

33 Paediatric surgery

33.1 Surgery in children, infants and neonates

There is an increasing proportion of children in LMICs. The number of specialized paediatric surgeons available in Europe is c.1:50,000 population; in South Africa 1:2,669,000, and elsewhere in Africa much less. These figures reflect the fact that in Africa well over 50% of the population are children.

More relevantly, there are c.500 live births per year per European paediatric surgeon; in South Africa over 35,000. So the specialist in Europe will wait 6yrs to see a case of oesophageal atresia (c.1:3,000 live births), whereas the specialist in South Africa sees one about every month! Taking this argument further, it is obvious that every child needing surgery cannot hope, in the poorer parts of the world, to be treated by a specialist. This is why we describe the simpler procedures here for the non-specialist.

CHILDREN ARE NOT SMALL ADULTS

You may be accustomed to operating on adults but find the child patient, especially the neonate, an unfamiliar and intimidating prospect. You may find also you have to pay special attention to the concerns of the parents and family.

“TO TREAT AN ADULT AS A CHILD IS NO MISDEMEANOUR; BUT TO TREAT A CHILD AS AN ADULT MOST OFTEN ENDS IN DISASTER”

Children differ from adults in every anatomical, physiological, pathological or psychological sense. They cannot handle fluids, blood sugar, electrolyte changes and heat like adults. Their fluid requirements are different and their anaesthetic requirements likewise. Neonates tolerate fluid and electrolyte loss particularly badly: they don't have the ability to retain sodium and water post-operatively or concentrate urine like adults, and they also easily become cold, dehydrated and hypoglycaemic. They bleed easily and have little physiological reserve, so they can deteriorate quickly, and don't tolerate delays in treatment (*e.g.* referral to a distant hospital).

Specific paediatric surgical problems are described in this chapter; other important aspects of paediatric surgery are described elsewhere (consult the list at the end of this chapter).

Trauma, which covers fractures, burns, snake bites, and other violence including sexual abuse, is covered in Volume 2.

Perioperative management

(1) POSITIONING AND WARMING

Whenever possible, keep the child next to his mother or nearest relative. If he is old enough, try to explain what is happening and don't lie by saying things won't hurt when they will! Wherever possible, let a neonate breastfeed; otherwise use glucose on a stick, finger or dummy to pacify him.

If you need to perform an invasive procedure, wrap the child in a warm blanket to immobilize him safely and to prevent him from fighting and kicking you! Prepare what you need beforehand, and get the mother and a nurse to assist you.

Make sure the theatre is warm. Place a child on a well-padded cross made of two splints, and bandage the arms and legs to it. Cover the rest of the body except for the part to be operated with cotton wool or commercially produced silver foil. Use warmed solutions for preparation, infusion and washouts.

(2) INTRAVENOUS LINES. An infant requiring GA should have a good IV access, namely, in the antecubital fossa, scalp, or neck. *Avoid using a tourniquet to bring up a vein:* the pressure of a finger in a small child suffices. You rarely need to make a cut-down, but may need a central venous line preferably using the subclavian route. *Never attempt this twice on the same side without checking that no iatrogenic pneumothorax has developed.* If you are unable to find a suitable vein to cannulate, try in the OT under good light and with the child sedated. Use ultrasound to help you, if possible.

(3) UMBILICAL VEIN CANNULATION is a good and reliable option; *avoid cannulating the femoral vein in children wearing nappies* because of the high risk of infection. If available, use ultrasound to detect veins suitable for cannulation and to facilitate insertion.

(4) INTRA-OSSEOUS CANNULATION into the tibia is a fast and reliable route in children of all ages. *Take care to avoid the epiphyseal plate.* Remember many substances are toxic to bone marrow as well as venous endothelium and can easily cause thrombosis or even extravasation in neonates. Therefore fix the cannula properly and re-check its position and functioning repeatedly.

(5) FLUIDS. Replace *all the initial fluid deficit* with Ringer's lactate or 0.9% saline during the 3-6h of pre-operative preparation time. Prescribe the post-operative fluids yourself. *Don't leave this to the nurses, and don't exceed 5ml/kg/h* unless the fluid deficit is uncorrected. Where possible, provide the fluid needed as half-strength Darrow's solution with 5% dextrose, or Ringer's lactate with 5% dextrose, or half-strength (0.45%) saline with 5% dextrose.

Fluid balance in a neonate is even more critical: take great care he gets enough *but not too much*. A term baby usually needs c.100ml/kg/day (less in the first 48-72h), and a premature baby 120-150ml/kg/day. For a child between 10-20kg, use 80ml/kg; and >20kg, 60ml/kg.

It is best to administer IV fluids as boluses rather than relying on an unreliable or unmonitored infusion.

For major surgery, make sure you monitor post-operative urine output. A child should pass 1-2ml/kg/h, and a neonate 2-4ml/kg/h. *You don't need an indwelling catheter unless you are absolutely sure the urine output will be observed and measured.* A urethral catheter is often not appropriate; so, for a boy, use a condom catheter (Paul's tubing: 27-3) of suitable size; alternatively, use a fine feeding tube. If you do need a urethral catheter, pass it yourself and take the precautions described (27.2), making sure a girl's hips are fully flexed and externally rotated for a good view. Make sure you have a good light, and can see the urethral orifice. In a boy, you might find the foreskin quite sticky with smegma: carefully clean it with sterile water while pulling back the foreskin gently. In girls, spread the labia to expose the vulva: the urethral orifice may be very difficult to see. It might help to push gently in the suprapubic area to cause some urine to come out: watch carefully from where it emerges! If you have accidentally put a catheter in the vagina, leave it there temporarily before trying again *with a new catheter* (to show you which is the wrong passage). Always be very careful, because you can tear the urethra if you use force!

Replace blood with blood ml for ml if you lose >10ml (or less in premature neonates); a child has a blood volume of approximately 75ml/kg, a neonate 85ml/kg and a premature baby up to 100ml/kg. Monitor and replace blood loss with the greatest care (3-1). Weigh all blood-soaked swabs accurately, on a scale borrowed from the pharmacy, if necessary. Try to avoid blood loss by all means! Make sure all neonates get 1mg vitamin K IM or IV pre-operatively.

(6) POTASSIUM. Normal daily maintenance is 1-3mmol/kg. No extra potassium is needed in the 1st 24h of life. If a child is not taking oral fluids by 24h, he needs a potassium supplement. Add 10mmol to 500ml of IV fluid (=20mM K⁺). *Don't infuse >10mmol/h or 3mmol/kg/day.* Or, use 5% dextrose in half-strength saline, which contains 18mM K⁺. *Potassium replacement can be very dangerous in children* if it is handled incorrectly. *Avoid adding potassium if renal function is disturbed.* If a child becomes drowsy post-operatively (and the glucose is correct), and the bowel becomes silent, suspect ileus, and add more potassium.

(7) SODIUM. Normal maintenance is 2-4mmol/kg/day. No extra sodium is needed in the 1st 24h of life. Replace nasogastric and intestinal losses carefully. A preterm infant may require up to 6mmol/kg/day in the 1st month of life.

(8) CALCIUM. Normal maintenance in the neonate is 2-25-4-5mmol/kg/day, which means 10-20ml/kg of 10% calcium gluconate per day.

(9) NASOGASTRIC TUBE (4.9)

Insert a well-lubricated tube of appropriate size, which has been stiffened by placing it for a short time in the freezer, through the nose with the neck fully flexed. In neonates, you can pass the tube through the mouth.

(10) NUTRITION

Interrupt feeding as little as you can. *Don't starve a child for >4h before an operation*, and restart feeding as soon afterwards as you can. Ask whether he has passed flatus. Listen every 4h for the return of bowel sounds, and note whether he has passed faeces or flatus; these signs show that feeding can start. Bowel sounds alone are not so reliable in children, so you can assess gastric emptying more accurately by aspirating the stomach hourly, before each intake of feed. Adjust the amount of feed tolerated according to the amount aspirated. Proceed with a staged feeding regime: start with ¼ of a normal (pre-operative) feed portion, diluted 1:2 with water; double this volume after 2h and then again after a further 2h, and then give the full undiluted feed after a further 2h. If the child brings up the feed, go back one stage, and try again. Most children are back on feeds 48h post surgery. If a child was starved <4h, and feeds are restarted soon, he is unlikely to be short of energy.

Where nutrition is going to be delayed for some time, you can provide 50% glucose through a central venous line, using it to replace the energy deficit resulting from starvation. Reckon that, if he cannot feed orally for more than 3 days, he needs 1-2g/kg/day. Test the urine and watch for glycosuria and an osmotic diuresis. Alternatively, dilute 25ml of 50% dextrose in 500ml of half-strength Darrow's solution and increase the concentration gradually.

In working out the energy content of various fluids, remember that 1L 10% dextrose contains 1700kJ (=400kcal). A child's daily post-operative energy needs are:

Neonate	3-10kg	10-25kg	25-35kg	35-60kg
420-840kJ/kg	250-330kJ/kg	190-270kJ/kg	145-190kJ/kg	125-145kJ/kg
(100-200kcal/kg)	(60-80kcal/kg)	(45-65kcal/kg)	(35-45kcal/kg)	(30-35kcal/kg)

Note that stress, cold, infection, trauma and surgery increase ordinary nutrition requirements; these should be 2-3g/kg protein and 10-15g/kg carbohydrate per day for the neonate.

CAUTION! If a child becomes drowsy, or unconscious, or behaves strangely, suspect hypoglycaemia, or, less commonly, water intoxication, or lack of electrolytes.

If you are operating proximal to the upper jejunum, a jejunostomy at the time of the operation is a good way to re-establish feeding (11.7).

(11) ANAESTHESIA. If you want to use LA on a neonate:
 (i) Dilute 1ml of 2% lidocaine to 10ml, and use half of this as the maximum dose.

(ii) Infiltrate the line of the incision with 0.25% bupivacaine, or 0.2% lidocaine. The maximum dose is about 5ml. *Avoid adrenaline in the neonate.*

Beware of using diazepam as pre-medication: its effects are unpredictable and may be paradoxical.

Ketamine is useful for short minor procedures for children >2yrs.

If a neonate requires an urgent operation, operate at 24h after birth, or as soon afterwards as possible. Lung function is poor if you operate before 24h, when the lungs are not yet fully expanded.

Aspirate the stomach, especially if there is pyloric obstruction (33.4) or bowel obstruction (33.2,3).

GA in neonates and small babies requires a skilful anaesthetist.

33.2 Neonatal alimentary tract obstruction

Most babies regurgitate or vomit a few times during the first few days of life, but bile-stained vomiting soon after birth almost always indicates bowel obstruction. When a newborn baby vomits repeatedly, he may have a medical condition such as:

- (1) Infection, typically arising from the umbilicus,
- (2) Meningitis,
- (3) Intracranial haemorrhage.

You must be able to distinguish these from true intestinal obstruction as the medical conditions are often readily treatable, if you diagnose them early.

A child's alimentary tract can obstruct at any level: oesophagus, stomach, duodenum, small bowel or, rarely, colon; but it most often obstructs at the rectum or anus.

Anorectal malformations form a separate group, and present as the failure to pass meconium, combined with abdominal distension, rather than vomiting (33.6).

Congenital atresia or stenosis of a child's small bowel presents as bilious vomiting shortly after birth, and often (but not always) the failure to pass meconium, which may be incomplete and irregular.

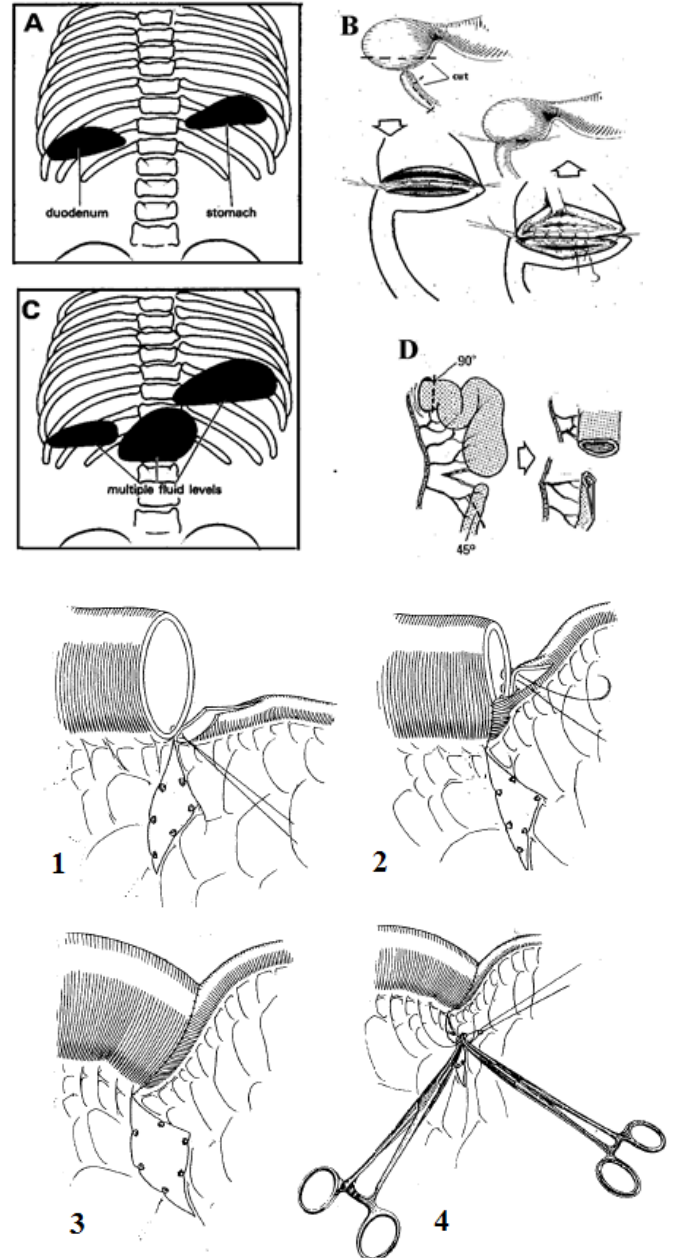
A minimum length of small bowel to survive is 25cm with an intact ileocaecal valve and colon, and 40cm without. However, without sophisticated support, some 100cm is probably the survivable limit.

If the obstruction is proximal to the middle of the small bowel, the abdomen does not distend significantly, but if the obstruction is below this point, it does. The distension may be localized or generalized. The child often presents only after 3-5 days, with severe dehydration.

An obstructed bowel is an emergency. Electrolyte and calorie loss affect a baby more dramatically than an adult, so he needs urgent treatment, within a few hours.

If you are a careful, dextrous operator, you may be able to save a few of these children despite not having sophisticated back-up: you will not have time to transfer these patients elsewhere.

2 CAUSES OF NEONATAL OBSTRUCTION



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Fig. 33-1 TWO CAUSES OF NEONATAL BOWEL OBSTRUCTION. A, radiograph showing the 'double bubble' of duodenal atresia. B, duodenal atresia relieved by a duodeno-duodenostomy. C, radiograph showing the multiple fluid levels of jejuno-ileal atresia. D, jejuno-ileal atresia showing how the proximal dilated loop is decompressed and resected at 90°, and the narrower distal segment divided at 45° with an added 'cutback'. E1-4, a single layer end-to-back anastomosis.

Partly after Mustard WJ, Ravitch MM (eds). *Paediatric Surgery 3rd ed 1978 Yearbook Medical*, with kind permission. E, after Lister J, Irving IM. *Neonatal Surgery 3rd ed, Butterworths, 1990 p.462 Fig. 32.8.*

Hypertrophic pyloric stenosis (33.4) presents later, as a previously healthy baby 3-6wks old, commonly male, who starts to vomit milk feeds, and *does not have diarrhoea*. This form of obstruction is readily treatable.

