

INTRODUCTION

Paediatric surgery is not a special branch of surgery, it is the whole of surgery applied to a child. Children should not be regarded as small adults, they are vastly different with different diseases, organ functions and responses to trauma and infection. Given a decent chance, they will outlive their operators.

The whole concept of paediatric surgery can be summarised in a nutshell.

"If this infant (on whom you are about to perform an operation) could speak it would beg imploringly of the surgeon: "Please exercise the greatest gentleness with my immature tissues and try to correct the deformity at the first operation. Give me blood and the proper amount of fluids and electrolytes, add plenty of oxygen to the anaesthesia, and I will show you that I can tolerate a terrific amount of surgery. You will be surprised at the speed of my recovery and I shall always be grateful to you."

Willis Potts The Surgeon and the Child.

The major causes of death in childhood are surgical problems

- Accidents
- Congenital malformations
- Malignancy
- Infections

Any person dealing with a sick child should be able to answer the following questions to the satisfaction of both child and family.

- What is the incidence and aetiology of the disease process.
- What are the presenting features as far as sex, age, typical history

and physical findings are concerned.

- What is the differential diagnosis
- What are the physiological derangements and complications
- What is the objective of the operative procedure
- What is the best time for surgery.
- What are the chances of normal growth and development.

The small baby as a surgical patient

The outstanding characteristics of the infant and young child are the rapidity with which life processes proceed and physiological status changes, e.g. Four hours in a neonate is equivalent to 8 hours in an infant, twelve hours in a toddler and 24 hours in an adult.

The newborn has great assets

- Tremendous vitality
- Abundance of maternal antibodies.
- Adaptation to extra-uterine life.
- Endocrine systems geared for stress.
- Freedom from acquired or degenerative diseases.
- Rapid recovery after surgery.

Liabilities of neonates

- Hypo: thermia, volaemia, glycaemia,
- calcaemia and prothombinaemia.
- Rapid responses to stress
- Rapid turnover of water and electrolytes
- Other major congenital abnormalities
- Susceptibility to infection
- Hyperbilirubinaemia

Neonatal surgery

Nearly all surgery in the first week of life is directed at correcting congenital abnormalities. Most numerous are congenital abnormalities of the gastro-intestinal tract, followed by congenital abnormalities of the respiratory tract.

Others of lesser incidence but equally important are those of the central nervous system and urinary tract.

It is important to recognise neonatal surgical emergencies early before clinical deterioration. These findings are the so-called

'RED FLAGS':

- Maternal polyhydramnios
- Prematurity
- Rapid respiration more than 40/min.
- Difficult respiration
- Cyanosis
- Excessive salivation
- Abdominal distension
- Abdominal mass
- Bile-stained vomiting
- Failure to pass meconium in first 24 hrs of life.
- Inability to void or poor urinary stream.
- Convulsions
- Lethargy
- Jaundice within the first day of life

The principles of management in neonatal surgery are

- Surgical recognition.
- The baby should be brought to the best physical state possible prior to surgery
- A correct surgical procedure should be done
- The baby should receive complete pre – intra – and postoperative care.

Main causes of death in neonates

- co-existing congenital abnormalities
- prematurity
- chromosomal abnormalities
- septicaemia

CONGENITAL DIAPHRAGMATIC HERNIA

The incidence is about 1:1000 - 1:3000 births. An arrest in diaphragmatic development during the

8 - 10th week of pregnancy causes incomplete separation of the pleura from the abdominal cavity by the diaphragm. There is a defect in the diaphragm usually in its left postero lateral aspect. During intra uterine life, abdominal viscera migrate through the opening filling the pleural cavity with stomach, small intestine, colon and spleen, thus inhibiting normal lung development.

Clinical Presentation

Progressive respiratory distress and an apparent dextrocardia. Bowel sounds may be heard in the chest and the abdomen may be scaphoid. The diagnosis is confirmed on chest x-ray noting bowel loops in the chest and mediastinal shift to the opposite side. Survival is related to the age of onset of symptoms, cardiac abnormalities, the degree of lung hypoplasia and prematurity.

Differential diagnosis

- (1) Tension pneumothorax, which can also deviate the mediastinum.
- (2) Cystic adenomatoid malformation of the lung which can give the mistaken impression of air-filled loops of bowel above the diaphragm.
- (3) Eventration of the diaphragm.
- (4) Sequestration of the lung

Management

Management consists of a 'trial of life'.

The babies are maximally supported with IV fluids, ventilation, correction of all biochemical and hematological abnormalities. Only when fully stabilized is the hernia repaired. The mortality is >50%.

OESOPHAGEAL ATRESIA AND TRACHEA OESOPHAGEAL FISTULA

The condition represents a form of generalised disturbance in embryogenesis. The VACTERL

association (vertebral, anorectal, cardiac, trachea-oesophageal fistula, renal abnormalities and limb dysplasia is frequently seen in association with oesophageal atresia).

