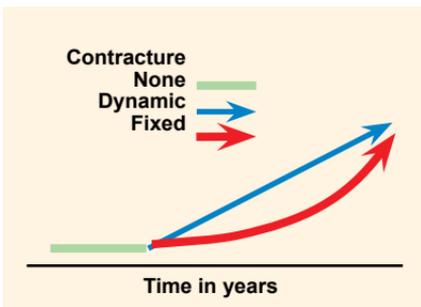
- *Ankle dorsiflexion* Measure with the knee flexed and extended ankle neutral.
- *Knee extension* Measure any loss of extension.
- *Popliteal angle* Assess with hip flexed as a measure of hamstring contractures.
- *Hip abduction* Assess using pelvis landmarks as bases of reference.

Prone measures

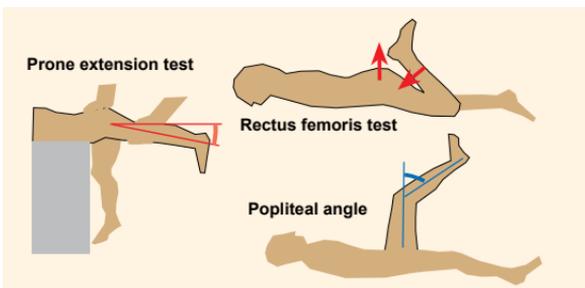
- *Rotational profile* includes hip rotation, thigh-foot angle and shape of foot. See page 70.
- *Prone extension test* for assessment of hip flexion deformity. See page 131. This test has been shown to be more reliable than the Thomas test in cerebral palsy.
- *Rectus femoris test* is performed by slowly flexing the knee and noting how much elevation of the buttocks occurs due to secondary hip flexion.

Other Measures

The value of assessing reflexes, reactions, and patterns is helpful for the very experienced clinician in developmental medicine [1 and 2 opposite page]. For most orthopedists, assessing motor development, head control, sitting, crawling, standing, and walking is most practical.



1 Effect of time on contracture formation. In early infancy hypotonia is often present and no contracture develops (green). With the onset of spasticity, dynamic deformity (blue arrow) develops. With time, this dynamic contracture becomes fixed (red arrow).



2 Assess contractures. These are common tests to assess contractures in cerebral palsy.

Management Options in Cerebral Palsy

Management is challenging as the disease is complex, extensive, permanent, and varied. Manage with the perspective that the long-term success places priorities on communication, socialization [1 next page], independence, and, finally, mobility. Optimal management requires considered application of the best of many choices of interventions for the particular child. Avoid ineffective treatments as they are harmful to the child and draining on the family’s energy and resources.

Therapy is valuable in assessment, providing family support, facilitating bonding, improving self-care skills, providing infant stimulation, promoting use of adaptive equipment, and facilitating family interaction with the child.

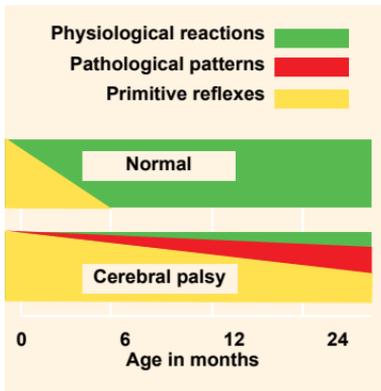
- *Neurodevelopmental therapy (NDT)* has been shown to be equal to infant stimulation programs and is declining in use.
- *Inhibitory casting* benefit is derived from immobilization in a functional position.

Adaptive equipment includes standing devices [2 next page] and aids for self-care. These devices are often very practical and effective in improving function.

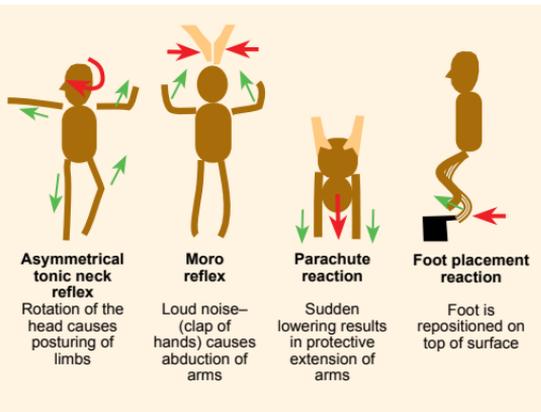
Mobility aids include wheelchairs [3 next page], carts, and motorized devices, which allow the child greater independent mobility.

Cast correction is effective as a temporary means of overcoming recently acquired fixed contractures.

Night splinting is sometimes useful in managing the child following surgery to prevent early recurrence and provide comfort.



1 Natural history of reactions, patterns, and reflexes. Note that primitive reflexes are replaced by normal reactions during normal development. In cerebral palsy, primitive reflexes and pathological patterns persist. These differences are useful for diagnosis determining severity of involvement.



2 Common reflexes and reactions. The test is initiated (red arrow) and the responses (green arrows) indicate a positive response.

Botulinum toxin injections into the muscle belly provide 3–6 months of reduction in muscle tone. The high cost of the agent and short duration of effect limit its value of this treatment.

Intrathecal baclofen Continuous infusion reduces spasticity in the upper and lower extremities, and improves upper extremity function and activities of daily living. Complications from the intrathecal catheter occur in 20% of patients and infection in about 5%.

Rhizotomy Selective dorsal rhizotomy is effective in reducing spasticity, especially in spastic diplegia. Orthopedic procedures are still required in about 70% of patients but usually should be postponed for 1–2 years after rhizotomy.

Orthotics have very limited usefulness. The ankle-foot orthosis (AFO) is the only brace shown to be useful [4]. Whether the ankle is articulated or solid is controversial.

Musculoskeletal surgery is appropriate to improve function, reduce discomfort, or facilitate care. Deformity alone is not an indication for surgery. Recurrent deformity is common.



1 Socialization and integration. The child with cerebral palsy needs the same experiences as other children.



2 Promoting upper extremity function. This may be done by positioning to free the hands and by occupational therapy to improve hand function.



3 Effective motility. This must be individualized. Wheelchairs often increase independence by making mobility safe, convenient, and energy efficient. This fortunate boy has a canine assistant (red arrow) who is not only a friend but enhances the boy's independence.



4 Braces and casts. These provide the child with stability following surgery and facilitate walking.

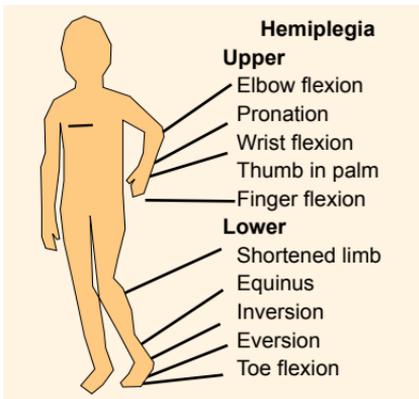
Hemiplegia

The spectrum of severity is broad. Sometimes a child is seen for intoeing or clumsiness and found to have mild hemiplegia or hemiparesis. The family may not have been aware of any underlying neurologic problem.

Clinical Features Contractures are most severe distal in the limb. Typical deformities include equinus and varus or valgus feet, and flexed elbow, wrist, fingers, and adducted thumb [1]. Proximal joints have less consistent involvement. Scoliosis is uncommon. Limb shortening is mild and proportional to severity. Function is generally fair and proportional to severity. Walking is slightly delayed. The involved hand disability is proportional to overall severity. Learning disability, seizures, and social problems are common. Sensory deficits are more significant than deformity in limiting hand function.

Management Tailor management based on the severity of the disability. Mainstream children when possible. As hand function requires sensation and fine motor function, disability in the upper limb is most severe.

- *Upper limb* encourage use of hands early. Value of early stretching, splinting, casting are controversial. Base operative indications on level of discriminatory sensibility, intelligence, motivation, and overall function. Delay operative correction until at mid or late childhood [2]. Transfer muscle under voluntary control to improve finger, thumb, or wrist extension. Fuse joints for stability.
- *Lower limb* Limb shortening insufficient to require epiphysiodesis. Patterns of involvement vary considerably [3] and require that management be individualized. Triceps, posterior tibialis, and hamstring lengths often necessary at about 4–6 years of age.