(a) Examination

Assess the distension of the abdomen, look for visible peristalsis. Feel for the distinctive mass of a hypertrophic pylorus (33.4). Examine for visible and palpable coils of terminal ileum that feel as if they might be filled with thickened meconium.

CAUTION! Neither the passage of meconium during the first 3 days, nor the absence of distension, excludes obstruction.

(b) Radiographs

After 12h the baby will have swallowed enough air to show air-fluid levels. If possible, take erect straight films before you start aspirating the stomach. The sign of intramural intestinal gas is diagnostic of necrotizing enterocolitis. Look for free air under the diaphragms to indicate a perforation.

Don't use contrast media introduced from above. In oesophageal atresia you may fill the lungs with it! It is also dangerous in obstruction lower than the jejunum, and may make it worse!

CAUTION!

(1) The neonatal jejunum, ileum, and colon all have the same smooth outline, and normally contain a few fluid levels. *They can often not be differentiated from each other radiologically.*

(2) *Don't use barium contrast media.*

(c) Differential diagnosis of neonatal bowel obstruction

Suggesting septicaemia: a site of origin for the infection, such as an infected umbilicus; child more ill than you would expect from obstruction alone; a +ve blood culture.

Suggesting meningitis: a stiff neck, headache, fever, irritability, fits and a +ve lumbar puncture.

Suggesting raised intracranial pressure: signs of cerebral irritation, a swollen fontanelle, photophobia; enlargement of the head and papilloedema are late signs.

Suggesting subarachnoid haemorrhage: impaired conscious level; also look for signs of hydrocephalus.

Suggesting some other cause of abdominal distension: a part of the abdomen which is dull to percussion. Causes include distension of the bladder in urethral obstruction, tumours, ascites, congenital cystic kidneys, and hydronephrosis.

Suggesting necrotizing enterocolitis (see below): a toxic lethargic preterm (<2kg) or HIV+ve baby, refusing feeds, with abdominal distension, vomiting and rectal bleeding (>25%), with radiological signs of intramural intestinal gas, free intra-peritoneal gas or gas bubbles in the portal vein.

(d) General management

As soon as you suspect the diagnosis, pass a nasogastric tube, strap it to the face, see that it is aspirated at least every 30mins, and let it decompress into a bag: *aspiration is a common cause of death in neonates.*

Start IV fluids to rehydrate the baby and correct electrolyte losses. Keep him warm. Perform gentle washouts with a rectal tube and prepare for surgery.

(e) Oesophageal atresia presents with regurgitation. It is usually associated with a tracheo-oesophageal fistula. The proximal oesophageal pouch fills with saliva, so there is excessive dribbling. Milk or water is likely to overflow into the trachea. The baby then froths, coughs, and becomes cyanotic with aspiration pneumonia. In order not to miss a case, you should pass a feeding tube on all neonates who regurgitate, especially underweight babies. Once the baby has an aspiration pneumonia, he is unlikely to survive surgery.

Confirm the diagnosis by passing, as far as it will go, a small (Ch8 in a full term and Ch5 preterm baby) nasogastric tube with a radio-opaque line in it, and then taking an AP chest radiograph. Make the tube firm by putting it beforehand in a freezer: it is then less likely to curl up; but *be very gentle so as not to perforate the oesophagus!*

The radiograph will show the oesophagus ending in a blind pouch; a lateral view is rarely necessary. *You don't need contrast medium.* Leave the tube reaching the blind end of the proximal oesophagus in place and aspirate frequently through it to prevent aspiration of saliva. Major thoracic intervention by an expert is necessary: generally, *if the weight is <2kg, or there is respiratory distress, it is not beneficial to intervene.* Rarely, there is no fistula, or a very narrow one, and the lungs remain clear: in this case, you might be able to bide time by forming a gastrostomy (13.9).

(f) Duodenal atresia or stenosis presents as vomiting on the 1st day of life. The vomit is usually bile-stained, because the obstruction is usually below the ampulla of Vater. The upper abdomen is distended. If the obstruction is above the ampulla of Vater, there will be no bile in the vomit. Erect AP radiographs and ultrasound show a characteristic 'double bubble', with no air (or very little) in the bowel beyond.

The bubble on the right is in the distended duodenal cap, and that on the left is in the stomach. There may also be other abnormal findings including Down's syndrome. A duodeno-duodenostomy or a duodeno-jejunosomy (33.3) will be necessary.

(g) Jejunio-ileal atresia or stenosis may occur at any point in the small bowel. Typically, it presents as bilious vomiting within 24h of birth, slightly later than with duodenal atresia, perhaps 1h after the first breastfeed; but it may be delayed for 2-3 days.

If there is jejunal stenosis (narrowing) rather than atresia (blockage), vomiting may be delayed for as long as 2wks. Obstruction in the upper jejunum is more common.

If the obstruction is low, it presents more slowly, with distension more evident than vomiting. About 50% of these children pass some meconium!

Hydramnios is common in the mother prenatally. Erect AP radiographs show considerable gaseous distension, ending at the site of obstruction, with *several* fluid levels. Unfortunately, by the time that several fluid levels are present, obstruction is advanced. A bowel resection (33.3) is necessary.

(h) Small bowel volvulus may present in older children (12.8) with sudden abdominal pain, distension and shock, or it may present in the 1st week of life, as an acute abdomen with bile-stained vomiting and abdominal distension. Volvulus usually involves the distal small bowel and proximal colon, and is due to a congenital malrotation of the intestine. If Ladd's bands (see below) are responsible, surgery is usually simple, because there is no need to resect bowel, if you managed to detect the obstruction early. *However, strangulation in volvulus develops rapidly*, and then resection is mandatory. This often means an extensive bowel removal, leading to short bowel syndrome.

(i) Necrotizing enterocolitis occurs in preterm (<2kg) or HIV+ve babies within the 2nd and 4th wks of life, and sometimes with duplication of the bowel (33.3). It is the most frequent cause of an acute abdomen in preterm babies, and is related to hypovolaemia, hypothermia, cardiac malformations, and umbilical catheterization. The fragile bowel wall becomes necrotic through a combination of effects of hypoperfusion and bacterial translocation, resulting in the classical presence of gas in the bowel wall. As the disease progresses, the bowel perforates and septicaemia ensues, characterized by erythema of the flanks, a generally tender abdomen. Radiological and ultrasound signs of gas in the bowel wall or in the portal vein, or free in the peritoneal cavity are typical of such deterioration. Look at bowel wall thickness and perfusion if you have Doppler ultrasound.

Start IV gentamicin, ampicillin and metronidazole, and monitor the baby carefully. Decompress free gas in the abdomen by a right hypochondrial puncture. If the condition improves, try to start breastmilk feeds early.

You should try to continue conservative treatment unless:

- (1) you see radiological signs of intramural intestinal gas or a persistent single dilated small bowel loop,
- (2) you feel a tender abdominal mass,
- (3) there is erythema of the abdominal wall,
- (4) you aspirate brown infected fluid from the peritoneal cavity,
- (5) you see sudden deterioration and progressive acidosis.

At laparotomy, you need to resect the affected bowel and fashion one or even multiple enterostomies (11.5). The mortality is high (up to 50%), and in severe cases, you might get better results by simple lavage and drainage of the abdomen. *Unless you can manage mechanical ventilation and careful intensive care of these babies*, their prognosis is very poor.

(j) Meconium ileus is rare: it occurs in babies with cystic fibrosis (mucoviscidosis) because absence of proteolytic enzymes from the pancreas makes the intestinal content putty-like, and failure of mucus production from the intestinal mucosa leaves it unlubricated.

The baby's sweat has high levels of sodium (>80mM). There is bile-stained vomiting and gross abdominal distension, with visible peristalsis and often palpable compressible bead-like stool in the intestines. Erect AP radiographs rarely show fluid levels, and distended loops of bowel may vary greatly from one part of the abdomen to another. You may be able to detect the hyperechoic intraluminal bowel content on ultrasound. Occasionally the baby is born with a red swollen abdomen, indicating a perforation *in utero* which has walled itself off.

You can often break up the thickened meconium with a warmed gastrografin enema (which may show a thin long micro-colon), but you need an IV infusion running to correct fluid shifts, and *must beware of bowel perforation*. *Never use barium* because this can block the bowel solid! Continue with repeated rectal washouts with warm water. Rarely you might need to perform a washout *via* an ileostomy (11-12G).

33.3 Operating on a neonatal acute abdomen

Anastomosing neonatal bowel is no easy task, but if you are surgically dextrous, are well experienced with adult bowel, and have the right sutures and devoted nurses, you may succeed. You have one advantage: the contents of a neonate's bowel are sterile, so that contamination of the abdominal cavity is less of a hazard than it is in an adult. Whatever the difficulties, you may be sure that if the operation is not done, death is certain.

LAPAROTOMY FOR NEONATAL BOWEL OBSTRUCTION (GRADE 3.5)

Make a transverse muscle-cutting incision (11-1), as this breaks down less often than a midline opening. Use the finest haemostats, and handle the bowel with the greatest care (11.3). Hold it with stay sutures, and *don't use bowel clamps* unless you have vascular 'bulldog or Blalock' clamps (3-2). Cover exposed bowel with moist warm swabs. Examine the whole bowel, because there may be more than one area of obstruction. Inject air into the distal bowel through the wall with a very fine needle and milk it along to see if there are any further sites of stenosis. Once you have opened both limbs of the bowel between stay sutures, insert a small tube and flush both clear of inspissated meconium completely and repeatedly, taking care not to spill the contents. By doing this, you will be able to check for any further areas of stenosis or atresia. Furthermore, in the dilated proximal segment you will note where bowel movement is present and where you can expect recovery. In the distal unused part, irrigation will provide some dilation and lubrication for subsequent passage of stools.

The proximal bowel is hugely distended and the distal bowel collapsed, so a straightforward anastomosis is difficult. Tailor the ends so that their sizes are a little more equal by dividing the distal part at 45° (33-1D) and make a cut on the anti-mesenteric border to give it a V-shape (33-1D). Make a single layer of instrument-tied 4/0 or 5/0 long-acting absorbable sutures (33-1E). Gently invert the posterior wall of the bowel, as you insert the first sutures.

If you can do this satisfactorily, inverting the anterior wall should not be difficult. As you go round the corner from the posterior wall of the anastomosis to the anterior, continue to invert the bowel wall. Suture the serosa if appropriate. Always check that the anastomosis is free of tension, watertight and well vascularized.

A child grows, so suture the bowel and the wound with absorbable sutures. Suture everything meticulously.

For DUODENAL ATRESIA, the best operation is a duodeno-duodenostomy. Bring the proximal (dilated) and distal (collapsed) duodenal loops next to each other by carefully separating adhesions to the liver, gallbladder, bile duct and pancreas. Where there is a significant gap between the atretic segments, you can usually mobilize the distal part, together with the duodeno-jejunal junction to the right behind the superior mesenteric vessels. If there is an annular pancreas (very rare), *don't divide it*: mobilize the duodenum enough to make an anastomosis in front of the pancreatic ring.

Aim to make a large (2-3 cm) diamond-shaped side-to-side anastomosis between them (33-1B), by making a transverse incision in the dilated and a longitudinal incision in the collapsed segment. If you encounter an intraluminal web, cut this out carefully leaving a thin rim circumferentially.

Pass a thin, soft, long nasojejunal tube through the anastomosis for feeding post-operatively. Stiff plastic tubes cause problems, so use a soft silicone tube (VP shunt tubes work well). You will also need a nasogastric tube for suction.

Occasionally, you will not be able to perform a duodeno-duodenostomy if the atresia is too distal; then an end-to-side retrocolic duodeno-jejunostomy is the alternative. *Don't fashion a gastrojejunostomy* as this will result in anastomotic peptic ulceration later.

If you cannot do either, fashion a feeding jejunostomy (11.7) to bide time till you can arrange the definitive surgery.

For JEJUNAL ATRESIA, you will have to anastomose the dilated jejunum above the stenosed segment to the collapsed jejunum below it. The blood supply of both ends of the affected bowel is often deficient, so resect it proximally at a point where it is about 1-1.5cm in diameter, *i.e.* normal. Fashion a 'fish-mouth' opening so as to make an 'end-to-back' anastomosis as described above (33-1D,E).

Don't make an end-to-side anastomosis because this results in the blind-loop syndrome (bacterial overgrowth leading to malabsorption).

Make quite sure there are no further stenotic segments by injecting saline into the distal bowel lumen and massaging it into the caecum. If there are (in 20%), try to include them all by doing one resection, if this does not remove excessive bowel. Otherwise make multiple resections or multiple stomas. Decompress the proximal intestine with suction through a Ch12 Foley catheter. *Don't try to milk intestinal contents into the stomach, as they may spill out through the oesophagus into the trachea.*

For VOLVULUS OF THE MIDGUT, when you open the abdomen, you will see distended coils of small bowel, which may be cyanotic and congested, and obscure the right colon. Deliver the small bowel to the surface, and protect it with warm moist packs. Examine the base of the mesentery to see which way it has twisted (usually clockwise). Untwist it (there may be 2 twists), and check that its normal colour returns.

There may be an incomplete malrotation where the caecum is found in the right hypochondrium fixed by Ladd's bands passing across the 2nd and 3rd parts of the duodenum to the lateral abdominal wall. This means the caecum is very close to the duodeno-jejunal flexure, and the base of the mesentery is narrow and so prone to twist. Divide the constricting bands (33-2): there are frequently more than one. Free up the ligament of Treitz, and so straighten the duodenal loop. Then carefully separate all adhesions between loops of bowel, and make sure the duodenum is not kinked, and is patent. Do this by manipulating the stomach contents through into the small bowel. Make sure that the caecum is now resting in the *left* iliac fossa. Remove the appendix attached, because of its subsequent abnormal position.

DIVIDING LADD'S BANDS

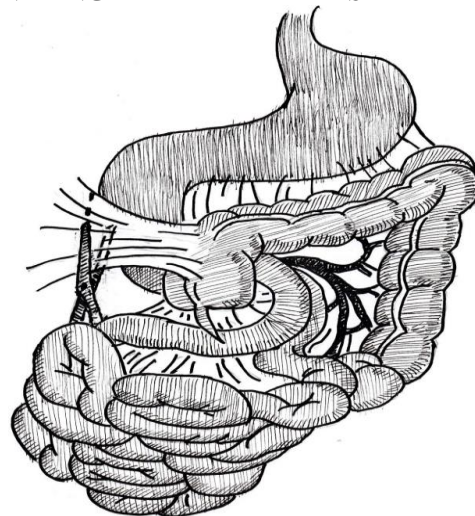


Fig. 33-2 DIVISION OF LADD'S BANDS. Incomplete malrotation where the caecum lies in the right hypochondrium, tethered by 'bands' to the right lateral abdominal wall; these may obstruct the duodenum.

After Greenfield LJ et al. Surgery. Lipincott 2nd ed 1997 p.2057 Fig 103-30.

Rarely, the malrotation is reversed so that the transverse colon lies behind the mesenteric vessels; usually it is possible to rotate the whole bowel through 360° in an anticlockwise manner to correct this.

If the bowel is not viable, resect the gangrenous part, decompress the proximal bowel and anastomose viable bowel or fashion a stoma. If you are not sure about the viability, place warm sponges over the bowel for 15mins and then reassess it. If you have to resect much bowel, the outlook is poor, because of the consequent short bowel syndrome.

There is nothing to be gained by fixing the caecum or colon in their new positions; this may actually do harm. However, do remove the appendix!

Occasionally, bowel twists around a remnant of the omphalomesenteric (vitelline) duct; divide this and assess the bowel viability as before.

Difficulties with neonatal bowel obstruction

If you find a tube (often solid) of duplicated small bowel, resect both the 'normal' and duplicated segments; *don't try to separate them* as the blood supply of the normal part will be compromised.