Clinical presentation

The incidence is 1/3000 live births

Polyhydramnios and prematurity are frequently seen. The newborn infant salivates excessively and on feeding may choke, cough or regurgitate the first feed. Cyanosis with feeds and apnoea can occur. Suspicion based on any of these signs or symptoms calls for immediate evaluation. The simplest and most effective way of excluding the diagnosis of oesophageal atresia is to pass a tube through the mouth or nose to the stomach. The tube should be relatively rigid and it usually meets an obstruction between 9 and 13 cm from the nares confirming the diagnosis of oesophageal atresia.

A plain x-ray of the chest and may abdomen would show air in the intestine which confirms a distal trachea-oesophageal fistula ($\pm 90\%$). A contrast study is unnecessary and potentially dangerous investigations. If on plain x-ray – there is no air in the intestine then a diagnosis of atresia without _____ is made (10%).

Treatment

The patient should be transported with the head up at approximately 30° to prevent gastric acid regurgitation into the bronchial tree. The saliva in the upper pouch should be aspirated via nasogastric tube. At no stage should the baby be fed, as this will result in aspiration pneumonia.

Management entails resuscitation and treatment of any pneumonia that may be present. A careful clinical assessment is made looking for other congenital anomalies, which are present in up to 40% of infants, especially cardiac and gastro-intestinal tract abnormalities. A right-sided aorta

is excluded on the x-ray and with echocardiography.

Surgery

The atresia is approached through a third or fourth intercostal space extrapleural thoractomy. The fistula in the distal segment is isolated, divided and sutured closed. The upper blind pouch is mobilised and an end-to-end anastomosis with the distal oesophagus is effected. In the baby with two blind ends, i.e. where no fistula is present, there is usually a large gap between the two blind ends which often precludes direct end-to-end anastomosis. Under these circumstances a feeding gastrostomy is required and the oesophageal anastomosis is delayed. If the blind ends are too far apart for oesophageal anastomosis, a functioning gullet is effected by bowel interposition grafting or gastric advancement. Survival is excellent and mortality is related to prematurity and cardiac abnormalities.

NEONATAL INTESTINAL OBSTRUCTION

Neonatal intestinal obstruction occurs in approximately 1/2000 live births and may result from:

- intrinsic developmental defects arising from disordered embryogenesis.
- abnormalities of peristalsis and/or intestinal contents.
- a secondary insult acquired *in utero*, after normal intestinal development.

It is now very uncommon for infants to succumb from intestinal obstruction *per se*; mortality is related to extreme low birth weight and associated severe congenital anomalies. Consequences of delay in recognition of neonatal bowel obstruction may be dire, however. Inhalation of vomit, sepsis, midgut infarction and enterocolitis are potentially lethal, but can be avoided by prompt and appropriate management.

Anomalies of rotation and fixation with volvulus

The incidence of malrotation is about 1/600 live births. Most errors occur during the second phase of rotation. The 1st and 2nd parts of the duodenum are situated normally, but the third and 4th parts descend vertically along the right side and the small bowel lies predominantly to the right of the mid-line. The caecum is central and the colon, doubled on itself, is on the left side of the abdomen.

Failure of fixation often occurs. Although thickened peritoneal attachments (Ladd's bands) may pass from the caecum across the duodenum to the right upper quadrant posterior abdominal wall, these seldom cause obstruction *per se*. The mid-gut segment is predisposed to volvulus because, as a result of the failure of fixation, the entire mid-gut is suspended in the abdominal cavity by a narrow pedicle (superior mesenteric artery, vein and lymphatics)

Diagnosis

Volvulus almost always occurs in a clockwise direction, symptoms usually resulting from episodes of varying degrees of volvulus. About 50% of cases present during the neonatal period, typically as well infants who have fed and stoolled normally but subsequently develop colic and bile-stained vomiting. Abdominal signs other than epigastric distension are absent initially, but become obvious once intestinal strangulation has occurred. The only evidence may be a history of bile-stained vomiting, which is often intermittent. Delay in recognition may lead to mortality or significant morbidity, with loss of much of the mid-gut as a result of strangulation and necrosis. Plain abdominal radiographs may show evidence of high small bowel obstruction with a dilated duodenum and stomach, but may be normal. A

barium meal excludes or confirms the diagnosis. Contrast enema may be misleading, because the caecum is often 'high' in neonates and may not be taken up in the volvulus. Dilated loops of small bowel are usually indicative of bowel infarction.

Management

At surgery, the bowel is untwisted anticlockwise, Ladd's bands and interloop adhesions are divided, and the distal duodenum is mobilised to the right, and the caecum to the left thus broadening the base of the mesentery. Patency of the lumen is checked. The bowel is then replaced in the abdomen in a non-rotated position. It is unnecessary to fix the bowel. The appendix may be removed.