1 Deformities in hemiplegia. Note that the deformities are mostly distal in the limbs.



2 Operative correction of thumb-in-palm deformity. Note the improved correction following surgery (arrow).

Type	Clinical pattern	Underlying abnormality
1	Minimal, drop foot pattern	Anterior tibialis weakness
2	Flexed knee	Tight gastrocnemius
3	Flexed knee and hip	Tight gastro and hip flexors
4	Knee hyperextension	Tight soleus
5	Knee and ankle hyperextension	Abnormal fore–aft shear force

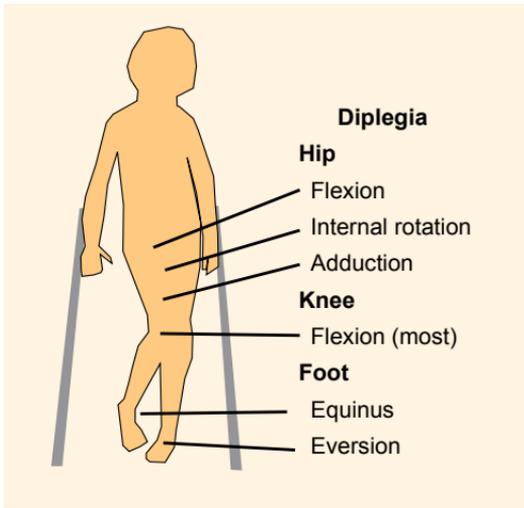
3 Gait patterns in hemiplegia. From Hullin (1996).

Spastic Diplegia

Spastic diplegia [1 and 2] is the most common form of cerebral palsy. About twothirds are associated with prematurity. Spasticity usually develops during the second year. Motor development is delayed but gradually improves to about age 7 years [3]. Independent walking usually occurs if the infant achieves a motor level of about 12 months by the chronological age of 36 months.

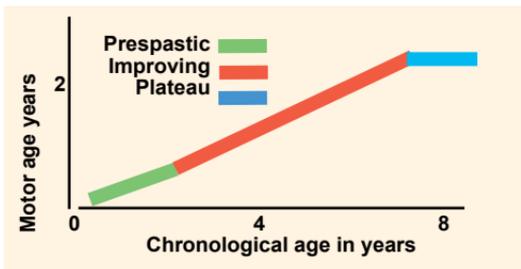
Clinical features include typical deformities of the lower limbs. Mild upper limb involvement is detected by careful examination. Severity varies. Often involvement is asymmetrical. Perform standard examination. Assess for hip subluxation and cause of a crouch gait [1 opposite page].

Management Consider providing walking aids, ankle-foot orthoses (AFOs), monitor the status of the hips for subluxation. This may require operative treatment. Operative procedures commonly indicated are adductor releases between 3–5 years, hamstring lengthening procedures between 6–10 years, and heel-cord lengthening after age 7. When possible correct equinus with gastrocnemius lengthening alone. Femoral rotational osteotomy may be helpful in correcting intoeing in the older child.



1 Common deformities in spastic diplegia.

2 Spastic diplegia.



3 Motor development in diplegic children. Based on Beals (1966).

Quadraplegia

Quadraplegia, or total body involvement, may be symmetrical or asymmetrical. Rarely, when asymmetrical with one limb minimally affected, the pattern is sometimes called *triplegia*.

Clinical features

Children with quadraplegia show greatest impairment in speech, mentation, nutrition, and self-care abilities, and are most demanding of caregivers. Deformities may be extensive [2].

Management

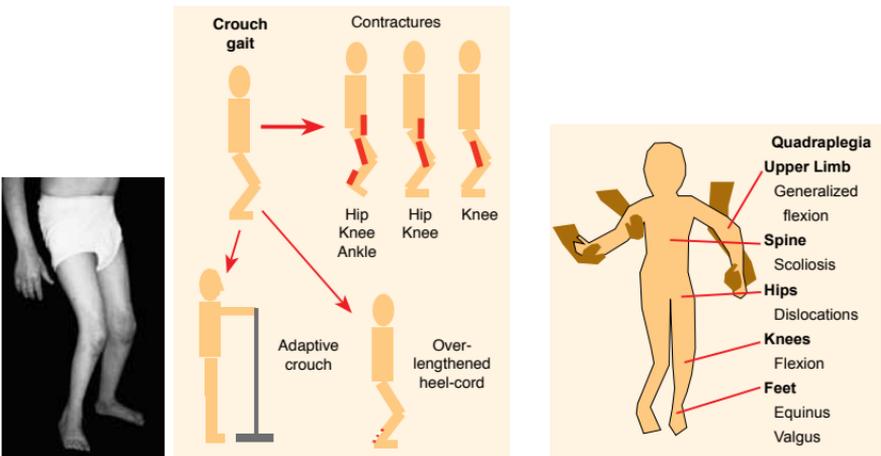
Management is usually best provided in a center that provides multidisciplinary care. The goals are to maximize comfort, self-care, and independence. Functional walking is uncommon. Major orthopedic problems include multiple, severe extremity deformities, hip dislocations, and scoliosis.

Multiple Severe Contractures

Severe deformities may cause considerable disability and may require operative release. Postoperative management may be complicated by severe pain that increases spasticity that may cause recurrent deformity. Following release, immobilize for comfort and plan night splinting to prevent recurrence. The results of these release procedures is only fair.

Hip Subluxation–Dislocations

Natural history The hip is normal at birth. Dynamic adduction and flexion deformity develops during late infancy and converts to fixed contractures during early childhood. These contractures and increased tone cause progressive subluxation of the hip and erosion of the lateral acetabular margin. Dislocation often occurs during midchildhood. As the dislocation becomes fixed, femoral head deformation occurs, and if unilateral, infrapelvic obliquity develops. The relationship between hip dislocation and scoliosis is inconsistent. Hip dislocations cause pain, pelvic obliquity, and complicate care.



1 Crouch gait. Determine the site(s) of contractures (red lines). Adaptive crouch position improves with support. An overlengthened heel-cord (dotted red line) causes a crouch without contractures.

2 Common deformities in quadraplegia.

Management Attempt to interrupt the progression of the deformity by early adductor lengthening procedures. Perform procedures bilaterally for to enhance symmetry and to reduce the risk of recurrent deformity [1]. Management depends upon the severity of deformity [2], and the child’s age and functional level.

Scoliosis

Scoliosis is most common in severely affected children. Scoliosis usually develops independent of hip dislocations and may cause suprapelvic obliquity.

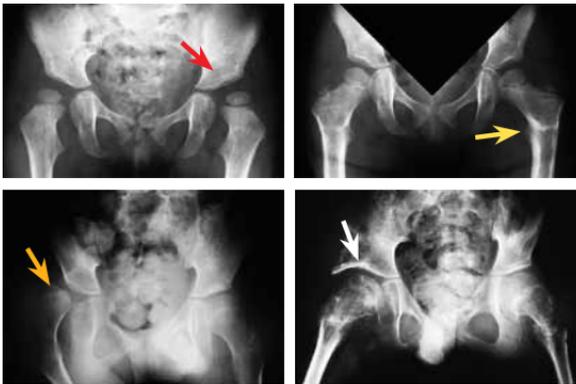
Progression Risk factors for progression include spinal curve of 40° before age 15 years, total body involvement, being bedridden, female gender, and thoracolumbar location. Progression after skeletal maturation is greatest if curves >50° with progression of about 1.5° per year.

Curve patterns fall into several categories [1 opposite page].

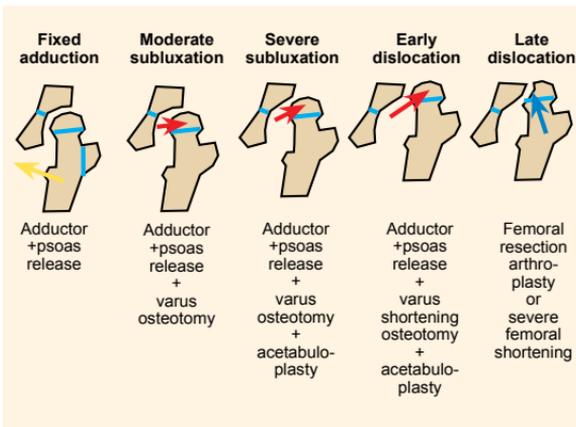
Disability is due to problems with sitting and care. Scoliosis is seldom a cause of decubiti or pulmonary compromise.

Orthotics and exercise treatments are not effective in altering the progression of the curve.

Operative correction maybe indicated for progressive severe curves to prevent progression and provide stability for sitting. Posterior fusion is adequate for curves below about 70°. Large curves require combined anterior-posterior releases, instrumentation, and fusion.



1 Hip dislocation in quadraplegia. Subluxation was noted at age 3 years (red arrow). A varus osteotomy was performed (yellow arrow) at age 5 years. At age 8, right hip subluxation developed (orange arrow), which was managed by bilateral femoral osteotomies and a shelf procedure on the right (white arrow).