Don't drain the duplication into the normal bowel as malignant change may result later on. If the duplication is one large cyst, open it and try to strip its mucosa.

If you find extensive or patchy gangrenous bowel without volvulus, this is neonatal necrotizing enterocolitis (33.2). Resect or exteriorize the gangrenous part, and make a spectacles colostomy (11-14). The prognosis is poor, particularly if there is HIV disease. Resuscitate aggressively, replace electrolyte deficits, correct acidosis and use IV gentamicin, ampicillin and metronidazole.

33.4 Omphalocele (exomphalos) and gastroschisis

Not uncommonly, a child is born with a defect in the abdominal wall which involves the umbilicus, and leaves the viscera exposed or covered only by a translucent layer. In **hernia into the cord**, there is a fascial opening where bowel protrudes into the umbilicus; in **omphalocele minor**, there are only a few loops of bowel inside a flimsy sac of peritoneum and amnion; but in **omphalocele major** (33-3), it may contain most of the abdominal organs, including even the liver. Often there are no other defects, but you should always look for them, especially cardiac anomalies.

The sac of an omphalocele is thin, and easily ruptures or becomes infected, causing peritonitis. It rarely ruptures during delivery, but then careful management is urgent.

If the umbilical cord appears hollow, a patent vitelline duct is attached (33.4,5). If it is lumpy, there may be a urachal remnant attached. There may be macroglossia and/or gigantism in <10%, and other serious abnormalities associated with the Beckwith-Wiedemann syndrome.

In **gastroschisis**, there is no membrane covering the viscera, which protrude to the *right* of the umbilicus. The stomach and intestines, but not the liver, herniate and are usually grossly thickened and shortened with a leathery appearance. There is rapid heat loss through the exposed intestines. There is often short bowel and sometimes ischaemic loops.

MANAGEMENT

You may have been able to make the diagnosis prenatally by ultrasound. If so, have everything prepared before delivery. As soon as the child is born, examine the baby carefully in a warm environment with sterile gloves.

If the membrane is intact and the viscera are covered, delay surgical intervention. Non-operative treatment is simple but needs particularly good nursing care, and has the disadvantage of not being able to inspect the internal organs.

Clean the sac with an antiseptic, and apply 70% alcohol or 1% gentian violet hourly for the 1st 48h, and then less often as a crust forms which takes about 3wks. The crust will separate from the periphery, as epithelialization takes place. This will then result in a large skin-covered ventral hernia (33-3H) which will need repair later (18.10).

N.B. Don't put dry dressings on an intact omphalocele: you may tear the thin membrane when you change the dressings. It is best left exposed (under a mosquito net). If, however, you intend to treat the baby as an outpatient, you will need to apply paraffin gauze covered by large padded dressings to avoid trauma to the thin sac.

If the membrane is ruptured or the defect is a gastroschisis, wrap the baby up well, preferably in a large sterile plastic bag encasing the legs with a string tied around the chest (33-4), and *keep him warm*. Start an infusion of warm IV saline, administer ampicillin 10mg/kg and metronidazole 7.5mg/kg IV. Insert a nasogastric tube and suction every 10mins (or with low-grade aspiration), and also pass a rectal tube and perform warm irrigations of the bowel. Prepare theatre.

Don't try to reduce the abdominal contents unless you find an omphalocele minor: just cover them with a silo, made from a pre-washed *sterilized female condom (without spermicide)*. This should be transparent so you can see what is happening inside, and of the smallest possible size to make a good fit around the exposed viscera.

Place these within the silo gently, taking time for oedema to reduce, making sure that you aspirate the stomach at the same time. Tuck the open end of the silo with its ring into the peritoneal cavity under the abdominal wall skin, and try to fix it under the abdominal wall. Extend the defect by incisions superiorly and inferiorly.

Retract the abdominal wall edges and suspend the silo gently from above the bed and wait for the bowel to reduce spontaneously (usually 5 days), and then wait 24h before removing the silo. The defect will close spontaneously if you pull the umbilical cord to the right over it. Monitor the neonate for respiratory problems from pulling up the diaphragm or increased intra-abdominal pressure, and start feeding as soon as the reduction is complete.

These babies are a serious challenge, and if you have the option of transfer to a specialist centre where the babies can be fed intravenously, do so! *But always keep the baby warm!*

OMPHALOCOELE MAJOR

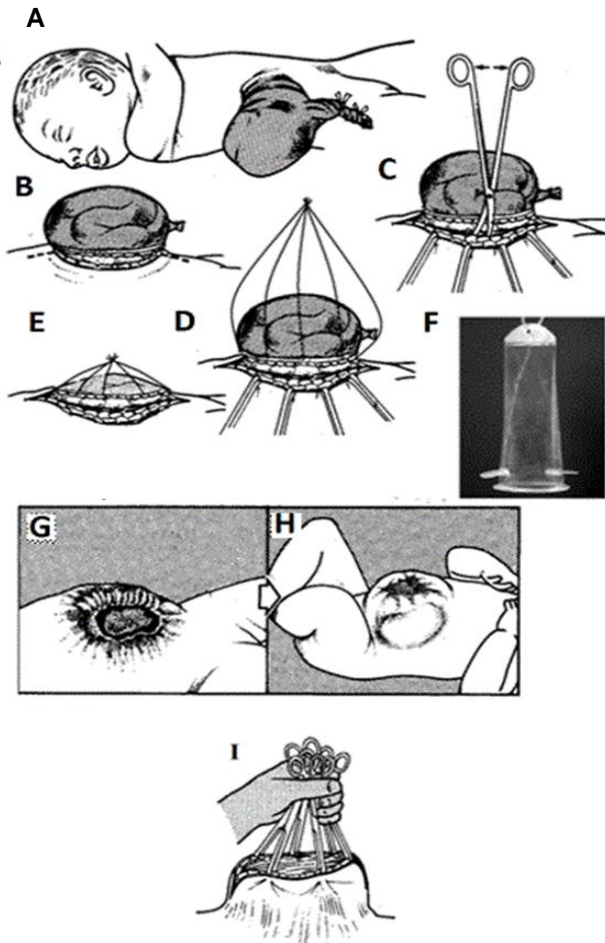


Fig. 33-3 OMPHALOCOELE.

A, this hernia is too large to reduce in a single stage (omphalocele major); its sac is intact. B, extend the opening superiorly and inferiorly. C, mobilize the edges of the sac. D, suture a silo pouch (*left transparent in this illustration*) to the edges of the defect. E, reduce the sac in volume gradually over days, by gently twisting the pouch. F, for gastroschisis, a silo made from a female condom with skin tabs attached at the sides to fix under the abdominal wall, works well. G,H, early and late stages in the non-operative treatment with gentian violet. I, when the edges come together readily, close the abdominal wall.

Partly after Mustard WJ, Ravitch MM (eds). *Paediatric Surgery. Yearbook Medical 3rd ed 1978, permission requested.*

If there is an omphalocele minor and you succeed in reducing the contents of the sac into the abdominal cavity with ease, you can proceed to close the defect; but do so only if you have good back-up, because the non-operative method works just as well.

However, operation may be difficult if the liver is adherent to the membrane, if there is a patent vitelline duct, and if reduction of the prolapsed viscera causes respiratory difficulty. You may, moreover, have little chance to do anything about any abnormalities found anyway!

GASTROSCHISIS TREATMENT USING A STERILE SILO

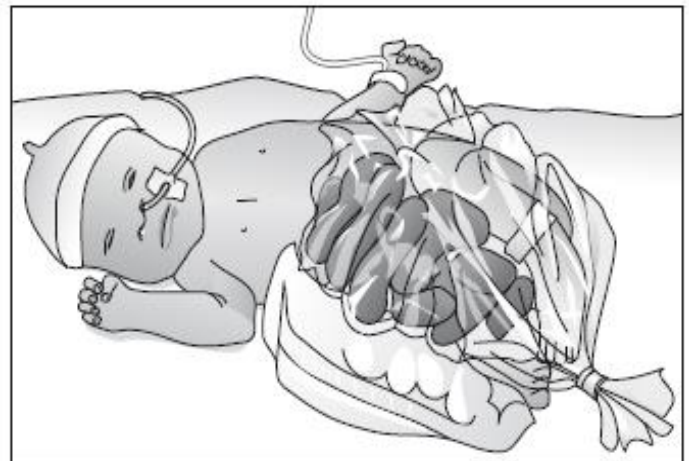
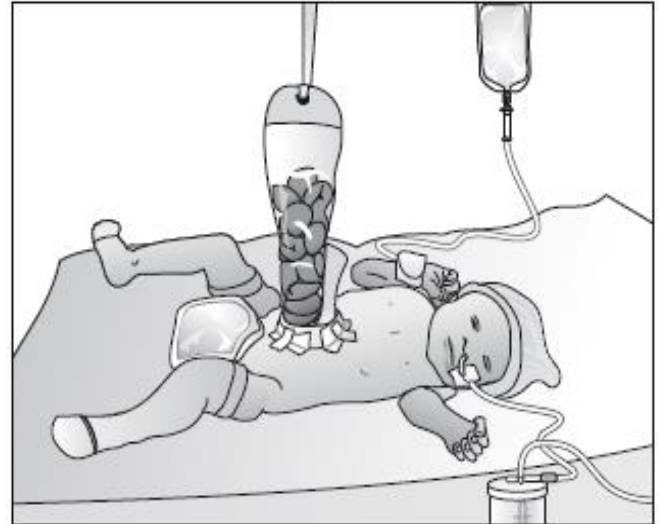


Fig. 33-4 EXPOSED BOWEL IN A NEONATE.

Use this method for ruptured omphalocele or gastroschisis: enclose the exposed bowel in a sterile silo suspended from above and make sure the baby stays warm (note head and legs covered). If the bowel does not fit in a suspended silo, encase the whole abdomen and legs in a sterile plastic bag.

SURGERY FOR OMPHALOCOELE (GRADE 3.3)

Prepare the skin, including the omphalocele sac. Tie and divide the cord, as it emerges from the sac, with '0' absorbable suture (monofilament cuts through too easily). Incise the skin ½cm, or less, from the edge of the defect.

At the edge of the sac, find and tie the umbilical arteries infero-laterally in the 5 & 7 o'clock positions; tie the umbilical vein superiorly (in the 12 o'clock position). Expose the edge of the fascia and peritoneum, and remove a ring of tissue. Excise any tissue of doubtful viability from the sac wall.

Reduce the hernia, and close the defect in three layers. If necessary, hold up the edges of the defect with haemostats. Close the abdomen with long-lasting absorbable sutures and the skin with simple sutures of 4/0 monofilament. Post-operatively, feed with expressed breast milk by nasogastric tube, until the baby is sucking well, usually in 2-3 days.

If you fail to reduce the contents of the sac, or if the sac is ruptured, leave as much of it intact as possible. Find and tie the vessels as above. Excise any tissue of doubtful viability from the wall of the sac, including the stumps of the vessels.

Stretch the abdominal wall with your fingers to allow as much of the viscera to reduce as possible; free any adhesions of the sac from the liver.

As for gastroschisis above, fix up a silo; gently twist this to reduce the visceral content after 5 days, every 2-3 days. When you can reduce the omphalocele easily, you can remove the bag and close the defect as above.

33.5 Disorders of the omphalomesenteric (vitelline) duct

In foetal life, the intestinal tract and the yolk sac are joined by the omphalomesenteric (vitelline) duct. Remnants of this may persist and present as:

- (1) A persistent discharge from the umbilicus which is occasionally faecal.
- (2) Gastrointestinal bleeding from ectopic gastric mucosa in a Meckel's diverticulum (33-5I, J).
- (3) Bowel obstruction caused by bowel twisting around a persistent vitelline duct (33.3, 33-5F).
- (4) Intussusception through a patent vitelline duct (rare). You may occasionally have to resect and anastomose bowel in connection with any of these, or, rarely, with the other abnormalities (33-5).

Difficulties with umbilical discharge

If a child discharges urine from the umbilicus and the urethra, there is a persistent connection to the bladder: a URACHUS (rare). Sometimes urine discharges in a small spurt during micturition. You may demonstrate this using a gentle probe or guide wire, or with contrast. Presentation may be at 1yr or later. *Dissecting out and excising the track does not require a full laparotomy.*

If there is a dirty umbilicus which discharges, smells, and occasionally bleeds, this is an UMBILICAL GRANULOMA (very common), caused by an infected remnant of the cord. Clean it with spirit daily, apply zinc powder, and keep it dry.

Treat an infected umbilicus (omphalitis) in the acute stage with gentamicin, and add metronidazole if it is smelly. *Watch out for signs of necrotizing fasciitis (6.23), and beware of bowel evisceration!*

ABNORMALITIES OF THE OMPHALOMESENTERIC DUCT

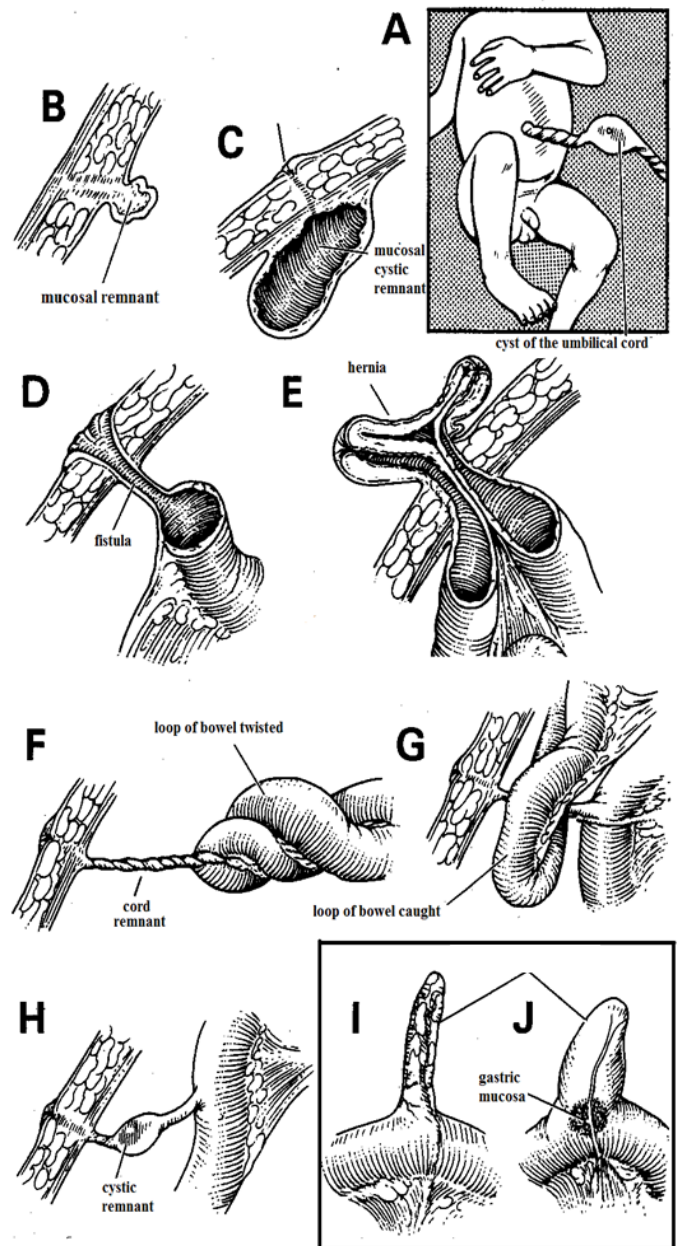


Fig. 33-5 ABNORMALITIES OF THE OMPHALOMESENTERIC (vitelline) DUCT are usually rare.

A, harmless cyst in the cord. B, umbilicus covered with red intestinal mucosa, which may dip down into the abdominal wall. *Don't confuse this with an umbilical granuloma, which is much more common.* C, mucosa-lined cyst communicating with the skin. D, communication between the ileum and the umbilicus. E, bowel herniating through this communication. F, persisting cord around which bowel may twist (33.3), or G, bowel may be caught. H, a solitary cyst within the cord. I, remnants of the duct persisting as Meckel's diverticulum, which may become inflamed. J, some gastric mucosa in the duct ulcerating and bleeding.

Adapted from Mustard WJ & Ravitch MM (eds). Paediatric Surgery, 3rd ed. 1978 Fig.88-17, Yearbook Medical, with kind permission.

33.6 Anorectal malformations

Anorectal malformations are relatively common. The child presents soon after birth with abdominal distension and the failure to pass meconium. Some lesions are incomplete, and present later with difficulty passing faeces, or distension. Around 15-40% of these children have one or more other anomalies, particularly vertebral, cardiac, oesophageal, renal and skeletal. Take time to record your findings carefully.

ANORECTAL MALFORMATIONS

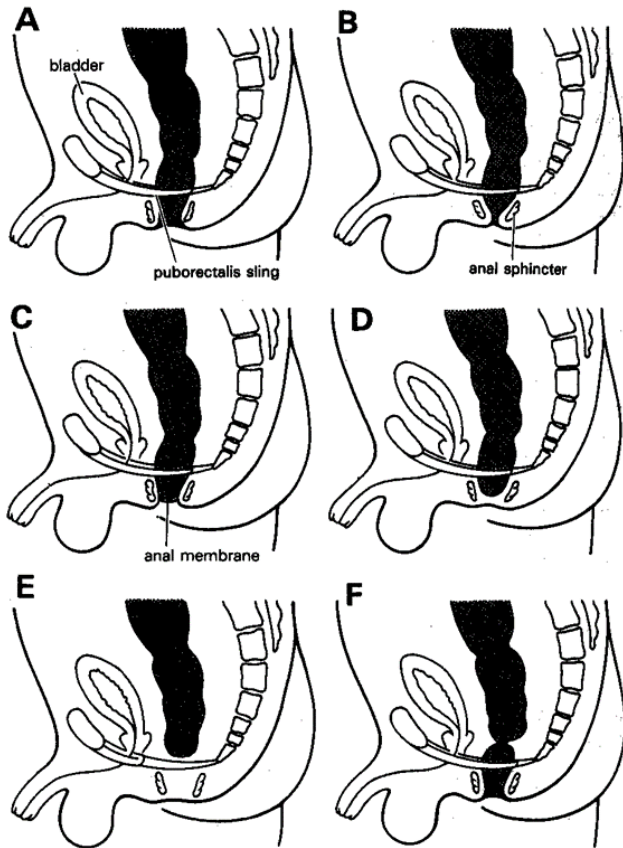


Fig. 33-6 ANORECTAL MALFORMATIONS.

Low lesions (B-D) have rectum extending below the *puborectalis* sling (*infra-levator*), high ones (E,F) remain above it (*supra-levator*). The low lesions are much easier to treat. Function a colostomy for the high lesions.

Adapted from Mustard WJ & Ravitch MM (eds). *Paediatric Surgery*. Yearbook Medical 3rd ed 1978 Fig. 98-2 with kind permission.

There are many kinds of lesions, but what matters most is whether a child's rectum ends close or far from the skin. If the rectum extends close (<1cm) to the skin (*infra-levator* lesions), you can create an anus, or dilate a stenosed anus relatively easily. But if the rectum ends further from the skin (*intra-* and *supra-levator* lesions), make a temporary colostomy, and arrange later for the sigmoid colon to be pulled through to the perineum, or an anorectoplasty to be performed. This is difficult surgery, and continence is often not perfect.

The anus or rectum may fail to develop entirely (agenesis), it may partly fail to develop (atresia), or the rectum or anus may be narrowed (stenosis). Agenesis (but not the other lesions) may be combined with fistulae between the rectum and the urinary or genital tracts of either sex. These variables combine to produce a complex series of lesions. Some fistulae are useful, because you may be able to dilate them to make an anus. About 75% of cases are low lesions with fistulae.

(a) Low anorectal lesions:

(1) Anal stenosis or imperforate anal membrane in boys or girls (33-6B,C).

(2) Anocutaneous fistula in boys and girls.

(3) Anovestibular fistula in girls (33-8).

(b) Intermediate anorectal lesions:

(1) Rectobulbar urethral fistula in boys; rectovestibular and rectovaginal fistula in girls.

(2) Anal agenesis without fistula in boys and girls (33-6D).

(c) High anorectal lesions:

(1) Anorectal agenesis with rectoprostatic, urethral or rectovesical fistula in boys; or rectovaginal fistula in girls.

(2) Anorectal agenesis without fistula in boys and girls (33-6E).

(3) Rectal atresia in boys and girls (33-6F). There is a normal-looking anus in the normal place, with low bowel obstruction (no meconium, abdominal distension and only vomiting late in the presentation).

N.B. A contrast enema is safer than probing in this situation.

(4) Cloaca (a combined opening of urethra, vagina and rectum) in girls. This looks like a 'one-hole' perineum.

You should be able to diagnose which kind of lesion a child has from the clinical signs and a simple radiograph (33-7) or ultrasound. The risk in trying to repair an anorectal lesion is that, *if the lesion is higher than you expect, you may significantly damage the rectal anatomy. Unless you find the rectal stump just underneath skin level, stop, make a colostomy, and arrange for repair later. Don't divide any muscle.* If you are in any doubt, a colostomy would be wiser, even though you may occasionally fashion one unnecessarily. *The penalty for failure is incontinence.* Because bowel washout for the definitive procedure is much more effective with a sigmoid colostomy, do this rather than a transverse colostomy.

Examination

Ask your midwives to examine the anus in all children. Ask them to refer:

(1) any child with an abnormal anus,

(2) any child who passes no faeces for 12h, and whose abdomen distends. Examine him, and if necessary, pass a rectal thermometer or a stiff catheter.

If there is no anus, make an opening before the bowel distends. Look for other congenital abnormalities.

If there is a mass of irregular epithelium where the anus should be, the diagnosis is almost certainly anal stenosis. Probe it and look for even a trace of meconium to confirm the presence of an opening, however tiny.

If there is a thin veil of epithelium overlying the anal orifice, surrounded by normal skin folds and *rugae*, there is an imperforate anal membrane.

If the anus looks normal, until you put a probe into it, when you find that the rectum is almost or completely blocked, the diagnosis is rectal atresia. You may find a very small hole which you can dilate.

If the anal skin is smooth, there must be some kind of lesion other than stenosis.

LOOK FOR FISTULAE:

Check for meconium in the urine, or in the vagina or vulvar vestibule. Look for a dark blob of meconium under the skin, especially in the midline *raphe*. A perineal ultrasound scan, demonstrating a fluid-filled track, may well help you in diagnosing a fistula.

INVERTOGRAMS OF IMPERFORATE ANUS

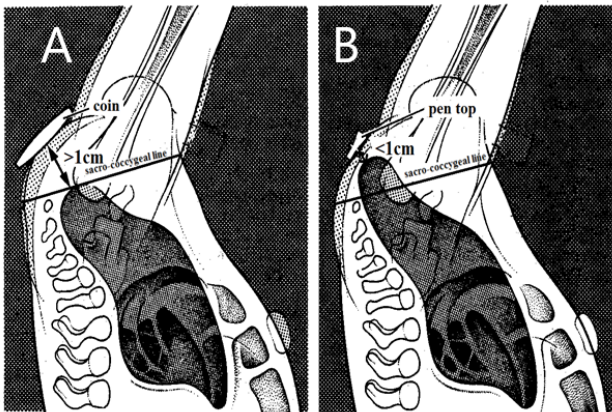


Fig. 33-7 RADIOGRAPHS OF AN 'IMPERFORATE' ANUS showing high (A) and low (B) rectal lesions. Lift up the child's legs and buttocks and stick a piece of metal over the anus. A, high lesion with the gas bubble in the rectum, below the sacro-coccygeal line, >1cm from a coin placed over the anus. B, low lesion with a pen top on the anus <1cm from the gas bubble.

Radiographs (INVERTOGRAM, 33-7)

Wait until 12-16h after birth, when the gas the child has swallowed has reached the blind lower end of the bowel. Place the baby prone on a pillow so that the anus is uppermost. Flex the knees. Strap a small piece of metal such as a paper clip flat on the skin where the anus should be.

Take a lateral film. The bubble of gas in the bowel will be uppermost. The distance between it and the metal will show you how much tissue there is between the bottom of the bowel and the skin.

Interpret the films as follows: if the gas bubble is <1cm from the anus, this is a low lesion; if it is >1cm from the skin, it probably is a high lesion. More accurately, draw a line between the posterior part of the pubis and the coccyx.

If you see any blind dilated bowel distal to this line, this is a low lesion. If it remains proximal to this line, the lesion is high. Anything in between is intermediate.

CAUTION!

Radiographs are useful *but not completely reliable*. There may not have been enough gas in the bowel, or there may be a fistula higher up.

Ultrasound shows the rectum as a hypo-echoic structure. There may be a rectovesical fistula and so gas producing a fluid level in the bladder (uncommon). Exclude this by taking a supine lateral film.

Intravenous urography is not indicated at the early stage.

A fistulogram may provide important information, though.

Management

If a child has a low lesion, operate early. The obstructed bowel is going to distend, so, as soon as he has swallowed enough air to help make the radiographic diagnosis, pass a nasogastric tube. Maintain hydration and add 1mg IM vitamin K₁.

CAUTION! If you are in doubt, fashion a colostomy.

(1) ANAL STENOSIS AND IMPERFORATE ANAL MEMBRANE (boys and girls)

ANAL DILATION (GRADE 1.2):

Incise the epithelial covering of the anus if there is one. If the anal opening is very small, use a filiform urethral catheter with a metal follower or the smallest Hegar dilator. Teach a parent to dilate the child's anus daily, then every other day, then weekly with the little finger, and provide a supply of disposable gloves to do this.

(2) ANOCUTANEOUS FISTULA (boys and girls) AND ANOVESTIBULAR FISTULA (girls)

CUTBACK OPERATION (33-8B, C) (GRADE 1.4):

Introduce a grooved dissector through the fistula and point it posteriorly strictly in the midline. Cut down onto the dissector to the point where the anus should be.

N.B. If you cut deeply, you may divide sphincter muscle!

Suture the anal mucosa to the skin, using interrupted long-lasting absorbable sutures. Dilate the anus as in (1) after 5 days. You can deal with the 'anterior ectopic anus' (really a type of anocutaneous fistula) in the same way with a cutback operation.

(3) RECTOVAGINAL AND RECTOVESTIBULAR FISTULA (girls)

In practice it is difficult to differentiate between the high and the intermediate anorectal agenesis. Where there is a rectovaginal fistula, the lesion tends to be lower. However, it is of no great consequence in the neonatal period. Dilate the fistula tract with Hegar's dilators. *Make sure the track does not stenose:* repeat a dilation every 3 months.

An anoplasty may be required when the child is >1yr. If this is impractical, it is not a disaster, but it will mean that she will always defecate from the fourchette. This is much less of a disability than it might seem; it is, for example, compatible with a normal sex life, and the husband may be quite unaware of it! However, some faecal incontinence may always be a problem.

ANOCUTANEOUS FISTULA

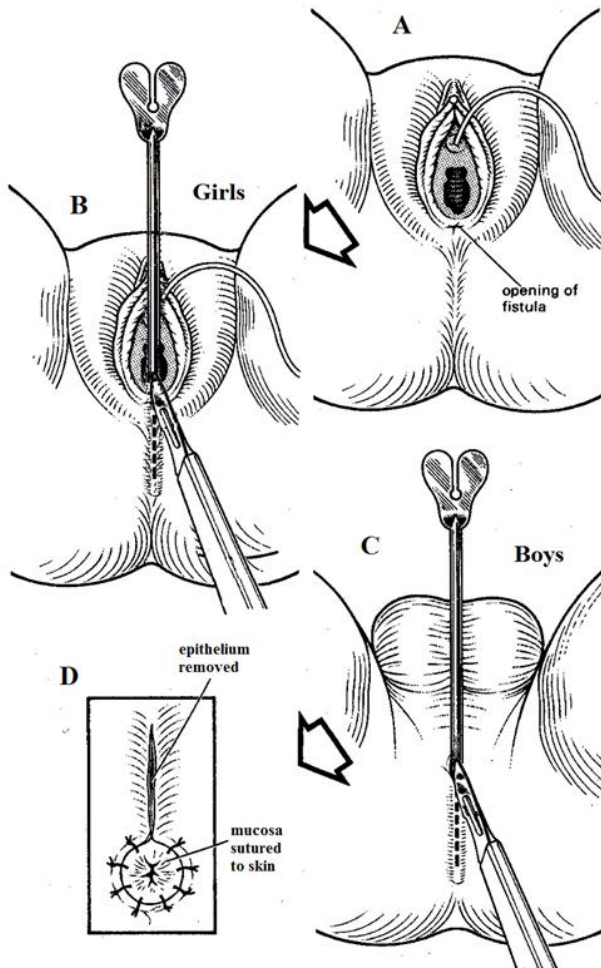


Fig. 33-8 ANOCUTANEOUS FISTULA. A, fistula opening close to the introitus. B,C, cutback operation; note that the director points posteriorly, and remains superficial. Cut along this grooved director to where the anus should be. D, remove epithelium of the track, pull out the anal mucosa and suture it to the skin.

Adapted from Mustard WJ & Ravitch MM (eds). *Paediatric Surgery*. 3rd ed 1978 Fig.98-4. Yearbook Medical, with kind permission.

(4) ALL OTHER ANORECTAL LESIONS

Create a SIGMOID COLOSTOMY (11.6). Make a small Lanz-type transverse incision (11-1) in the left lower abdomen and carefully retract the muscle layers until you reach the peritoneum. Open this very carefully; underneath you should find colon; gently pull some out, perforate it and aspirate its contents. This will reduce abdominal distension and hopefully allow you to extract enough bowel to make a skin bridge to let you really divide the colon. If you cannot do this, fashion a loop colostomy.

Wash the distal colon free of meconium with warm saline: *if you don't do this early, the meconium will solidify like concrete!* Make sure you do frequent washouts if there is a recto-urinary fistula in order to minimize urinary tract infection.

Don't perform a formal laparotomy: it is difficult and unnecessary and the baby may well not survive it.

You don't need stoma bags for a neonate! Wrap the nappy over the stoma, and advise mother to clean gently around the stoma as she would for the anus.

Don't be tempted to operate on the anus of any intermediate or high anorectal agenesis; leave fashioning a neat anoplasty to an expert. This will not be possible if you have damaged the sphincter musculature.

Difficulties with anorectal malformations

If the anus becomes stenosed, it has become fibrosed. Unfortunately, regular dilation perpetuates the cause of the fibrosis. A formal anoplasty will eventually be needed.

If there is a mucosal prolapse or skin excoriation after an anal operation, apply talc powder and carefully prevent soiling of the perineum. The prolapse will reduce spontaneously.

If excess fluid is lost from a colostomy, wait for 14 days, while adding extra fluid to maintain a satisfactory fluid balance. The stools will usually become formed.

For other colostomy complications, see 11.6.

33.7 Hirschsprung's disease

In this not-uncommon disease, the neuroganglion cells in the wall of the rectum are absent, so that faeces are not propelled onwards as they should be. The length of the aganglionic segment varies although it is usually confined to the rectosigmoid in 75%, but may be tiny (ultra-short segment) or extend to the caecum or beyond (c.5%). This results in dilation and hypertrophy of the proximal (neurologically intact) segment with transition to a normal-sized or narrowed distal (aganglionic) segment. The child becomes constipated, and the abdomen distends; and in acute cases the bowel may obstruct. There may be a meconium plug in the rectum.