2 Hip dysplasia and management. Hip deformity in cerebral palsy is usually progressive. With only adduction deformity (yellow arrow) soft tissue release is appropriate. Tailor the extent of bony correction to the severity of subluxation–dislocation and acetabular dysplasia (red arrows). Salvage long-standing dislocation with femoral head deformity (blue arrow) by a resection type of arthroplasty.

Athetosis

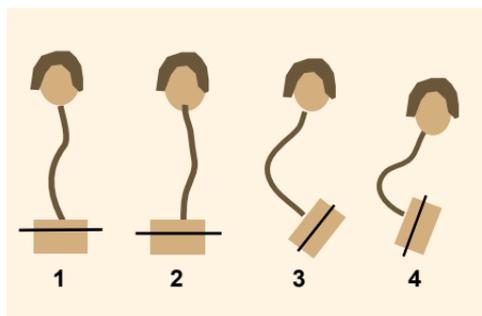
Athetosis has become uncommon because of improvements in obstretrical practices and neonatal care.

Clinical features

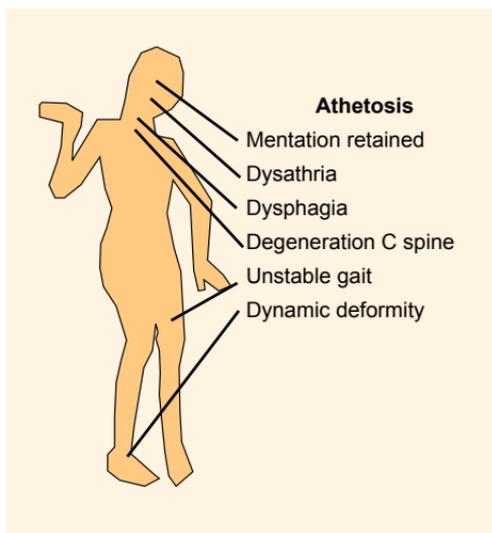
Athetosis causes dyskinesia with involuntary movement and labored volitional function [2]. Because of the excessive movement, contractures are uncommon except in combined forms, which include an element of spasticity. Scoliosis may develop. Intelligence is often normal.

Management

Traditional orthopedic procedures are usually not required. Provide effective mobility with an electric wheelchair, and adaptive equipment to facilitate self-care and independence. Speech and occupational therapy are most helpful. Teach computer skills and use of adaptive devices early.



1 Curve patterns in cerebral palsy. Based on Lonstein (1994).



2 Problems in athetosis.

Complications of Cerebral Palsy

Nutritional deficiencies

Nutritional problems should be resolved before severely affected patients undergo extensive releases or spinal surgery. Children with serum albumin <3.5 mg% and lymphocyte count of <1500 cells/cc are at risk for postoperative infections and for a prolonged hospitalization.

Postoperative Pain Syndrome

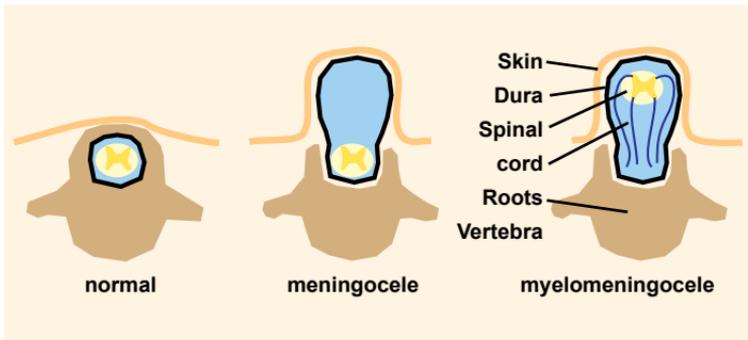
This distressing complication commonly occurs when children are mobilized following extensive operative procedures. Pain, irritability, increased tone, and recurrent deformity may occur. The pain may be due to stretch on the sciatic nerve following hamstring lengthening. Manage by gradual mobilization, pain relief, sedation, and patience.

Pressure Sores

Reduce the risk by applying extra cast padding, utilizing careful technique in cast application, relieving pressure over bony prominences, and periodically inspecting the skin.

Aspiration

Severely affected children in spica casts when positioned supine are at risk for aspiration. Prone or side-lying positioning may prevent this complication. In poorly nourished children, consider placing a nasogastric tube before surgery.



1 Pathology of meningocele and myelomeningocele.



2 Myelomeningocele. These lesions may be large. In the past (red arrow) large lesions were not repaired. Currently repairs are performed in the neonatal period (yellow arrow).

Myelodysplasia

Myelodysplasia is part of a spectrum of deformities [1 opposite page] resulting from failure of closure of the neural tube late in the first month following conception [2 opposite page and 1 this page].

Etiology

Factors include genetic, geography, and drugs (valproic acid and carbamazepine). The prevalence is reduced by periconceptional folic acid supplementation. The severity of paralysis is reduced by cesarean delivery before rupture of amniotic membranes and onset of labor.

Pathology

The spectrum of defects is broad with localized or extensive neurological defects.

Brain defects include hydrocephalus, and the varied Chiari malformations that may include herniation of the cerebellum into the upper cervical canal [2]. Mental retardation is most common in higher level lesions [1 next page].

Cord defect Lesions may be incomplete or complete, open or closed, and occur at different levels. An autonomous functioning cord below the primary lesion may result in segmental spasticity. Diastatomyelia, tethered cords, lipoma, and syringomyelia may be coexisting problems. The level of the defect usually determines the neurological level and pattern of musculoskeletal deformities [2 next page].

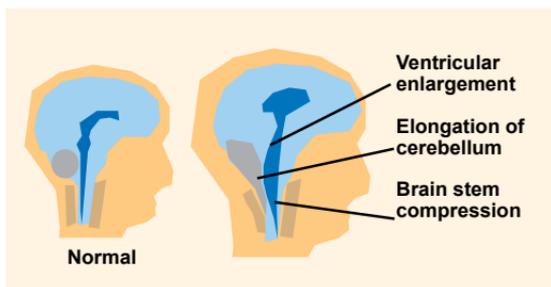
Multiple system involvement is common. Major problems include incontinence, urinary infections, and nutritional problems.

Bone and joint deformity are common associated problems. These include clubfeet, vertical talus, knee flexion or extension deformities, hip dislocations, and spinal kyphosis or scoliosis.

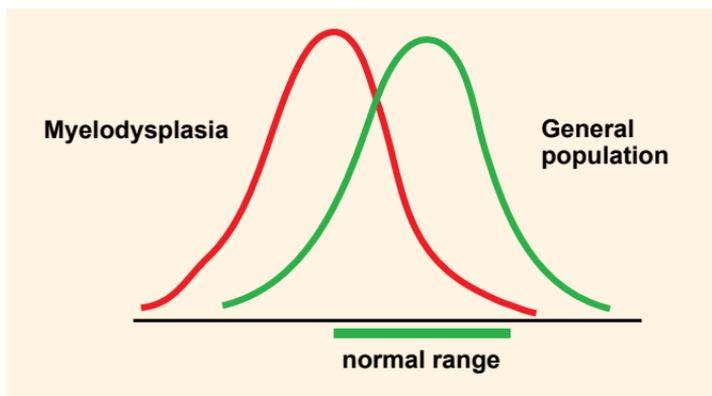
Secondary skeletal defects often develop with time and muscle imbalance. These include hip dislocations, progressive scoliosis, and lower limb deformities.



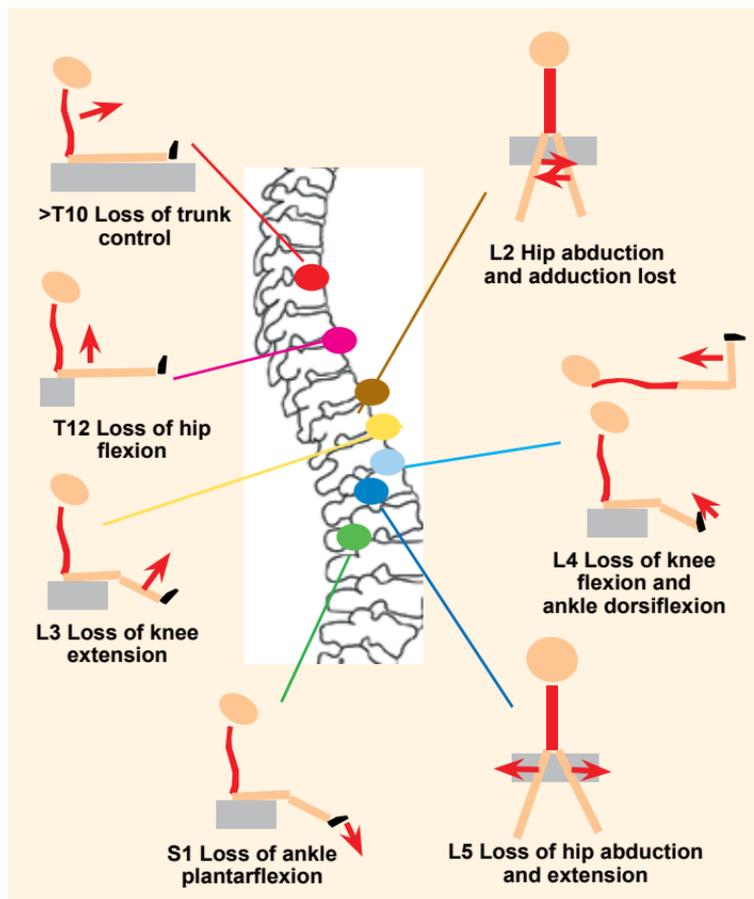
1 Spina bifida. The vertebral defect includes widening of the canal (yellow arrows) and loss of posterior elements.