(a) Subacute presentation: a child presents soon after birth with subacute intestinal obstruction; its onset is more gradual than with complete mechanical obstruction, but may still be fairly acute. There is usually a history of delayed passage of meconium >24h after birth.

Rectal examination often results in explosive projectile passage of meconium or faeces, which may result in visible deflation of the abdomen. Abdominal distension is always present, and may be quite alarming if there is enterocolitis or bowel perforation (20%), resulting in a TOXIC MEGACOLON. There are then signs of peritoneal inflammation. *In this situation, repeated washouts will make the situation worse!*

(b) Late presentation: an older child has a chronic history of constipation *without laxative abuse or psychosocial problems*. The abdominal distension may be gross and affect breathing. Intermittent bowel actions tend to be explosively incontinent and infrequent.

(c) Investigations

Abdominal radiographs show multiple loops of distended bowel occupying the whole abdomen, with only occasional air-fluid levels and minimal or no air in the rectum; if there are gas bubbles in the bowel wall, this is a sign of enterocolitis. If there is a pneumoperitoneum (which may be huge), this is a sign of bowel perforation.

A barium (or, safer, gastrografin, which does not become solid) enema may conclusively make the diagnosis: look for a dilated proximal segment of colon and narrow distal rectum. This delineates the 'transition zone'. Repeat the film after 24h to see if there is retained barium proximally. A post-evacuation film can be particularly helpful.

Check the thyroid function: hypothyroidism may totally mimic Hirschsprung's disease.

(d) Full-thickness rectal biopsy (GRADE 1.4)

You need this investigation to make a definitive diagnosis. If the presentation is acute, do it at the same time as a sigmoid colostomy (11.6).

Put the child in lithotomy position under anaesthesia (e.g. ketamine), and dilate the anus with Hegar's dilators. Clean the anorectum with non-alcoholic antiseptic solution, and leave a swab soaked with this inside the rectum, proximal to the operation site. *Remember to record this swab inside!* It is best if you tie a suture to it to hang outside the anus on a mosquito forceps.

Evert the posterior wall of the anus on both sides with Allis forceps and place all-layer stay sutures postero-laterally on both sides at the level of the internal sphincter (33-9A). Retract the anterior wall of the anus upwards, exposing the rectal wall.

Make an incision and excise a 1.5cm triangular 5mm wide full-thickness segment with fine scissors (33-9B), and close this with 4/0 long-acting absorbable *through all layers* of the rectal wall. Remove the gauze you put in the rectum.

FULL-THICKNESS RECTAL BIOPSY

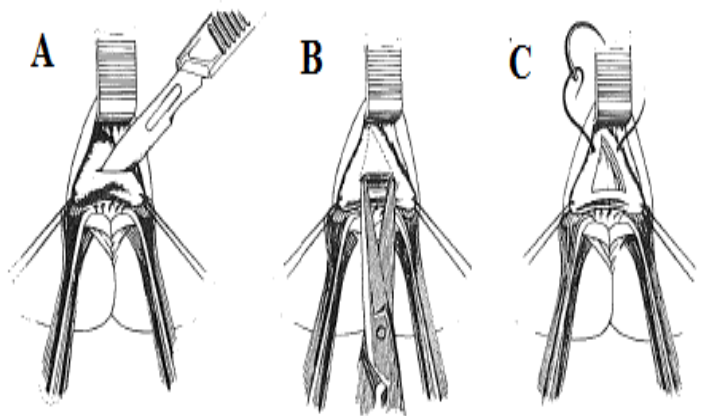


Fig. 33-9 FULL-THICKNESS RECTAL BIOPSY.

A, exposure of rectal mucosa and incision. B, excision of a triangular piece of rectal wall. C, suture of rectal wall defect.

After Lister J, Irving IM. Neonatal Surgery, Butterworth 3rd ed 1990 p.534 Fig 37.10.

N.B. There are more sophisticated methods of getting histological diagnosis; consult colleagues if these are available because a full-thickness rectal biopsy may make the definitive pull-through operation more difficult.

Try to refer such a child for definitive 'pull-through' surgery to an expert, because a repeat intervention for problems hardly ever gives a very good result. In the meantime, show the parents how to perform bd washouts with a soft catheter, using warm clean water. If the delay will be significant, perform a defunctioning transverse colostomy (11.5,6).

33.8 Neonatal jaundice

Most neonatal jaundice is 'medical', and only occasionally is it 'surgical'. Physiological jaundice (with levels of unconjugated bilirubin $>85\mu\text{M}$) fades after 1-2wks; however, levels $>220\mu\text{M}$ (or $>255\mu\text{M}$ in preterm babies) are *not* physiological. *Jaundice after 2wks of life is not physiological.* However, elevated *conjugated* bilirubin levels are surgically significant. The biliary tract can be blocked by epithelial debris or biliary sand (less common), in which case it may be temporary and clear spontaneously, or by atresia of the biliary tree (more common), for which the only hope is surgery. This blockage appears to be caused by a sclerosing inflammatory process starting *in utero*, and is not due to a simple single stenosis. Hepatomegaly is common, and stools may be pigmented initially before becoming white.

If much of the bile duct is not patent (as in 25% of cases), nothing can be done other than very major surgery (the Kasai procedure) by an expert.

If however the bile duct is sufficiently patent for the gallbladder to be distended, and in connection with the proximal duct system (75%), it can be anastomosed to the proximal jejunum.

BILIARY ATRESIA

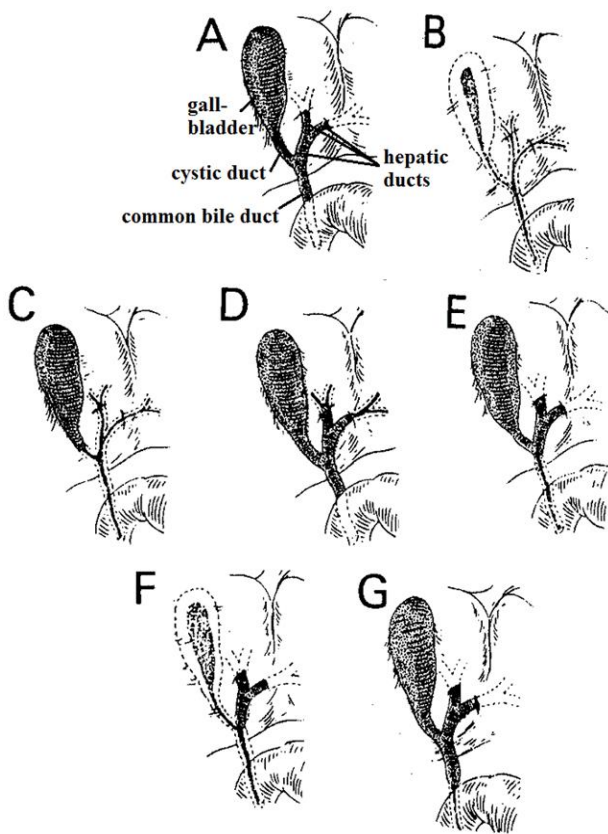


Fig. 33-10 BILE-DUCT ATRESIA.

A, normal. B, total atresia of all ducts. C, atresia of all ducts except the gallbladder. D, intrahepatic atresia. E, atresia of the common bile ducts. F, atresia of the common bile duct and gallbladder. G, segmental atresia of the distal common bile duct.

After Schwartz *SI. Surgical Disease of the Liver*. McGraw-Hill 1964, permission requested.

If there is an extra-hepatic stump, this too can be anastomosed. Without surgery, death is inevitable. Even in experienced hands the outlook is poor, and 30% of children develop liver failure.

Ultrasound (38.2c) can help with the diagnosis: you should be able to differentiate biliary atresia from dilation of the common bile duct (known as a **choledochal cyst**), which itself may obstruct the proximal common bile duct.

The time of onset is of great diagnostic value.

If a child starts to become jaundiced in the 1st 6-24h, jaundice is likely to be haemolytic from:

- (1) Septicaemia from an umbilical infection. Look for signs of infection of the cord and the surrounding tissues (septicaemia from cord infection can also occur later).
- (2) Haemolytic disease of the newborn. Usually, the baby is Rh D+ve, the mother is D-ve and has anti-D antibodies as the result of having had a previous D+ve gestation, or of having been given D+ve blood. Other blood group incompatibilities may have the same effect (*e.g.* ABO).
- (3) Congenital syphilis (severe infection).

If mild jaundice starts at 24-72h, this is likely to be 'physiological jaundice' owing to a deficiency of glucuronidase (common, especially in 'small for dates' babies). This usually clears spontaneously, but can be helped by sunlight and phenobarbitone 1-2mg/kg bd.

If jaundice starts at 24-72h, and becomes severe by the 3rd-5th day, there may be G6PD (glucose-6-phosphate dehydrogenase) deficiency. This is common, especially in Southeast Asia and the Mediterranean littoral; a less severe form occurs in parts of sub-Saharan Africa.

If there is little or no jaundice till 3-6wks, by which time the liver is enlarged, and the stools are clay-coloured, there probably is some 'surgical' reason for the jaundice. Sometimes there is slight jaundice at birth, or a few days later; the urine is dark brown early.

If there are recurrent bouts of cholangitis, there may be a congenital dilation of the common bile duct (choledochal cyst) which may be saccular or diverticular. The best treatment for this is a Roux-en-Y choledochojejunostomy.

33.9 Hypospadias

If a boy's urethra opens on the ventral surface (underside) of the penis, this is a HYPOSPADIAS. There may be a glandular, penile, scrotal or perineal opening (or anywhere in between), often with a chordee (tight band) causing ventral curvature of the penis, especially seen in erection.

If there is perineal hypospadias with undescended testes, examine carefully regarding gender.

HYPOSPADIAS REPAIR: STAGE 1

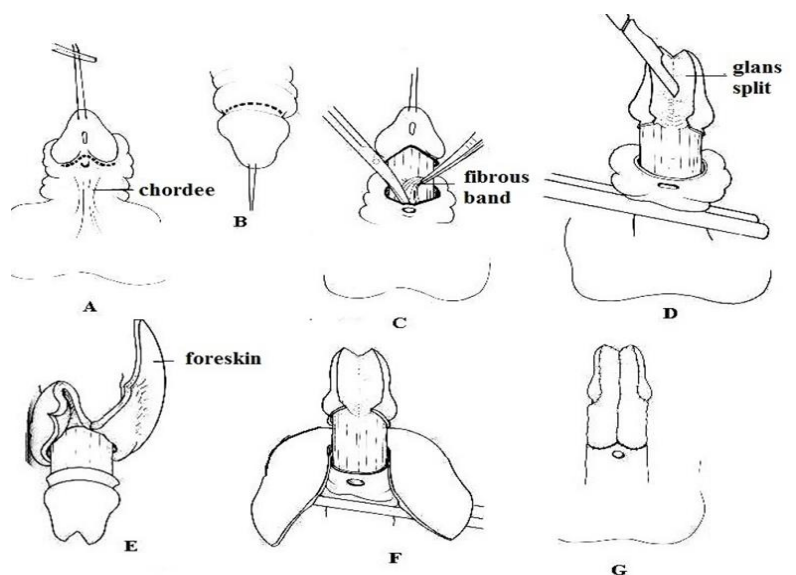


Fig. 33-11 HYPOSPADIAS CORRECTION: 1ST STAGE.

Chordee correction. A, apply a tourniquet. B, correct the chordee: circumferential incision. C, excise the fibrous band. D, slit the glans penis. E,F, divide the dorsal foreskin into 2 long flaps. G, bring flaps round to cover raw ventral penile surface.

After Blandy *J. Operative Urology*, Blackwell 2nd ed 1986 p.206 Figs 14.2-6.

Never circumcise the hooded prepuce in hypospadias patients because it is needed for urethroplasty repair. Do this corrective surgery around 1-2yrs. If there is a chordee, the two-stage procedure is more reliable for the non-expert.

PENO-SCROTAL HYPOSPADIAS CORRECTION: 1ST STAGE (GRADE 3.4)

Make sure the child is thoroughly bathed with antiseptic and bowel action is well controlled.

Place a stay suture at the tip of the penis and a tourniquet at its base. Widen the meatus if it is stenosed either by dilation or a meatotomy (27-29).

Make an incision right round the coronal sulcus (33-11A,B), and dissect the urethra off the *corpora cavernosa*. Firm bands may tether the urethra and these must be excised to correct chordee (33-11C).

See if you have achieved correction by injecting saline in the corpus to create an erection: if not, complete the dissection proximally to the bulb. Then open up the glans penis by incising it up to the tip on the ventral surface in the midline until it looks quite flat (33-11D), and then divide the hooded foreskin on the dorsal surface in the midline, separating its inner and outer layers (33-11E,F).

Bring the two flaps round to cover the raw ventral surface (33-11G) of the penis with 5/0 absorbable sutures and leave in a Ch6 Foley urethral catheter or feeding tube for 4-5 days.

2ND STAGE (GRADE 3.5)

About 6 months later, making sure the child is thoroughly bathed, fashion a suprapubic cystotomy (33-12A). Then mobilize the skin on the ventral surface of the penis and wrap it snugly round a Ch8 feeding tube with 6/0 absorbable sutures, trying to keep the suture line off midline. Try swinging around an intermediate layer of *dartos* tissue from the dorsum over the suture line. Close the outside skin around this suture line, preferably avoiding one suture line lying over the other. Keep the catheter *in situ* 7-10 days with urine dripping freely: *if you attach a urine collection bag, it is likely to pull on your repair and disrupt it!*

Nurse the child in a 'double nappy' with a hole for the genitalia in the inner nappy, in order to separate stools from the operative site.

HYPOSPADIAS REPAIR: STAGE 2

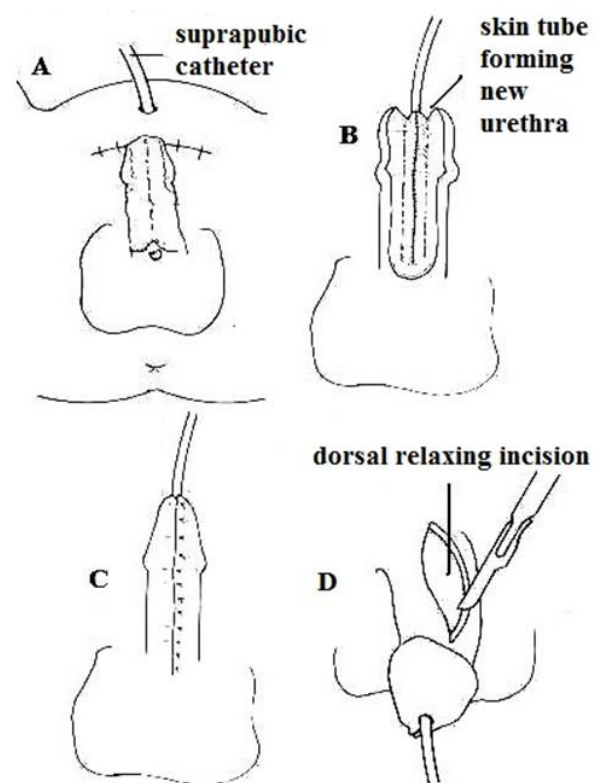


Fig. 33-12 HYPOSPADIAS CORRECTION: 2ND STAGE.

A, insert a suprapubic catheter. B, outline a full-thickness skin tube using a suitable catheter as template for its width. Suture a skin tube over the catheter, preferably not in the midline. C, close glans and penile skin over the skin tube. D, make a dorsal relaxing incision.

After Blandy J. *Operative Urology*, Blackwell 2nd ed 1986 p.208 Figs 14.8-11.

DIFFICULTIES WITH HYPOSPADIAS CORRECTION

If a small urinary fistula develops, make a Y-shaped incision over the hole (33-13A), and mobilize the skin thoroughly to expose the fistula hole (33-13B). Close the hole with an inverting long-acting absorbable 4/0 suture, and then advance the skin over the hole to cover it (33-13C).

If there is complex scarring, chordee and fistulae, the whole scarred urethral segment must be excised and a new urethra created using a pedicled *dartos* scrotal skin flap. This is something for an expert.

URETHRAL FISTULA CLOSURE

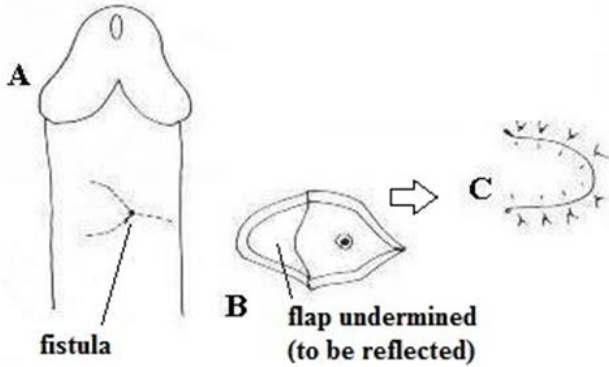


Fig. 33-13 CLOSURE OF URETHRAL FISTULA.
A, Y-shaped incision. B, mobilize the skin to expose the fistula.
C, U-closure of skin.

After Blandy J. *Operative Urology*, Blackwell 2nd ed 1986 p.208 Fig 14.12.

33.10 Spina bifida & encephalocele

Congenital abnormalities of the spinal cord and vertebral column are not uncommon. You may see:

(a) **Spina bifida occulta**, in which the arches of the vertebrae remain open but the skin is closed, usually in the lumbar region. There is often a brownish spot over the defect (this is less easily seen in dark skin, but it is there if you look for it), and/or some extra hair and fatty tissue. Spina bifida occulta is usually symptomless, but the child may develop a tethered cord as he grows, particularly during growth spurt periods: so watch him carefully. If the legs become weak or urinary or faecal incontinence develop, he will need prompt untethering of his spinal cord: leave this to the experts!

(b) A **meningocele** is an extension of the spinal canal, filled with CSF but without any spinal cord or spinal nerves in it; commonly in the lumbar region, and usually associated with spina bifida. The cord is normal and there is no neurological abnormality. It is a relatively simple procedure to obliterate the sac by closing the dura, and then to close the skin. This is a closed lesion, so there is no hurry.

(c) A **myelomeningocele** is more common than a simple meningocele, and takes two forms:

- (1) There is a closed swelling containing spinal cord and/or spinal nerves.
- (2) More often, the spinal canal is open and leaks CSF, with the flattened cord forming a plaque on its surface.

Both varieties may occur in the cervical (rare), thoracolumbar, lumbar, or lumbosacral regions, and other abnormalities are frequent, particularly hydrocephalus. Many children have irreversible paralysis of their legs, and loss of sphincter control, but some can still achieve quality of life through surgery and good follow-up. The decision of whether to operate or not on these children is sometimes difficult and must be made with each family. *Don't operate without discussing the long-term orthopaedic, urinary and faecal continence and psychological problems extensively and repeatedly.*

N.B. A myelomeningocele may be obscured by a lipoma: beware!

(d) An **encephalocele** is a condition where the brain herniates through a cranial defect, usually occipital or naso-frontal. The neural function may be entirely normal, though large defects may be associated with microcephaly and other brain anomalies. Occipital defects without involvement of the medulla oblongata are easier to repair while naso-frontal ones, especially if affecting the olfactory nerve, are much more difficult. Try to get an ultrasound to delineate the true situation, and decide if you had better leave this intervention to an expert.

MENINGOCOELES

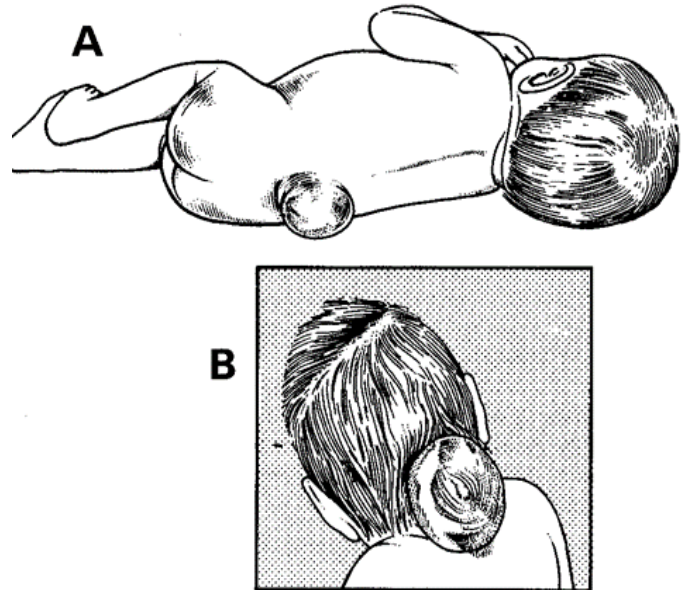


Fig. 33-14 MENINGOCOELES.
A, skin-covered sacrolumbar meningocele. You may be justified in operating on this child. B, cervical meningocele. *Leave this for the experts.*

Diagnosis

Make sure you examine the baby carefully in a warm room, repeatedly if necessary. Look for other abnormalities, especially hydrocephalus (present in 80%: measure the head circumference), and cardiac defects. Ultrasound is very helpful if you can interpret the images.

If the swelling on the child's back is covered by skin, the legs are not weak, the anus is not lax, and he micturates normally, he probably has a simple meningocele or lipomeningocele. Otherwise, you cannot tell a meningocele from a myelomeningocele from its site, or its covering.

If you transilluminate it, you may occasionally see nerves outlined inside it. Ultrasound will give you an idea how much fluid is inside.

If there are any neurological signs, there probably is a myelomeningocele. If it is open, there will almost certainly be some neurological defect such as:

(1) Partial or complete paralysis of buttock, thigh, leg or feet muscles, often with deformities such as *talipes equinovarus* (32.10) or *genu recurvatum*.

(2) An absent or meagre response to tickling or pin-prick (*again tested in a warm, relaxed environment*).

(3) A lax anal sphincter, an absent anal reflex, or a rectal prolapse.

(4) Urinary retention which can be expressed by forceful suprapubic pressure, associated with overflow dribbling, or rarely constant dribbling with an empty bladder.

N.B. Differentiation between voluntary and reflex movement is difficult; if the baby is cold, voluntary movements may disappear!

If there is a low solid sacral lesion which on pressure does not distend the anterior fontanelle, this is likely to be a sacrococcygeal teratoma (33.13). Unlike spina bifida, it tends to displace the anus forwards, and can extend into the abdomen when it is bimanually palpable.

If there is a solid spinal tumour infiltrating bone, this is probably a chordoma, a rare slow-growing tumour of notochord. Resection is the only hope for cure.

Management of meningoceles

If the meningocele is covered with normal skin (usual), surgery is not urgent. Close the defect when the baby is bigger (>6-9 months). This should be done by an expert if the lesion is in the cervical or thoracic region. If the lesion is not fluid-filled, it is likely a lipomeningocele: a very complex lesion that must be repaired within the first 2yrs of life, but only by an expert.

If there is an open meningocele or a myelomeningocele with severe neurological signs (commonly in the thoracolumbar or lumbar regions), there is nothing to be done except compassionate palliation.

Some movement in the legs and anal tone imply incomplete nerve damage: you must explain carefully then to the parents the prospects of the child needing difficult and extensive rehabilitation before embarking on operative intervention. Once you start, you signal a commitment to continue!

Note that closing the defect will *not* improve any neurological deficit. Therefore discuss the situation fully with the family before deciding on surgery.

MYELOMENINGOCELE CLOSURE

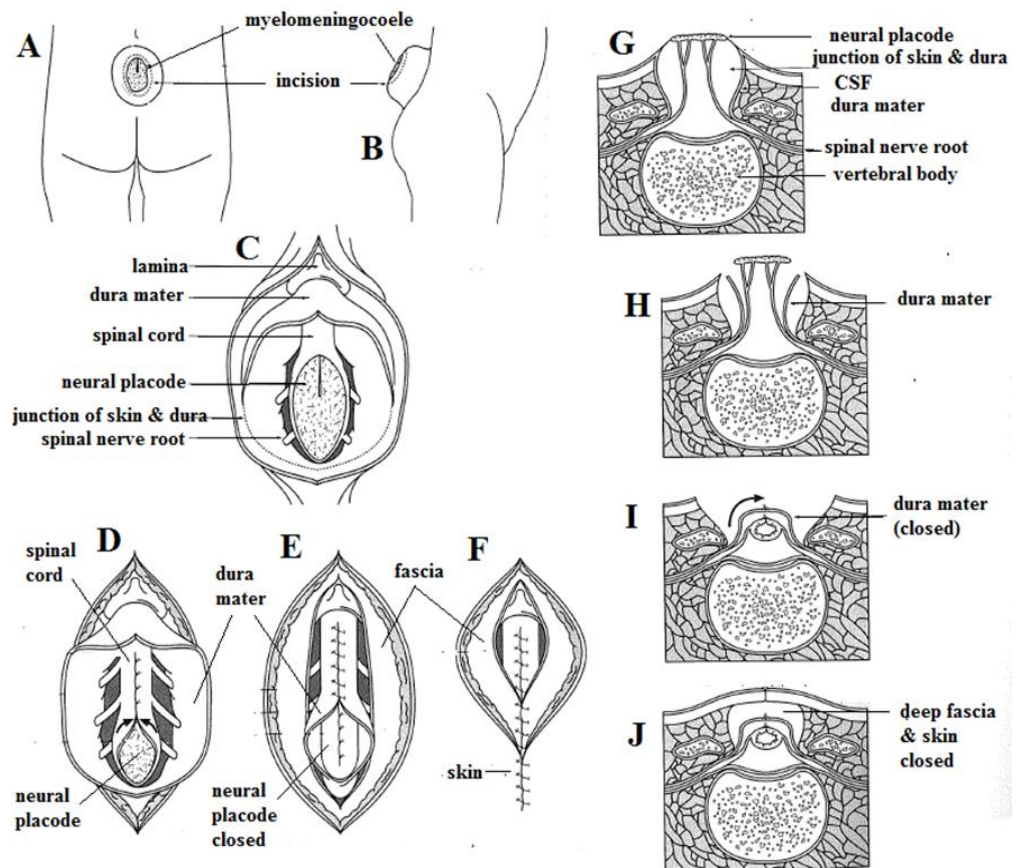


Fig. 33-15 MYELOMENINGOCELE REPAIR.

A,B,G, line of excision at edge of neural plaque (placode). C, free the elements of the defect to show the anatomy. D,H, fold over the placode to create a neural tube. E,I, close the dura. F,J, close the deep fascia and skin. G-J coronal views: G, before dissection.

After Rosenfeld JV, Watters DAK. *Neurosurgery in the Tropics*. Macmillan, 2000 p.386-8 Fig.11.8 with kind permission.

If there is a lumbo-sacral myelomeningocele, with reasonable power in the legs (normal sphincter control is unusual), operation may be justified. You will probably have to deal with variations of club foot deformity (32.10) later, as well as the development of hydrocephalus in many.

CLOSING A MYELOMENINGOCELE OR MENINGOCELE (GRADE 3.5)

INDICATIONS

A fairly small lumbo-sacral meningocele, with minimal neurological signs, no hydrocephalus, *and no other congenital abnormalities*.

CONTRAINDICATIONS

- (1) Complete or virtually complete denervation below the level of the lesion.
- (2) Progressive hydrocephalus.
- (3) A very large lesion, in which you will have difficulty closing the skin and subcutaneous tissues without tension.
- (4) Lesions in sites other than the lumbar or sacral region.
- (5) Untreated HIV disease.
- (6) A severely infected lesion: *drain the pus first!*

PREPARATION. Take the strictest aseptic precautions and *be careful not to damage the cord*. Lay the child prone with the head of the table depressed at least 30°, to minimize the loss of CSF when you incise the sac. Prepare and drape a wide operative field. Clean the sac meticulously with aqueous disinfectant.

OPERATION

If the swelling is covered by skin, make a longitudinal elliptical incision, through normal skin at the base of the swelling (33-15A,B). Cut through the subcutaneous tissue to the deep fascia, and define the neck of the sac by blunt dissection. The defect in the fascia will usually be quite small, and easy to expose. Free all surfaces of the sac, and open it over its dome. Send the CSF for culture if possible. If there are no nerve filaments, amputate the sac at its base, and close it with continuous 4/0 or 5/0 non-absorbable suture.

If you find nerve filaments or the meningocele is open, preserve the nerve filaments with the greatest care, and try to free them from the sac (33-15C). *Don't excise more healthy skin than you need*, but be sure to remove all the skin covering the placode in order to prevent the development of an epithelial remnant cyst later. If possible, after you have freed the neural plaque (placode), fold it over and close it along its axis (33-15D). Then close the dura over it with 4/0 or 5/0 prolene after freeing it on both sides of the plaque (33-15E).

Now free up the thick fibrous layer overlying the deformed spinal laminae on each side of the defect, and approximate these with the musculofascial layer (33-15F) so that you can obtain tension-free and solid skin cover.

Seal the wound meticulously, and nurse in the prone position. Apply a corset of orthopaedic strapping so that the abdomen is pulled upwards (33-16). This keeps tension off the suture line and allows faeces and urine to drain away from the wound. Leave a nasogastric tube *in situ* for 24h, and then start feeding. Keep the prone position for 10-14 days till you remove sutures. If there is a neurogenic bladder, make sure you train the mother to empty the bladder regularly by suprapubic pressure. You will then later have to teach her intermittent catheterization. *Keep faeces away from the wound!*

If the child also has hydrocephalus, a shunt (33.11) or ventriculostomy will be necessary, but wait at least 1wk after the back closure, till the risk of infection is diminished. Proceed when the back is healed, the ultrasound (38.2m) shows enlarging hydrocephalus, the child is well, and the CSF (from a ventricular tap) is clear.

The long-term management of neurologically impaired children with spina bifida is extensive, requiring bladder evaluation with possible clean intermittent catheterization, renal follow-up with ultrasound, bowel management, and occupational therapy. Find a centre where such care exists: the International Federation for Spina Bifida and Hydrocephalus can assist in the process.

SUSPENDING A BABY AFTER MYELOMENINGOCOELE REPAIR

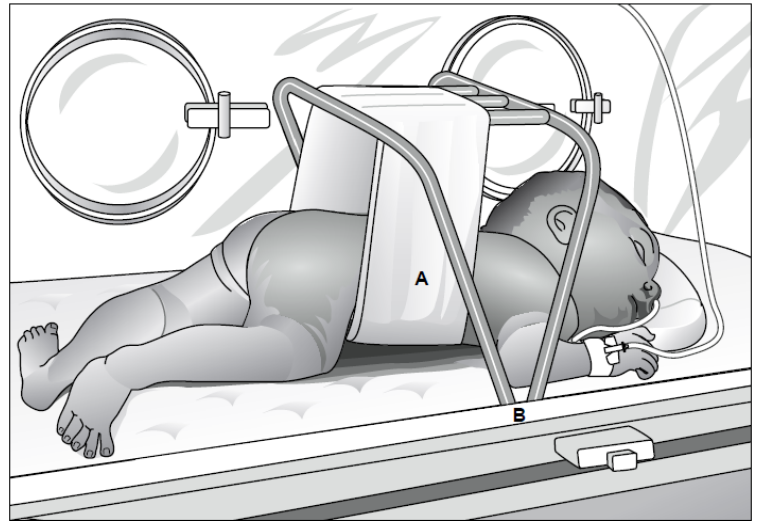


Fig. 33-16 POST-OP SUSPENSION OF A BABY AFTER MYELOMENINGOCOELE REPAIR.