2 Chiari malformation. This deformity is common in myelodysplasia and includes several characteristic features as shown.



1 Intelligence in myelodysplasia.



2 Weakness related to level of spinal defect. These tests are useful in determining the level of the defect. Function is lost distal to the defect. Typical functional losses for each level are shown.

Clinical Features

Prenatal diagnosis is by alpha-fetoprotein determination and ultrasound examination.

Initial assessment is best provided in a multidisciplinary facility with neurodevelopmental, neurosurgical, urologic, and orthopedic consultants available. Make certain management is whole-child oriented. Determine neurological level by neurological examination and muscle testing. Unilateral or incomplete lesions improve prognosis. Often the orthopedic concerns are the least of the child's problems.

Periodic evaluations are necessary throughout childhood [1]. During each visit, assess overall function, the spine and pelvis for symmetry, the skin for ulcers, and address specific problems identified by the parents.

Hip deformity

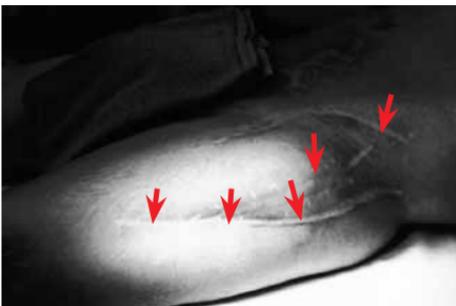
Hip deformity is related to the neurological level. In the past hip deformity was often over-treated [2].

Flexion contractures increase with time and may require release.

Hip dislocations are most common in upper lumbar paralysis with muscle imbalance. Hip dislocation itself does not affect the ability to walk. Operative indications include painful dysplasia in the ambulatory patient and fixed pelvic obliquity, which makes sitting difficult or skin care unmanageable. Operative complications including recurrent deformity, stiffness, pathological fractures, and skin ulcers are common.



1 Evaluation throughout childhood. Plan to see the child periodically throughout childhood. Become acquainted with the family.



2 Ineffective management. Hip reduction procedures are seldom helpful as recurrence is common, reoperations are frequent, complications are distressing, and functional improvement is minimal. This patient had several procedures (red arrows) with little or no improvement.

Spinal Deformity

Spinal deformity is most common in the more severely affected child. Progression is most likely when associated with cord tethering or hip contractures.

Kyphosis is usually congenital and may be severe [1 and 2]. Resection is indicated if the deformity prevents skin closure during neonatal repair of the defect or later if skin breakdown occurs over the apex of the deformity.

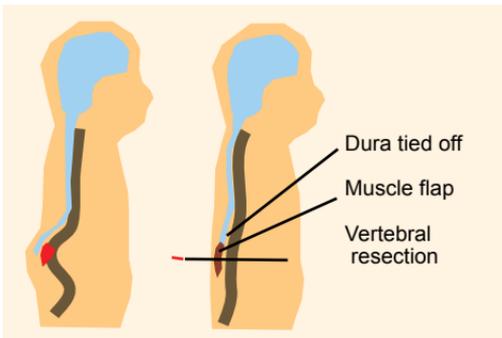
Scoliosis may cause suprapelvic obliquity increasing the risk of decubitus ulcer formation, sitting problems, and impaired hand function. Correct severe or progressive deformity and level the pelvis to distribute skin loading evenly under the pelvis. Manage with a focus on disability rather than deformity. Be aware that poor soft tissue coverage, contractures, impaired sensation, fragile bone, and deficient posterior elements complicate treatment. Pseudoarthrosis rates are decreased with combined anterior–posterior fusions and rigid segmental instrumentation.

Knee Deformity

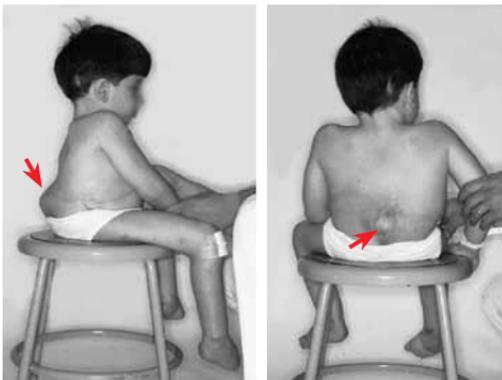
Knee deformities may be congenital or develop during childhood.

Flexion deformity may make walking difficult or impossible. Correct significant disability with soft tissue releases, which often requires a posterior capsulotomy. Extension osteotomy may be necessary in the older child or adolescent.

Extension deformity may complicate sitting. Release persisting congenital contractures by early percutaneous release.



1 Kyphosis repair. Repair usually requires bony and soft tissue procedures.



2 Lumbar kyphosis. This prominence makes the overlying skin vulnerable to breakdown.

Foot Deformities

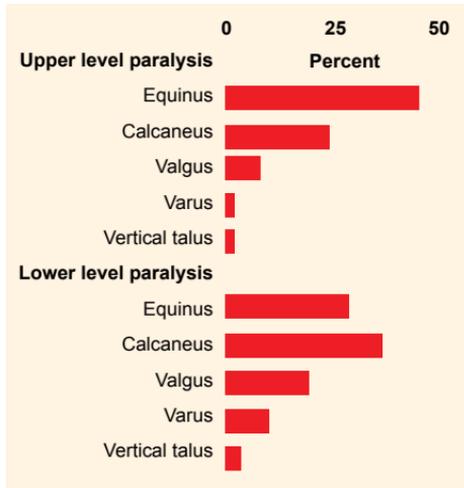
Foot deformities are common for both upper- and lower-level lesions [1]. They occur in nearly 90% of upper-level paralysis and 60–70% of those with lower lesions. Spastic segments are common in high level lesions and cause progressive deformity.

Operative procedures on the foot are often necessary, effective, and successful. Foot ulcers are common problems if the foot is non-plantigrade and stiff. Attempt to preserve motion and create a plantigrade position for standing or for positioning the footrests of a wheelchair. Avoid arthrodesing procedures whenever possible.

Calcaneus deformity results from relative overpull of the anterior tibi-
alis [2]. Manage with an orthotic, anterior tibialis and ankle release,
tenodesis of the tendo-achilles, or transfer of the anterior tibialis to the
Achillis tendon.

Clubfeet are common and require operative correction at about a year
of age. Correct by extensive posteromedial release or talectomy. Recur-
rent deformity may require a decancellation procedure. Avoid arthrodesing
procedures. Residual varus may be a problem [3].

Ankle valgus is secondary to a triangular distal tibial epiphysis. Manage
by placing a medial malleolar screw that restricts medial physeal growth
to correct the valgus. Remove the screw when the ankle is neutral [1 next
page]. A valgus deformity at the end of growth may require a distal tibial
osteotomy for correction.



1 Foot deformities in high-and low-level myelodysplasia. From data of Broughton (1994) and Frawley (1998).



2 Calcaneus deformity.



3 Foot deformities complicating wheelchair sitting. Correction of foot deformities is necessary whether the child walks or is mobile in a wheelchair.

Planovalgus foot is best managed by calcaneal lengthening, which provides correction and maintains foot mobility.

Vertical talus deformities require operative correction during the first year. Correct with a single-stage procedure.