A, orthopaedic strapping. B, special supporting frame. This keeps tension off the suture line; *don't dress the wound but make sure faeces and urine drain away from it.*

33.11 Hydrocephalus

When there is inadequate absorption of CSF in the head, its volume builds up in the ventricular system and a child's head consequently expands in size. When the CSF is unable to flow from the ventricular system to the subarachnoid space, it is described as a non-communicating hydrocephalus.

Congenital hydrocephalus usually arises from obstruction of the aqueduct of Sylvius, and in children with spina bifida from cerebellar and medullar herniation through the foramen magnum (the Chiari II malformation). Acquired hydrocephalus commonly arises from meningitis, but may result from intraventricular haemorrhage of prematurity, posterior fossa tumours or ventricular cysts.

(a) Symptoms in toddlers and children

Vomiting, drowsiness, irritability, fever, headache and loss of cognitive function or coordination are the commonest symptoms.

(b) Signs

The anterior fontanelle is bulging and tense, and scalp veins may be prominent; skull sutures separate and may become palpable, and the head may give a 'cracked pot' sound on percussion. In advanced cases, the child becomes lethargic and anorexic. When the 3rd ventricle expands, pressure on the oculomotor nerves causes down-turned ('setting sun') eyes. Cranial nerve palsies (especially VIth causing a squint, or Xth causing stridor) are not uncommon.

(c) **Ultrasound** (38.2m) will demonstrate dilated ventricles, as well as showing up intraventricular haemorrhage, cysts in the 3rd ventricle or a fourth ventricle tumour. You may be able to detect hydrocephalus antenatally. You can measure the thickness of the cerebral cortex: if this is <20mm, shunting will almost certainly be required, although the relationship of intelligence and cortical thickness is by no means proportional.

(d) Differential diagnosis

Distinguish hydrocephalus from **craniosynostosis**, where there is premature fusion of cranial suture lines and an odd-shaped enlarged head, but no bulging fontanelle.

ANATOMY OF THE VENTRICULAR SYSTEM

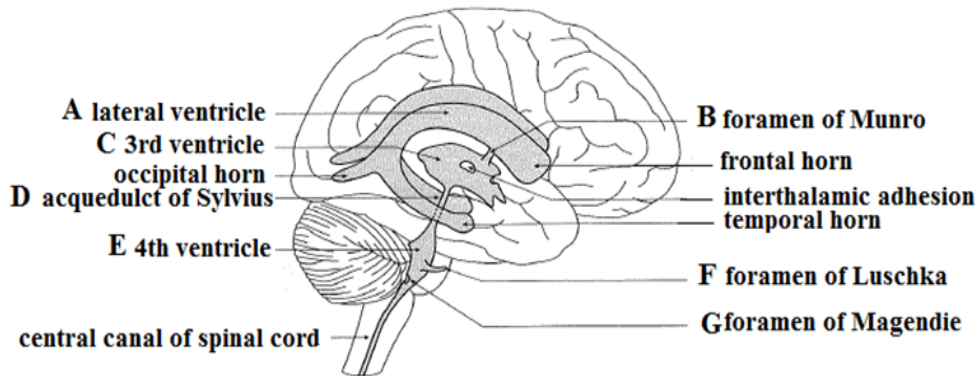


Fig. 33-17 ANATOMY OF THE VENTRICULAR SYSTEM.

About 20ml/h of CSF is actively secreted in the choroid plexuses in the lateral ventricle (A), the 3rd ventricle in the diencephalon between the 2 thalami (C). It leaves these by the aqueduct of Sylvius (D) to the 4th ventricle in the hindbrain (E), where more CSF is secreted. From here it passes through the 2 lateral foramina of Luschka (F) and the midline foramen of Magendie (G) to bathe the brain and spinal cord in the subarachnoid space. The total volume of CSF in an adult is c.130ml. After Rosenfeld JV, Watters DAK. *Neurosurgery in the Tropics*, Macmillan 2000 p.90 Fig 4.1 with kind permission.

You should measure the head against a standard head circumference chart:

OCCIPITAL CIRCUMFERENCE

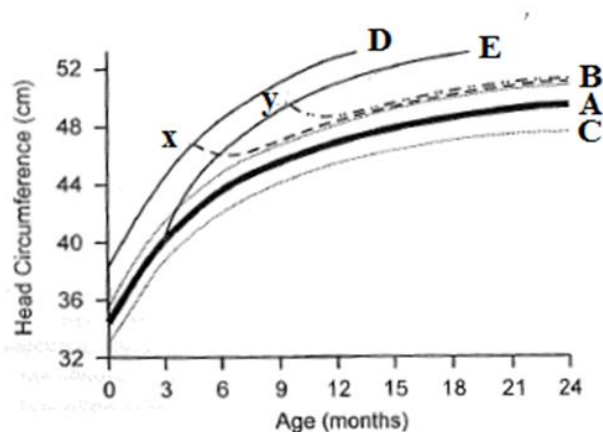


Fig. 33-18 HEAD CIRCUMFERENCE CHART, measured horizontally around the occiput just above the level of the eyebrows. A, 50th centile. B, 98th centile. C, 2nd centile. D, congenital hydrocephalus treated at point 'x'. E, hydrocephalus, acquired at age 3 months, treated at point 'y'.

After Rosenfeld JV & Watters DAK. *Neurosurgery in the Tropics*, Macmillan 2000 p.92 Fig 4.2 with kind permission.

(e) **Ventricular tap** is useful: shave and clean the head and disinfect the site carefully; aspirate through the anterior fontanelle with a 20G needle and syringe. Start at the level of the coronal suture just off the midline, aim slightly away from the midline, advance while aspirating until CSF is found. If the pressure is high, aspirate until the fontanelle is flat. *Don't try to do this slowly: it is unnecessary and you will anyway fail!* Send the fluid for CSF analysis. You can measure the CSF pressure by attaching the needle to an infusion line and measuring the level above a zero point at the level of the head.

(f) Management

The commonest treatment for hydrocephalus is to insert a shunt to drain the CSF either into the peritoneal cavity or rarely into the atrium. The ventriculo-peritoneal (VP) shunt is simpler to insert, but will probably not be effective if there has been peritonitis previously, because of adhesions.

Various types of shunt exist, with different valve mechanisms, but it is not necessary to use commercially produced shunts. An affordable shunt is the Chhabra shunt from India

(provided free to qualified centres by the International Federation for Spina Bifida and Hydrocephalus).

Don't delay shunt insertion in hydrocephalus, except until intraventricular haemorrhage or meningitis has recovered and the CSF is clear (<10 cells/ml) with a low protein content.

Don't attempt to treat a child with a head circumference >60cm if there is gross neurological deficit.

VENTRICULO-PERITONEAL SHUNT FOR HYDROCEPHALUS (GRADE 3.4)

PREPARATION

Shave the scalp and clean it thoroughly with alcohol and betadine before operation. Administer prophylactic cefalosporin, or gentamicin and ampicillin.

Position the head turned laterally on a head-ring, with the table elevated 10° and a support under the shoulders to keep the neck extended so that there is a straight line from head to abdomen along the anterior chest wall in front of the clavicle (33-19B).

INCISION

Make a semicircular flap 3cm above the centre of the pinna and 4cm behind its top edge, in the occipito-parietal area (33-19A). *Don't make this incision too low* where you may hit the transverse venous sinus.

Make a burr hole (or if the bone is very thin, nibble it away with forceps or scalpel) but don't open the dura; before you do so, make an opening in the peritoneum through a small transverse right hypochondrial incision and make sure you are actually inside the peritoneal cavity by attaching haemostats to the edges of the peritoneal layer.

VENTRICULO-PERITONEAL SHUNTING

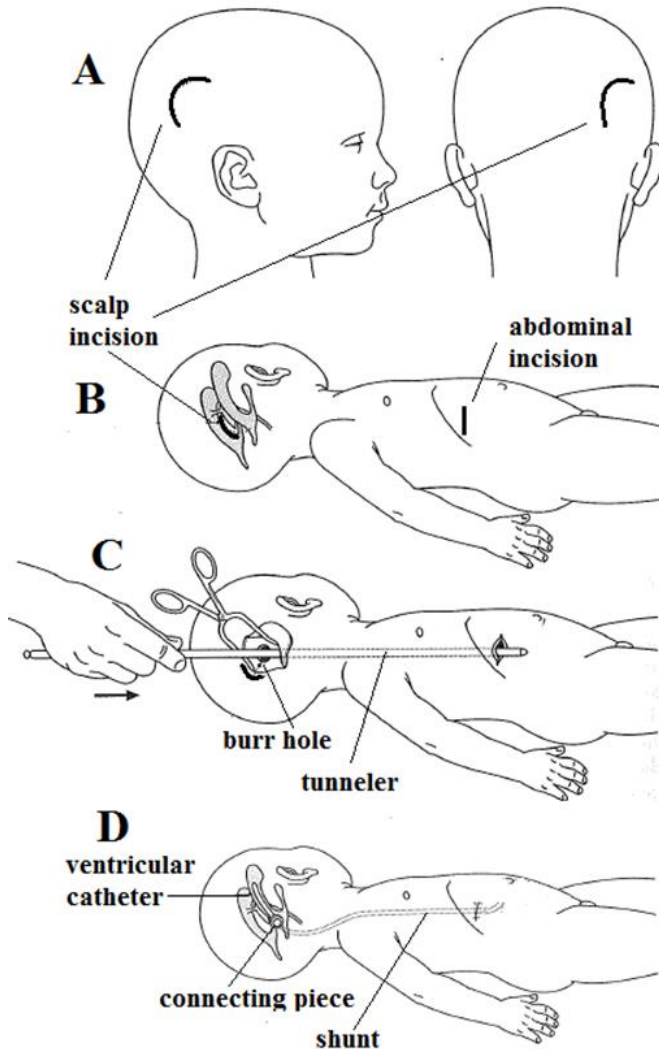


Fig. 33-19 PLACEMENT OF A VENTRICULO-PERITONEAL SHUNT.

A, scalp incision. B, operative position, showing orientation of ventricles and site of right upper quadrant abdominal incision. C, passing the tunneler. D, final placement of shunt.

After Rosenfeld JV & Watters DAK. *Neurosurgery in the Tropics*, Macmillan 2000 p.380 Fig 11.6 with kind permission.

Pass a tunneler subcutaneously between the 2 wounds (33-19C), either from above to below or *vice versa*. If you don't have a tunneler long enough, you may need to make an extra incision in the neck.

Attach the distal shunt tubing to the tunneler and pass it under the skin from neck to abdomen, but leave it outside the peritoneal cavity. Check its patency with saline. When it is correctly in place, remove the tunneler and fix the shunt tubing to the valve or connecting L-piece.

Then make a small cruciate opening in the dura just big enough to pass the shunt through. (An incision too big will allow CSF to leak.) With the proximal shunt mounted on a stilette, guide it forwards towards the inner canthus (corner) of the opposite eye (felt through the drapes). As soon as the shunt has entered the ventricle, you will see CSF emerge; remove the stilette: you should see a substantial flow of clear CSF. Send this for culture, if possible.

Then, check with ultrasound, if you can, that the shunt is well placed in the anterior horn of the lateral ventricle, withdraw it 2cm and attach it to the valve, or a connecting L-piece, trying not to handle the shunt except with instruments.

Fix the valve, or L-piece, with non-absorbable sutures to the skull periosteum. Make sure that CSF is dripping regularly from the distal end of the shunt, and then pass this into the peritoneal cavity either over the liver or deep into the pelvis.

Fix it to the peritoneum with an absorbable suture and close the abdomen taking care not to include the shunt tubing in your stitch or kink its position. Close the scalp flap over the tubing so that sutures lie well away from it.

DIFFICULTIES WITH VENTRICULO-PERITONEAL SHUNTING

Advise the parents to return the child in case of any serious symptoms: *late presentation of complications is the commonest cause of death and long-term disability in these children!* Complications are common, and usually occur in >30% of cases! You must warn parents that you may have to replace the shunt several times, and particularly as he grows.

If the shunt blocks, it may do so at the ventricular end (where the choroid plexus adheres to the tubing) or the peritoneal end (where the omentum or adhesions may occlude its end). Symptoms and signs depend on the rate and degree of the blockage, but essentially are worsening of the original hydrocephalus problems, especially vomiting, headache and loss of neurological function.

To treat the blockage, you need to explore the shunt, disconnect it and test the flow through it at the peritoneal and ventricular ends. You may be able to unblock the ventricular catheter by introducing a stilette down it, and passing a diathermy current along it. If it remains blocked, and remains stuck, it is best to place another shunt next to it rather than forcibly removing it.

If the shunt disconnects or migrates (which may be visible on simple radiographs), especially during a growth spurt, the peritoneal end may end up disconnected inside the abdomen, or under the skin. Reconnection with new tubing will solve the problem.

If the shunt becomes infected, either *de novo* or more commonly within a few months as a result of sepsis spreading from elsewhere (e.g. teeth, throat etc.), the child deteriorates with headache, fever, vomiting and maybe fits.

Try antibiotics alone, but if this fails, disconnect the shunt tubing and drain the CSF externally into a closed drainage system (an external shunt). When the CSF is clear and no longer infected, re-insert the distal end of the shunt. However, it is frequently necessary to remove the shunt entirely and replace it with a new one.

If the skin over the shunt necroses, it has probably been closed under tension. The skin needs debridement, and usually you will have to reposition the shunt on the opposite side of the head.

If the child develops peritonitis, the distal peritoneal end of the shunt needs to be removed from the abdomen, and allowed to drain into an external closed drainage system till the inflammation has settled down.

If there was only localized peritonitis (e.g. from appendicitis), the abdomen can probably still be used for drainage, but if the peritoneal inflammation was generalized, it may no longer absorb properly. In this case, a ventriculo-atrial shunt may be necessary.

If the abdomen distends with fluid, the CSF may not be absorbed properly as above. Alternatively the fluid may collect into a loculated 'pseudocyst' which is visible on ultrasound (38.21). In this case, perform a laparotomy to break down the cyst walls and reposition the shunt if it remains patent.

N.B. More sophisticated surgery for hydrocephalus consists of endoscopic 3rd ventriculostomy, which has much fewer complications and is effective in the majority of cases. This procedure is not that difficult to grasp and has been effectively performed up-country in Mbale, Uganda. You need a flexible paediatric endoscope like a cystoscope, and to be shown how to do the procedure by an expert.

33.12 Congenital vascular lesions

Congenital vascular lesions are not uncommon, and may worry a parent, so you should know how to advise. Some will regress, some will grow as the child grows, and some increase in size alarmingly. Try to identify them correctly to advise the right treatment. Differentiate between angiomas (which are tumours) and vascular malformations (which are not). You may be able to diagnose cystic lesions prenatally with ultrasound.

A **capillary haemangioma** is, characteristically, a bright red, raised lesion varying from mms to cms in diameter. It is well circumscribed and only partially compressible.

A **cavernous haemangioma** is nodular and may be very large in diameter and depth. It is often bluish or purple and easily compressible. Mixed variants exist; they are histologically benign neoplasms of proliferative endothelium.

A **capillary cavernous haemangioma** consists of abnormal capillaries, arteries, and veins, and is partly compressible. It is usually present at birth, and commonly occurs on a child's face, axillae, or neck, where it may extend into the mediastinum. It may occasionally resolve spontaneously over several years (unusual), or it may enlarge rapidly.

Occasionally it connects with arteries and pulsates, expanding progressively: the feeding vessels then need ligation. Excision is indicated if there is functional disability (e.g. amblyopia from an eyelid haemangioma, 28.9), gross disfigurement or significant haemorrhage, or skin necrosis overlying the lesion.

CONGENITAL VASCULAR MALFORMATIONS

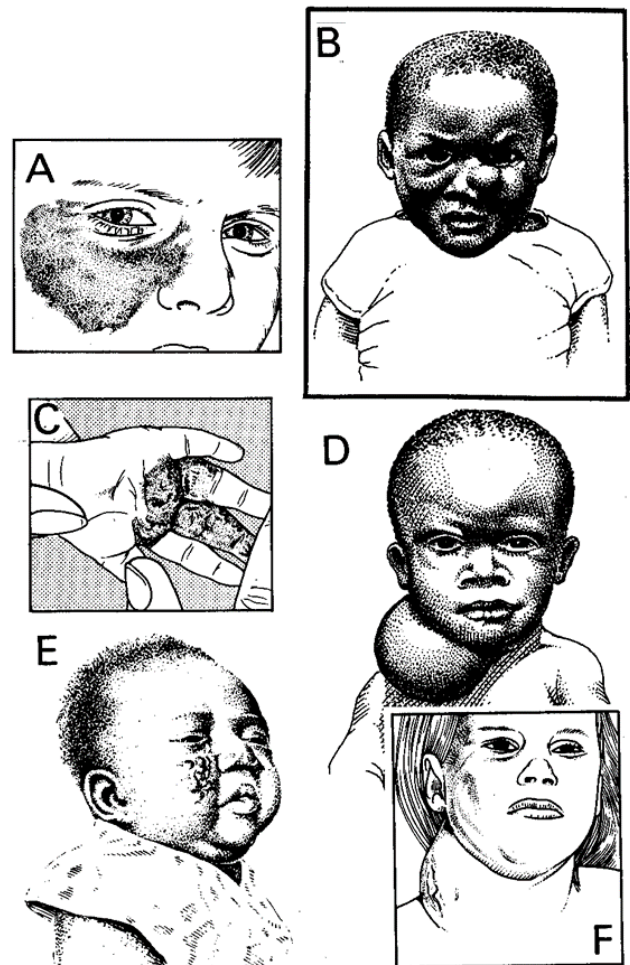


Fig. 33-20 CONGENITAL VASCULAR ANOMALIES.

A, a port-wine stain. B, a cavernous haemangioma. C, a capillary cavernous haemangioma of the hand. E, a capillary haemangioma of the face. D,F, cystic hygromas.

A,D,E, after Bowsman C. *Surgery and Clinical Pathology in the Tropics*, Livingstone 1960, permission requested.

The so-called '**strawberry naevus**' (33-20E) is a haemangioma skin lesion, and is commonest on the face. It is bright red, and a little raised, sometimes quite markedly. It may occur on the mucous membranes and may bleed severely. The lesions appear 1-4wks after birth, and enlarge for a few months.

Rapid growth is unusual after 6 months, but slow growth is common up to 1yr. *If you leave them alone, 95% will resolve spontaneously.* The first sign that this is happening is the appearance of a lighter, flatter area. Resolution may be precipitated by trauma, but this usually causes only minor scarring.

Advise the mother that the lesion will probably disappear slowly. Warn her not to allow traditional healers to scarify the lesion, which may cause bleeding, infection, and worse scarring. If resolution is slow, and parents pressurize you, refer the child to a plastic surgeon specialist because the final results of excision may otherwise be very disfiguring.

Lesions on the face, in the area of distribution of the ophthalmic and maxillary branches of the Vth nerve, may be associated with vascular abnormalities of the cerebral cortex (Sturge-Weber syndrome), and present with seizures. Glaucoma of the ipsilateral eye sometimes occurs, and is treatable (28.6).

The so-called ‘**port-wine stain**’ or ‘**flame naevus**’ (33-20A) is a malformation of cavernous channels, and usually occurs on the face or neck, but is not uncommon on the trunk. It is usually present at birth and does not progress, but it may be quite extensive. The texture of the skin is normal, and is not usually thickened; occasionally there is some hypertrophy and irregularity. If a lesion is particularly prominent, as in light skin, cosmetic creams may help. Otherwise, there is no treatment. Reassure the parent that the lesion will not enlarge.

The so-called **lymphangioma** is actually a malformation of cystic cavities filled with clear or straw-coloured fluid (actually lymph) which grow slowly, often infiltrating or surrounding adjacent structures. Occasionally it regresses, but it may grow alarmingly especially when infected.

It occurs usually in the neck and axilla, but may also be in the mediastinum, retroperitoneum, or the groin. It usually appears early, and may be present at birth. The mass is not attached to skin and is fluctuant. It may be very large, being known as a **cystic hygroma** (33-20D,F), where it may cause respiratory distress due to pressure effects on the airway.

Review the child carefully, and at each visit use a measuring tape to record the exact size of the lesion in 2 dimensions at right angles. At each visit record changes in appearance and complications, and if possible take photographs, measuring the size of the lesion.

Complications are ulceration, infection and bleeding, and rarely a consumptive coagulopathy and thrombocytopenia (Kasabach-Merritt syndrome).

You can try to cause thrombosis and regression by injection of 50% dextrose into the lesion. Simple aspiration will reduce the size of a lymphangioma temporarily, but with the risk of introducing infection.

Surgery is likely to be difficult though, and bleeding severe. In the neck, post-operative haemorrhage may cause acute neck swelling and respiratory compromise.

Differential diagnosis

Suggesting cystic degeneration in a lymph node: HIV disease, multiple asymmetric lesions in typical sites of lymphadenopathy.

Suggesting a branchial cyst: unilocular and low in the neck, along the anterior border of the sternomastoid; aspiration yielding a thicker opalescent fluid, instead of the thin, clear, watery fluid from a hygroma. If you are a careful operator, and have diathermy available, try excising this.

Suggesting a haematoma: associated with torticollis from neck injury at birth.

33.13 Other paediatric problems

If a male neonate, infant or occasionally older child has retention of urine with overflow, the neck of the bladder is probably obstructed by URETHRAL VALVES. These usually present in the 1st 6 months of life. They may present as retention, urinary tract infection, dribbling incontinence or renal failure. Sometimes the urinary symptoms are overlooked and the child presents with vomiting, failure to thrive, uraemia and acidosis. The urine may leak from the kidneys into the peritoneum (urinary ascites). You may feel that the bladder is distended, and you may be able to feel the kidneys. Ultrasound will demonstrate a distended bladder (38.2i) and perhaps also hydro-ureters and hydronephroses (38.2f). The distended bladder will disappear on catheterization. Pass a urethral catheter (a Ch6-8 feeding tube is suitable) under ketamine. Treat any infection. If endoscopic resection of the valves is not available, you can try to pass a Fogarty arterial balloon catheter in an attempt to disrupt the valves, though try to arrange endoscopy which is better. To temporize the problem, you may need to drain the bladder with a Foley catheter or suprapubic cystostomy long-term.

If a neonate has a fleshy swelling on the lower anterior abdomen weeping urine, this is a BLADDER EXSTROPHY (ectopia vesicae). In the male, there is complete epispadias, *i.e.* the urethra opening on the dorsal (exterior) side of the penis. The scrotum is wide often with maldescended testes and inguinal herniae. The anus is anteriorly placed and there is often sphincter laxity with rectal prolapse. Occasionally there is a cloacal exstrophy which includes a rectal agenesis.

Bladder reconstruction for exstrophy is a complex procedure, with results especially poor in older children. Surgery may include pelvic osteotomy and require several stages; this needs an expert! If the final bladder capacity is likely to be small, urinary diversion may be the best long-term option.

If a neonate has ambiguous genitalia, examine the baby carefully with good light in a warm room. If there is a small phallus, and a vagina or bifid scrotum, there is doubt as to the gender of the baby. Two conditions are most common:

(1) *severe hypospadias with undescended testes*, where the phallus is normal with a urethral opening in the perineum; there is no vagina or uterus.

(2) *congenital adrenal hyperplasia (CAH)*, where there is a small phallus with normal vagina and uterus: this is caused by 21-hydroxylase deficiency leading to virilization. It has an autosomal recessive inheritance and so may occur more than once in the same family, and more commonly where there is consanguinity.

This also leads to low cortisol levels: glucose, IV fluids and hydrocortisone are needed urgently!

N.B. Females with CAH have a normal reproductive potential.

A specific diagnosis in intersex states may be difficult without sophisticated karyotyping. You must be frank with parents, however, especially with regard to fertility and the sex of rearing. *You should never refer to a child as an 'in-between'*; dispel fears that malformations might lead to homosexuality.

You should aim to limit psychological disturbance in the child: before suggesting a child is a male, think of the viability of rearing him as such (especially if there is a micro-phallus, bifid scrotum or vagina) especially in terms of micturating standing up, possible sexual intercourse and social embarrassment.

Surgically, it is easier to construct female appearances than male, though this may of course not be the most important criterion. All this needs an expert. Whilst some traditions recognize a 'third gender', the choice of how to bring up a child with ambiguous gender remains complex.

If a neonate is born with a large solid mass below the sacrum, it is probably a SACROCOCCYGEAL TERATOMA. This is a solid mass protruding usually from the coccyx, and displacing the anus anteriorly. It may extend into the pelvis, and its malignant potential increases rapidly with age (>30% at 1 month) and with internal extension. There may be rudimentary limbs growing from the teratoma, and it may contain well-differentiated foreign tissues, especially teeth but also including brain! Elevated levels of α -fetoprotein strongly suggest malignant change.

If possible, preserve a blood sample for an initial α -fetoprotein baseline level. Ultrasound will demonstrate pelvic or intra-abdominal extension, and distinguish from a sacral myelomeningocele (33.10) which is higher up. Pressure on a teratoma will cause no bulging of the anterior fontanelle.

If the tumour is intact and the baby stable, there is no need for immediate operation. You will reduce the risk of infection by operating within 24h of birth because the bowel is not yet colonized, but surgery will be more technically difficult.

Don't wait long though, because a tumour, benign at birth, is often malignant by the age of 2 months.

EXCISION OF SACROCOCCYGEAL TERATOMA (GRADE 3.5)

Place the baby prone with legs abducted and the lower abdomen raised on a small sandbag (33-21).

Pack the rectum with paraffin gauze to ease its identification in the operation. Make sure you cross-match blood.

REMOVAL OF SACROCOCCYGEAL TERATOMA

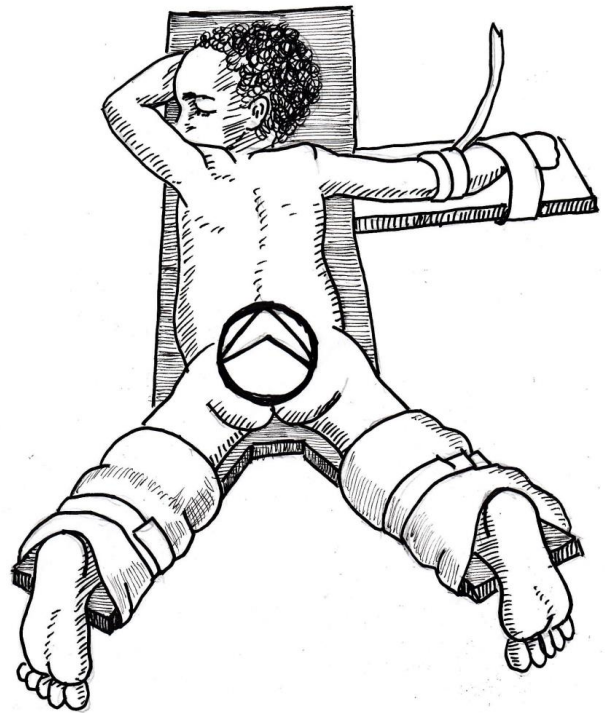


Fig. 33-21 OPERATION FOR SACROCOCCYGEAL TERATOMA. Showing the upper chevron incision, a 2cm-long apical extension, and a lower chevron incision.

After Lister J, Irving IM. Neonatal Surgery, Butterworth 3rd ed 1990 p.148 Fig 14.11.

Make a chevron (inverted V) incision at the upper limit of the teratoma, and extend it cranially at the apex for 2cm. Divide between the 4th and 5th segments of the sacrum, removing tumour with the coccyx and 5th sacral segment. Ligate and divide the middle sacral vessels, taking care to avoid heavy bleeding, and then dissect the tumour from the lateral skin flaps. Good clearance is necessary, and this is difficult when the tumour is wrapped closely round the ventral aspect of the rectum, and when the tumour cannot be readily distinguished from ischio-rectal fat.

Mobilize the tumour completely, and then incise the skin at its lower border with another chevron, some distance above the anus, and so remove the teratoma. Repair the *levator ani* (26-1) behind the rectum and hitch it up to the presacral fascia. Close the skin in an inverted Y shape with 4/0 nylon, with a suction drain *in situ*, and leave it usually in place for 48h.

Nurse the baby prone post-operatively (as for myelomeningocele, 33-16) till you remove sutures after 10-14 days. Do 6-monthly follow-up rectal examinations to detect presacral recurrences, as well as serial α -fetoprotein levels, if possible.

If a neonate (or older child) has respiratory difficulties and a radiograph shows bowel or stomach in the chest, this is due to a DIAPHRAGMATIC HERNIA. This may be congenital or, rarely, traumatic.

Presentation is usually later in childhood with respiratory distress on exercise. Diagnosis is evident on chest radiography and ultrasound. *Avoid ventilating the child by a mask,* because air inevitably introduced into the stomach will make thoracic compression of the lungs worse.

Repair the defect in the diaphragm through the abdomen with non-absorbable mattress sutures, *taking care not to damage branches of the phrenic nerve.* Pulmonary hypoplasia or pneumothorax are hazards and often demand ventilatory support. Older children with this condition do better, as their lungs are better developed.

If two neonates are conjoined ('Siamese twins'), this is a very rare instance. The join may be trivial or involve vital structures like heart, lungs, brain etc. The publicity gained by such cases may be advantageous to bring support to your hospital! Prompt investigation and treatment is mandatory before you can decide what sort of surgery is feasible.

If a baby is born with extra limbs, these may be surprisingly easy to remove. However, get some radiographs done and try to get advice. If there is a good chance of improving function, and at the same time making the baby look normal, encourage the parent.

CAUTION! This does not apply to fingers and toes: don't be tempted to remove extra digits until a child is older (32.21).

N.B. Other childhood problems are discussed elsewhere:

amblyopia and squint (28.9),
ascaris infestation (12.5),
 bladder stones (27.17),
 Burkitt's lymphoma (17.6),
 cancer pain control (37.2),
 cancrum oris (31.5),
 circumcision (27.29),
 cleft lip (31.7),
 club foot (32.10),
 congenital glaucoma (28.6),
 contractures (32.1),
 corneal scarring (28.4),
 corrosive oesophagitis (30.3),
 deciduous teeth (31.1),
 extra digits (32.21),
 eye tests (28.1),
 foreign bodies in the ear (29.6),
 foreign bodies in the nose (29.11),
 foreign bodies in the throat (30.1),
 haematocolpos (23.17),
 hearing tests (29.2),
 hypertrophic pyloric stenosis (13.6),
 inguinal hernia and hydrocoele (18.5),
 intussusception (12.7),
 maldescended testis (27.27),
 myopia (28.8),
 nephroblastoma (27.35),
 noma (31.5),
 osteomyelitis (7.3),
 otitis media (29.4),
 painful hip and limp (32.14),
 pelvi-ureteric junction obstruction (27.14),
 phimosis and paraphimosis (27.30),
 physiological goitre (25.4),
 poliomyelitis (32.7),
 proptosis (28.11),
 pyomyositis (7.1),
 rectal prolapse (26.8),
 rectovaginal fistula (23.17),
 retinoblastoma (28.16),
 spinal TB (32.4),
 testicular torsion (27.25),
 tongue tie (31.9),
 tracheostomy (29.15),
 trachoma (28.13),
 umbilical hernia (18.10),
 urethral stones (27.18).