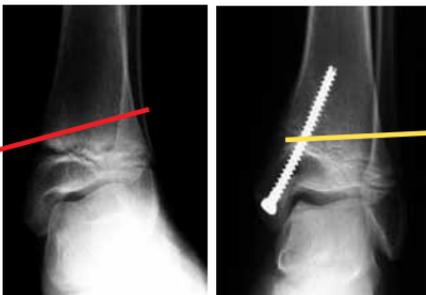
Cavus deformity often causes skin breakdown and significant disability. Manage in two stages. First perform a plantar medial release. Follow with osteotomies and tendon transfers to maintain correction.

Toe deformities include dorsal bunion, clawing, or simple flexion deformities. Manage by releases and osteotomies to preserve function. Avoid fusions when possible.

External rotation deformity is nearly always secondary to external tibial torsion. Correct by a supramalleolar medial rotational osteotomy and fix with a short plate and screws.

Walking

Walking ability is related to level of paralysis, knee flexion deformity, mental status, family compliance, and gait training. It is not related to hip reduction status. Most sacral, many lumbar, and a few thoracic level patients walk [2]. Walking often deteriorates in late childhood and adolescence, when the body weight increases more than muscle mass. Provide walkers for gait training and crutches for long-term use. Walking must be energy efficient, convenient, and comfortable to be maintained. Combinations of independent and wheelchair mobility often provide a good long term solution for mobility.



1 Correction of ankle valgus by temporary physal screw placement. Correct ankle valgus (red line) by placing a medial malleolar screw to inhibit medial physal growth. Remove the screw when the valgus deformity is corrected (yellow line).



2 Crutch walking in myelodysplasia. This is a common mode of walking in children with myelodysplasia.

Problems

Pathologic fractures may follow minimal trauma, manipulative treatments, or operative procedures [1]. Pathologic fractures may be confused with osteomyelitis. Avoid casts and splint until comfortable. Avoid long-term immobilization. Physeal stress fractures may occur in the distal femur and upper tibia [2].

Cord tethering is suggested by loss of function, increasing deformity or pain [3]. Untethering often does not arrest deformity progression or eliminate the need for additional orthopedic procedures.

Skin breakdown includes sacral decubitus and foot ulcers [4]. These are common, serious problems that cause considerable disability. Reduce risk by correcting rigid foot deformities and severe pelvic obliquity.

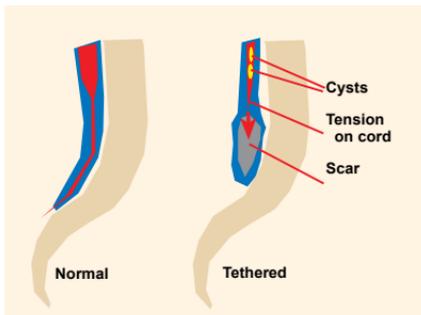
Latex allergy occurs in about 5% of patients. Screen patients for a history of latex allergy. Create a latex-free environment in both the hospital and at home. Before any operative procedure make certain the problem is accessed by an anesthesiologist and considered for preoperative prophylaxis.



1 Pathological fractures. Acute fractures (arrow) often follow a period of immobilization for treatment.



2 Physeal injuries. Physeal stress fractures result from repeated microtrauma. If the injury is asymmetrical, an angulatory deformity develops (arrow).



3 Tethered cord in myelodysplasia. These deformities are often complex with scar, adhesions, traction on the cord and cyst formation.



4 Skin breakdown in myelodysplasia. Skin problems are common due to poor sensation. Manage by applying a heavily padded cast that extends beyond the toes to protect the skin. The child may then be mobile.

Muscular Dystrophy

Muscular dystrophies are a group of uncommon, genetically determined primary myopathies characterized by progressive muscle wasting [1]. The natural history is variable [1 opposite page].

Duchenne Muscular Dystrophy

Duchenne muscular dystrophy (DMD) is most common and caused by absence or impaired dystrophin. The dystrophin gene is located on the X chromosome. Dystrophin stabilizes the cell membrane and protein complexes within the muscle cell. A loss of this function causes the disease. Prenatal diagnosis can sometimes be made by special molecular studies or fetal muscle biopsy.

Clinical features include males with an onset in early childhood, a wide base gait, impaired running, positive Gowers sign, delayed motor development, intellectual slowness, calf pseudohypertrophy [2 opposite page], and often a positive family history. About onethird are new mutants.

Diagnosis Serum CPK is elevated 200–300 times normal values. Muscle biopsy shows variation in fiber size, loss of fibers, increased fat, and sometimes centralization of nuclei. EMG shows myopathic changes.

Natural history Most boys show progressive deterioration with flexion–abduction contractures of the hips, and flexion contractures of the knees and ankle with loss of walking about 10 years of age. Once walking ability is lost, scoliosis develops progressively. Cardiomyopathy and pulmonary compromise progress, and death usually occurs in late teens.

Lower limb management Use stretching exercises and night AFO splinting to delay contracture formation. Attempt to preserve walking by releasing contractures just before the ability to walk is lost. Release the Achilles tendon and consider an anterior transfer of the posterior tibialis tendon. Release knee flexion and hip flexion–abduction contractures if they limit function. Place in a long-leg brace made before surgery and mobilize on the first postoperative day [1 on page 462]. This aggressive management may delay wheelchair dependence by 1–4 years.

Mobility Provide effective mobility. An electric wheelchair and a van for the family are useful. Make the home wheelchair accessible. Foot deformities should be corrected before the child becomes wheelchair dependent [2 on page 462].

Muscular Dystrophy

X-chromosomal

Duchenne muscular dystrophy
Becker muscular dystrophy
Emery–Dreifuss muscular dystrophy

Autosomal dominant

myotonic muscular dystrophy
fascioscapulohumeral dystrophy

Autosomal recessive

limb–girdle muscular dystrophy
congenital muscular dystrophy

1 Types of muscular dystrophy.

Spine management Prepare the family in advance of the onset of scoliosis. Progressive scoliosis develops once the child becomes non-ambulatory [1 on page 463]. Avoid bracing. Between about 11 and 13 years, when the curve approaches 20° , and before the vital capacity falls below 30%, perform a segmental instrumentation and fusion from T2 to L5 or the sacrum.

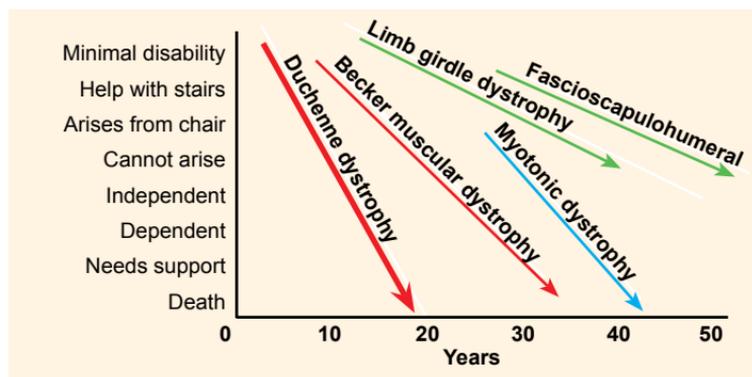
Family support This disease causes guilt in the mother, stress in the family, and depression in the child. Provide a support system and make available counseling as necessary.

Long-term ventilatory support Prolongation of survival is technically possible but creates many ethical issues.

Dystrophin replacement may be feasible in the future.

Becker Muscular Dystrophy

Becker muscular dystrophy (BMD) is a uncommon, mild, muscular dystrophy due to dystrophin abnormality with clinical findings similar to DMD but much less severe. Early features of BMD include leg cramps and gait problems during childhood. Scoliosis is rare. Survival into midadult life is usual. In some cases, the diagnosis is not established until adult life. Management problems are similar to DMD but required at an older age. Provide physical therapy, release of contractures, and effective mobility.



1 Natural history of muscular dystrophies. The arrows show the average clinical course with level of disability and age.



2 Clinical features of Duchenne muscular dystrophy. Note the calf pseudohypertrophy (red arrows) and positive Ober test (yellow arrow). This positive Ober test demonstrates a tensor fascia contracture.

Emery-Dreifuss Muscular Dystrophy

This is a rare sex-linked recessive muscular dystrophy with elbow, triceps, and postero-cervical muscle contractures, slowly progressive muscle wasting, and cardiomyopathy. Weakness becomes apparent during the first decade, and more pronounced during adolescence. Differentiate this from BMD and DMD by only mildly elevated CPK levels and by dystrophin testing. Patients may require contracture releases, spinal stabilization and referral to a cardiologist for insertion of a cardiac pacemaker.

Autosomal Dominant Muscular Dystrophies

Myotonic dystrophies include a heterogeneous group of diseases characterized by myotonia. Myotonia is the inability of the muscle to relax after contracting. Severe forms are seen.

- *Myotonia congenita* has an onset in infancy but becomes more evident during adolescence. Clinical features include generalized muscle hypertrophy, absence of skeletal deformities, and minimal long-term disability.
- *Congenital myotonic dystrophy* shows variable expressions with potential for severe early involvement. Features include hypotonia, delayed development, feeding and respiratory difficulties, and mild mental retardation. Walking is delayed. Cataracts may develop in adolescence. Orthopedic problems include clubfeet, hip dislocations, and lower limb contractures [2 opposite page].
- *Myotonic dystrophy* onset is during adult life. Clinical features include encephalopathy, facies myopathica, paresthesia, atrophy, myotonia, mental retardation, cataract, diabetes, and cardiac conduction defects.

Fascioscapulohumeral dystrophy (FSHD) is characterized by progressive weakness and atrophy of the facial, shoulder-girdle and upper arm muscles, and occasional later pelvic-girdle and lower limb involvement. Shoulder instability may be improved by scapulothoracic fusion [3 opposite page].



1 Long-leg braces may prolong walking.



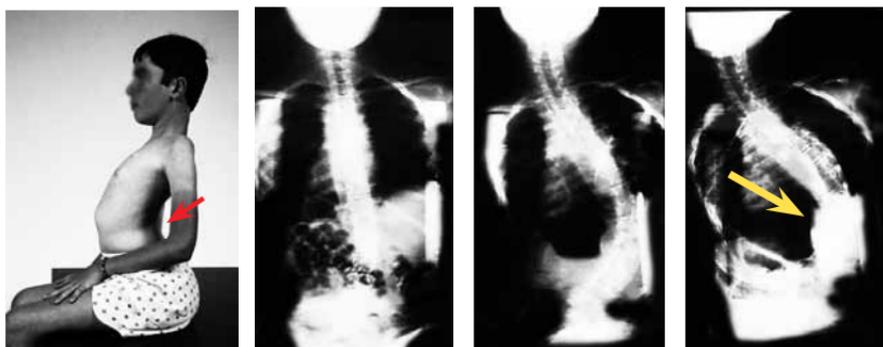
2 Heel-cord contractures in DMD makes foot positioning difficult. These equinovarus deformities (red arrows) make positioning of the feet on the footrests difficult.



Autosomal Recessive Muscular Dystrophies

Congenital muscular dystrophy (CMD) includes a heterogeneous group of congenital disorders characterized by marked hypotonia, generalized muscle weakness, and frequently multiple contractures. Four categories of CMD include: (1) the classic form without severe impairment of intellectual development; (2) the Fukuyama type CMD with muscle and structural brain abnormalities; (3) the milder Finnish type, and (4) severe Walker-Warburg syndrome (CMD IV).

Limb-girdle muscular dystrophy is similar to FSHD but without involvement of the facial muscles. Problems are similar to DMD and BMD except scoliosis is mild. Longevity is limited to middle age.



1 Progressive scoliosis in DMD. Child has thoracolumbar lordosis (red arrow). Scoliosis becomes progressive once the patient becomes nonambulatory. Instrument and fused before curves become severe (yellow arrow).



2 Unusual forms of muscular disorders. Congenital muscular dystrophy (red arrow) and heel-cord contracture in congenital myotonia (yellow arrow).



3 Scapulothoracic fusion. Shoulder stabilization improves upper extremity function in FSHD.

Poliomyelitis

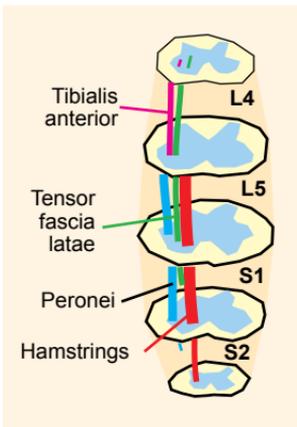
Acute poliomyelitis is a viral infection that damages the anterior horn cells and brain stem motor nuclei, causing paralysis. From a human host, the disease is spread by the oropharyngeal route. Most develop only a mild gastroenteritis. About 1% develop paralysis.

Stages of Poliomyelitis

Acute stage After an incubation period of 1–3 weeks, a systemic flu-like illness develops. Rarely the infections spreads to the nervous system causing inflammatory changes with varying degrees of neuronal degeneration. Over a period of 1–2 weeks a progressively increasing paralysis develops without sensory involvement. Muscles with motor nuclei extending over several segments are most likely to be affected [1]. About twice as many muscles become weak as become totally paralyzed.

Convalescent stage This recovery phase extends over a period of about 2 years. Most recovery occurs during the first few months. Contractures start to develop during this period.

Chronic stage After about two years the disease becomes chronic. Muscle imbalance, contractures, and growth cause increasing deformities. Most severe deformities are seen in severely affected younger patients with many years of growth. Limb atrophy and shortening are characteristic deformities [2].



1 Relative length of cell columns in spinal cord. Based on Sharrard (1955).



2 Lower-limb atrophy due to poliomyelitis. This atrophy is associated with limb shortening (red arrows).

Management Principles

Sensation and IQ are unaffected, improving prognosis as compared with children with cerebral palsy or myelodysplasia.

Prognosis is better for the young with minimal paralysis.

Contractures develop with time All soft tissue components— including muscle, tendon, joint capsules, fascia, and neurovascular structures—become contracted.

Gentle stretching and splinting An overcorrected position may improve or prevent progression of deformity [1]. Stretching must be performed carefully to avoid fractures of the fragile bone.

Operative correction is useful to correct deformity and provide stability. Sometimes function can be improved by tendon transfers.

Basic principles of tendon surgery are well established [2].

Simple osteotomy fixation techniques are frequently utilized as these procedures are most commonly performed in developing countries with limited medical resources.

Bony procedures are best delayed until the end of growth to prevent recurrent deformity.



1 Stretching casts for knee flexion deformity.

Principles of tendon transfers

- Transfer only muscles of fair (3) or good (4) grade
- Insure muscle excursion suitable
- Loss of function from original site acceptable
- Joint with adequate free passive motion
- Transfer cannot overcome fixed deformity
- Insure unrestricted motion of joints at new site
- Preserve muscle neurovascular status
- Transfer in straight line
- Attach into bone preferable
- Place transfer under proper tension

2 Principles of tendon transfers. Based on Mayer (1956).



3 Assessing hip abduction contracture in poliomyelitis. From Gautam 1998.

Bracing is often useful to provide stability and to facilitate walking.

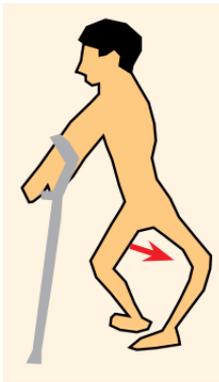
Postpoliomyelitis syndrome (PPS) causes slowly progressive weakness, atrophy, muscle pain, and fasciculations that occur 15 or more years following the original disease. Management is usually conservative.

Orthopedic Management

Assessment requires a careful evaluation, which includes grading strength of muscle groups, determining the range of active and passive motion, assessing contractures [3 previous page], determine limb length differences, and documentation of deformity [1]. Function may be improved by tendon transfers. This requires a preoperative assessment of specific muscles to determine which have adequate strength to function effectively in a rerouted position.

Upper limb The objective is to place the hand in a position for optimum function and for stability to facilitate transfers and crutch walking. Children with little hand function may still use crutches and have prehension between the arm and the chest. These important adaptive functions should be preserved.

- *Shoulder* stability is more important than mobility.
- *Elbow and hand* require mobility for optimal function. Establish motion and correct deformity before performing tendon transfers [2].



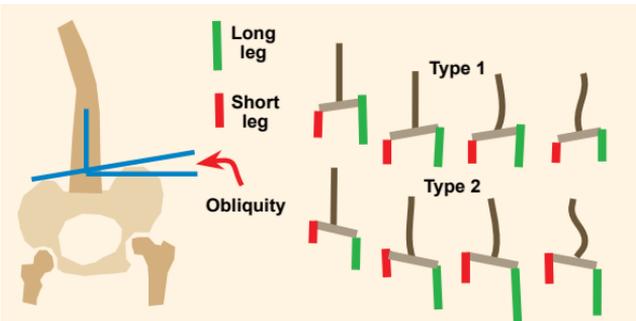
1 Hyperextension knee deformity in poliomyelitis.



2 Muscle transfers may improve hand function.



3 Scoliosis from poliomyelitis. Note the scoliosis and pelvic obliquity.



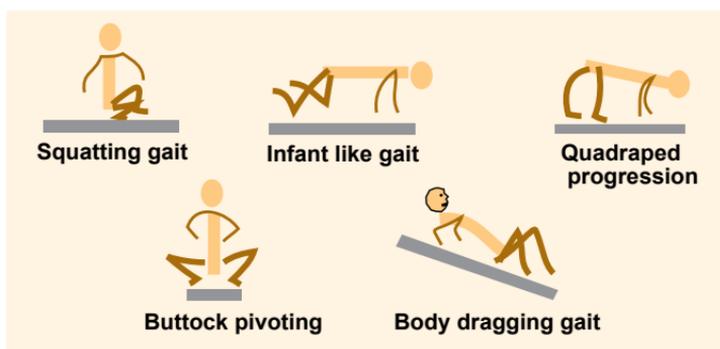
4 Pelvic obliquity in poliomyelitis. Note that a variety of curve patterns occur with pelvic obliquity and limb shortening. Based on Lee (1997).

Spine Scoliosis [3 opposite page] occurs in about a third of children with paralysis. Curve patterns are usually either a double-major or long paralytic types. Pelvic obliquity is common [4 opposite page]. Bracing may slow progression for 20° – 40° curves. For curves 40° – 60° a posterior segmental instrumentation and fusion is often indicated. Curves $>60^{\circ}$ may require anterior and posterior instrumentation and fusion.

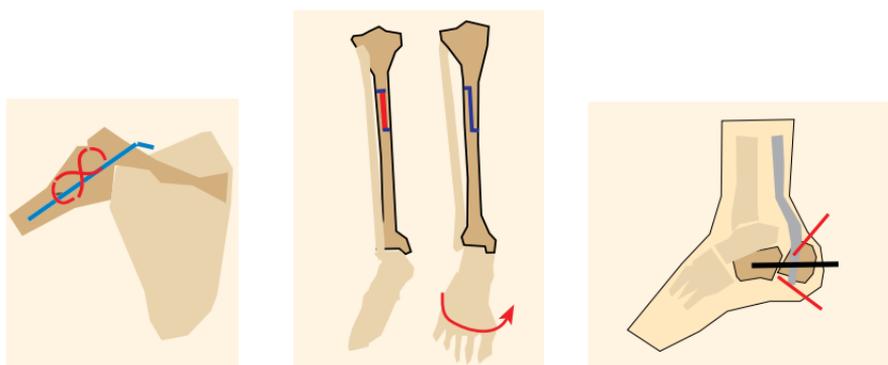
Crawling is a form of mobility commonly used by children without medical care. With good medical care, about 60% of these children can become community and 30% household ambulators [1].

Lower limb requires most attention as paralysis is more common and corrective procedures most effective. The objective is to provide stability and symmetry for walking with or without a brace. The foot should be plantigrade, the knee extended, and the hip stable. Symmetry requires an absence of significant pelvic obliquity and leg length inequality.

Common procedures in poliomyelitis include shoulder fusions [2]; tensor fascia releases; correction of knee flexion contractures, rotational osteotomies [3], correction of calcaneus [4], and cavus deformities; and limb length equalization procedures.



1 Patterns of crawling in poliomyelitis. Based on Arora (1999).



2 Shoulder fusion. Using fig. 8 wiring and K wire fixation. From Mohammed (1998).

3 Tibial rotational osteotomy. Removal of an anterior bone wedge (red) produces rotation. From Asirvatham (1990).

4 Calcaneal osteotomy and tendon sling for calcaneus deformity. Osteotomy is fixed with a K wire. From Pandy, (1989).

Arthrogyriposis

Arthrogyriposis multiplex congenita includes a heterogenous group of disorders characterized by multiple congenital joint contractures. The disorder occurs in about 1/3000 births. Decreased fetal movement due to fetal or maternal abnormalities cause the deformities. The causes of fetal akinesia include disorders of nerves, muscles, or connective tissues, maternal disease, intrauterine constraint, or vascular compromise. The earlier and longer this loss of movement, the more severe the deformities. The most common deformities include clubfeet and hip dislocations. Most deformities are nonprogressive [1].

Evaluation

More than 100 disorders are included in the differential diagnosis of multiple congenital contractures [2]. This differentiation may be complex and in some instances the exact diagnosis cannot be established. Deformities may be classified based on whether they primarily affect the limbs, include systemic involvement, or a clear neurological etiology. Certain types are most likely to be seen by the orthopedist.

Amyoplasia is the classic form of arthrogyriposis making up about a third of the cases. The common features include clubfeet, flexed or extended knees, dislocated hips, internally rotated and abducted shoulders, flexed or extended elbows, pronated forearms, and flexed wrists and fingers. The trunk is less commonly affected. Muscles are hypoplastic or absent, joints are fibrotic and stiff. Joints show a loss of creases and dimpling. IQ is normal, sensation is intact, potential for walking is good, and most become independent and productive in adult life.



1 Arthrogyriposis in different age groups. The natural history is favorable. Multiple contractures at birth (left), treatments early childhood (middle), and a successful outcome in most patients (right).

Classification of arthrogyriposis

- I Mainly limbs**
 - Amyoplasia
 - Distal arthrogyriposis type I
- II Limbs and other body areas**
 - Freeman–Sheldon syndrome
 - Diastrophic dysplasia
- III Limbs and central nervous system**
 - Fetal alcohol syndrome
 - Trisomy 21

2 Classification of arthrogyriposis – major categories and examples. From Hall (1997).

Distal arthrogryposis includes 6 subtypes [1], often inherited and primarily involves the hands and feet. Fingers are flexed, medially deviated, and overlapping, and fist is clenched. Clubfeet or vertical tali are common.

Contractual arachnodactyly or Beal syndrome is an autosomal- dominant disorder with long extremities, joint contractures, and ear crumpling.

Pterygium syndromes include a group of varied disorders with characteristic features [2 and 3].

Freeman–Sheldon syndrome or whistling face syndrome is a familial disorder with a characteristic *puckered appearance* to the face and multiple joint contractures.

Diastrophic dysplasia is a syndrome that includes short stature, multiple contractures, clubfeet, proximally placed thumbs, and progressive kyphoscoliosis.

Management

Management principles may be applied to most of the patients.

Accurate diagnosis is necessary to advise the family about the risk of recurrence.

Family counseling Deal with family guilt, which is usually present and unless resolved will interfere with management. Provide information regarding local or national arthrogryposis support groups. For parents of infants with amyoplasia, provide information regarding the favorable natural history with progressive reductions in deformity and the potential for an independent and productive life.

Classification of distal arthrogryposis

I	Overlapping, ulnar deviation fingers
IIa	Short stature, cleft palate
IIb	Hard muscles, ptosis
IIc	Cleft lip and palate
IId	Scoliosis
IIe	Trismus

1 Classification of distal arthrogryposis. Based on Hall (1997).



2 Popliteal pterygium. Note the severe knee flexion deformities (arrows).

Multiple pterygium syndromes

- Popliteal pterygium
- Antecubital pterygium
- Multiple pterygium – autosomal recessive
- Multiple pterygium – autosomal dominant
- Lethal multiple pterygium
- Lethal popliteal pterygium
- Pterygium and malignant hyperthermia

3 Classification of multiple pterygium syndrome. Based on Hall (1997).

Physical therapy should be started early to reduce contractures and facilitate bonding. Encourage the family to provide the treatment. This is most convenient and economical for the family, and the parent-child interaction is emotionally therapeutic. Stretching should be gentle and atraumatic.

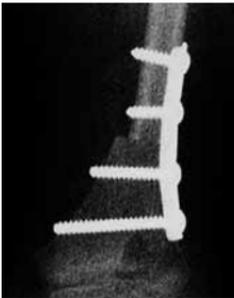
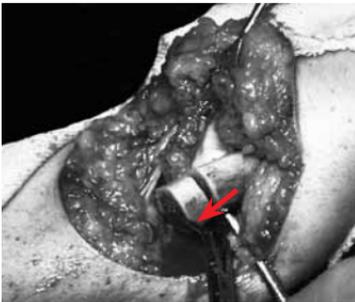
Bracing facilitates function by providing joint stabilization for standing and walking. Lightweight nonarticulated plastic AFO or KFO are most useful. Allow bracefree periods during the day for the child to crawl. Splinting at night is often essential to prevent recurrence of deformity once corrected by surgery.

Adaptive equipment such as walkers, electric wheelchairs, and devices to facilitate self-care are very valuable.

Operative correction is usually necessary to correct clubfeet, knee contractures, or hip dislocations. Minimizing the duration of immobilization during infancy and childhood by combining procedures, minimal duration of postoperative immobilization, and avoiding repeated procedures. Delay operative correction of upper-extremity deformities until early childhood when disability is clear. Surgery is necessary only to enhance function not correct deformity per se.

Clubfeet

Manage by early stretching, casting, percutaneous releases, repeated casting. Correct residual deformity by extensive posteromedial release or talectomy. Provide night splinting for several years following correction to prevent recurrence. If recurrence does occur, correct with serial casting and reinstitute night splinting. Provide stretching exercises during the day. Avoid repeated procedures. Try to delay reoperation for rigid recurrent deformity until the end of growth.



1 Correction of knee flexion contracture. Correct severe contracture with femoral shortening (red arrow) and plate fixation.

Knee Flexion-Extension Deformity

Manage by early stretching to correct less fixed deformity. Correct fixed deformity by hamstring lengthening, capsulectomy, and femoral shortening if necessary [1 opposite page]. Correct hyperextension deformity by quadriceps lengthening to center arc of motion to about 15° of flexion.

Hip Dislocation

Reduce dislocations in infancy [1] by a medial approach open reduction and shorten postoperative immobilization to about 5 weeks. See page 407. Combine with other procedures when possible. Avoid repeated or extensive procedures that may cause stiffness. Establish symmetry and maintain mobility.

Upper limbs

Provide early manipulation to improve motion and retain with night splints. Teach use of adaptive devices to facilitate self-care. In early childhood, perform procedures to correct deformity that interferes with function. Because of good hand sensibility, function is often surprisingly good [2].



1 Hip dislocations (arrow) should be reduced in early infancy.



2 Upper extremity function is surprisingly good. This is due to normal sensation and excellent adaptive mechanisms.

Miscellaneous Disorders

Hereditary Sensory Motor Neuropathies

This is a group of conditions that involve the sensory nerves, are often familial, and sometimes so mild in the parents that they may not be aware that their problem is a genetic disorder. The disability starts in childhood with gait disturbances; later, scoliosis and hand dysfunction develop. Nerve condition is delayed. These disorders are progressive and recurrence often follows operative correction. This disease is commonly classified into six groups [1]. The first three are of most orthopedic significance.

Charcot-Marie-Tooth Disease (CMT) develops during childhood, is autosomal dominant (30% new mutations), often seen first for pes cavus. Check for loss of vibratory sense and foot eversion weakness. Examine parents as sometimes they show signs of the disease without being aware of the diagnosis.

- Foot* Bilateral pes cavovarus is the most common foot deformity [2]. The specific components include hindfoot varus, anterior or forefoot cavus, and, often, claw toes. The deformity is caused by the tibialis posterior overpowering peroneus brevis coupled with peroneus longus overpowering of the tibialis anterior. Manage pes cavus as outlined on page 111.

Hereditary Sensory Motor Neuropathies (HSMNs I–VI)

Type I	Charcot–Marie–Tooth (CMT) disease
Type II	Axonal neuropathy
Type III	Hypertrophic interstitial neuropathy (Dejerine–Sottas disease)
Type IV	Refsum disease
Type V	Hereditary sensory neuropathy with spastic paraplegia
Type VI	Hereditary sensory neuropathy with optic atrophy

1 Classification of hereditary sensory motor neuropathies.



2 Foot deformity in CMT disease.

- *Spine* Scoliosis, kyphoscoliosis, or kyphosis develops in about half by the end of growth.
- *Hand* disability is increasing likely with advancing age. Disability results from decreased conduction velocity of the nerves, lack of opposition, weak pinch, and clawing of the fingers. Tendon transfers may be helpful in improving function.

Axon Neuropathy Like CMT disease but milder.

Hypertrophic Interstitial Neuropathy This condition may be associated with clubfeet. Weakness and sensory loss are progressive. Thickening of the peripheral nerves may be felt.

Spinal Muscular Atrophy

Spinal muscular atrophy (SMA) includes a heterogenous group of disorders that are rare, autosomal recessive, with anterior horn cell degeneration, causing muscle weakness and atrophy. Sensation and mentation are unaffected. SMA was traditionally classified by the age of onset but currently is being classified based on severity [1].

SMA type I Infants are severely affected at birth, profound hypotonia, fragile, may have pathologic fractures. Maintain range of motion. Aggressive orthopedic care not appropriate.

Spinal Muscular Atrophy Classification

Traditional classification

Type I	Werdnig–Hoffman disease acute	onset <6 mo.
Type II	Werdnig–Hoffman disease chronic	onset 6–24 mo.
Type III	Kugelberg–Welander disease	onset >24 mo.

Current classification

Type I	Severe – unable to sit, usually dies in infancy
Type II	Moderate – diagnosis in infancy, unable to walk, dies mid–adult life
Type III	Mild – diagnosis after age 2, marginal walkers, independent adults

1 Classification of spinal muscular atrophy. Two different classifications are in use. The traditional classification is based on age of onset. The current and more commonly used classification is based on severity with prognostic implications.

SMA type II Children with this intermediate form live many decades with considerable disability. Prolongation of survival is possible with ventilatory assistance despite severe respiratory and bulbar muscle dysfunction. Common orthopedic problems include hip dislocations and spinal deformity.

- *Hip subluxation or dislocation* may occur due to muscle imbalance, hypotonia, pelvic obliquity, and poor muscle tone. Attempt to maintain free mobile symmetrical motion. Avoid operative procedures as disability is minimal and recurrence is common.
- *Scoliosis* occurs in nearly all of these patients. It occurs during the first decade and progresses at a rate related to the severity of the weakness. Bracing or modifications of seating may be helpful in supporting an upright posture. It is doubtful that bracing significantly affects progression. Posterior segmental fusion is indicated for curves approaching 40° and when forced vital capacity is still above 40%.

SMA type III Attempt to maintain walking as long as possible. Rarely hip dysplasia progresses making correction by femoral and pelvic osteotomy necessary. Progression of scoliosis is less severe than for type II but may require fusion. Be aware that long fusions may adversely affect walking.

Friedreich Ataxia

Friedreich ataxia (FRDA) is an autosomal recessive, progressive, spinocerebellar degeneration with a trinucleotide repeat defect found in chromosome 9.



1 Spinal muscular atrophy. Disability in severe form is profound with severe muscle weakness and disability. Note the lack of head control.



2 Progressive scoliosis in Friedreich ataxia. Curves at age 9 and 14 years.

Clinical features Friedreich ataxia is the most common cause of recessive ataxias. Neurological examination shows ataxia, dysarthria, areflexia, pyramidal signs, and sensory deficits. Cardiomyopathy and diabetes mellitus are common. Scoliosis and pes cavus are primary orthopedic problems. Walking becomes progressively more difficult and ceases to be functional by about 20 years of age. The disease progresses causing cardiopulmonary failure and usually death in mid-adult life.

Management Individualize management. Scoliosis is progressive [2 opposite page]. Brace curves in the 25°–35° range if the bracing does not interfere with walking. Larger curves may require segmental instrumentation and fusion. Cavus foot deformities may be improved by plantar-medial release followed by tendon transfers and osteotomies. Provide effective mobility with electric wheelchairs.

Congenital Myopathies

These heterogenous disorders cause hypotonicity and muscle weakness in infants and children [2]. These disorders are non or slowly progressive. The diagnosis is established by histochemical analysis and electron microscopy of muscle biopsy specimens. Currently the classic list [1]) has been expanded to more than 40 disorders.

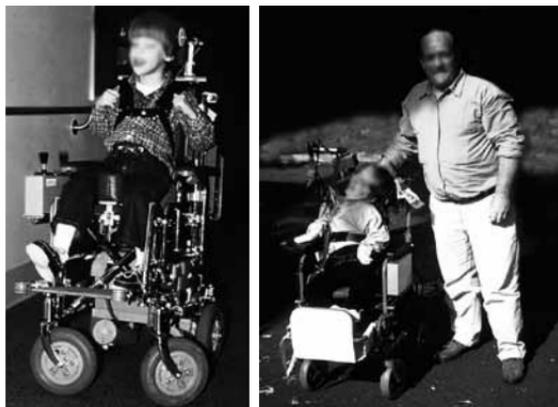
Dermatomyositis

Juvenile dermatomyositis is a multisystem disease causing inflammation affecting primarily the skin and muscle producing symmetrical weakness, and typical skin rashes that affect the face and hands. Confirm the diagnosis by an elevated CPK and muscle biopsy. Treatment often requires steroids or methotrexate.

Congenital Myopathies

Central core disease
Nemaline myopathy
Myotubular myopathy
Congenital fiber-type disproportion
Metabolic myopathies

1 Classic classification of congenital myopathies.



2 Mobility aids in children with profound muscle weakness. Electric wheelchairs provide effective mobility and increased independence.



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