INTESTINAL ATRESIA AND STENOSIS

The term 'atresia' denotes complete intrinsic occlusion of the intestinal lumen as a result of anomalous development of the intestinal walls. The term 'stenosis' refers to narrowing of the lumen, which causes incomplete obstruction. Approximately 30% of infants presenting with neonatal intestinal obstruction have atresia or stenosis.

Atretic lesions are distributed as follows:

- duodenum - 45%
- jejunum - 25%
- ileum - 15%
- colon - 5%
- multiple - 10%

Duodenal atresia

The lumen of the duodenum passes through a solid phase between the 5th and 8th weeks of embryonic life in about 30% of fetuses. Failure of recanalization could, theoretically, cause duodenal atresias. The frequent coexistence of other anomalies, however, suggests that an

insult affects the embryo as a whole early in embryogenesis.

Most duodenal atresias occur in the mid anatomical 2nd part of the duodenum. Several types are recognised.

- Proximal and distal segments may end blindly and adjacent, or separated by a gap or fibrous cord which is usually 'filled-in' with pancreatic tissue.
- A diaphragm, often with a small perforation, is not uncommon; multiple diaphragms seldom occur. A thin diaphragm, which has ballooned distally, is known as a 'windsock'.
- The distal biliary tree is often abnormal; it usually opens at the level of the diaphragm, via a T-shaped or even a Y-shaped insertion proximal and/or distal to the atresia. Pancreatic tissue overlying or surrounding the duodenum at this level is regarded as an indication of associated failure of duodenal development, rather than true annular pancreas.

Atresia of the more distal bowel occurs usually as a result of infarction of the bowel later in foetal life. In about 30% of patients, there is evidence at surgery of a 'foetal accident' (volvulus, intussusception, snaring at the umbilical ring, torsion around a congenital band, and, with meconium ileus, intra-uterine perforation or localised volvulus proximal to the obstructed segment). Interruption of the blood supply may produce all the varieties of bowel atresia found in infants.

Stenosis - there is a localised narrowing of the bowel or a diaphragm (membrane) with a small perforation.

Atresia type 1 is characterised by a thin complete mucosal diaphragm. The bowel is normal in length.

Atresia type 2 is characterised by blind ends joined by a fibrous band. There is seldom a corresponding defect in the mesentery; when this occurs, the length of bowel may be subnormal.

Atresia type 3 is characterised by disconnected blind ends, with a gap in an otherwise normal mesentery, and often considerably shortened small bowel. Type 3 is divided into 2 subtypes:

- In type **3a**, the ends are blind and there is a single gap. Loss of tissue from both bowel and mesentery occurs, and the length of the small bowel is subnormal.
- Type **3b** is usually characterised by upper jejunal atresia with extensive loss of bowel length, associated with a gross mesenteric defect. Variable lengths of distal ileum remain viable, supplied by a single abnormal artery arising in continuity from the right and/or middle colic vessels. Type 3b atresia is thought to develop subsequent to antenatal mid-gut volvulus.

Atresia type 4 - multiple intestinal occlusions occur. There may be 2 or more atresia or stenoses of the same or different types, and considerable shortening of the bowel. Type 4 lesions may result from intermittent torsion or placental emboli. Familial types have also been described.

Associated anomalies

Prematurity, growth retardation and co-existent malformation are common, particularly in patients with duodenal atresias. Almost 50% of duodenal atresias are associated with some other anomaly (e.g. cardiac, genitourinary or anorectal), and up to 30% of such patients have Trisomy 21. Less than 10% of infants with jejuno-ileal atresia, however, suffer from coexistent extra-abdominal malformations.

Diagnosis

The diagnosis of intestinal obstruction may be suspected during antenatal ultrasound examination. Polyhydramnios may be an indication of high intestinal obstruction, because the normal mechanism for re-absorption of the liquor through foetal swallowing is disturbed. Dilated fluid-filled loops of bowel may also be seen on antenatal ultrasound, and associated anomalies (e.g. Trisomy 21 and heart anomalies with duodenal obstruction) may be discovered. Some conditions (e.g. cystic fibrosis, Hirschsprung's disease) have an inherited basis, and may be suspected when siblings have been affected. Symptoms and signs of intestinal obstruction - vomiting (clear, or bile-stained if obstruction occurs distal to the ampulla of Vater), abdominal distension and a disturbed stooling pattern - are usually present from birth, but may vary depending on the level and completeness of obstruction. In incomplete obstructions, diagnosis may be delayed until well beyond the neonatal period.

Plain radiology, with air contrast introduced by a nasogastric tube if necessary, is sufficient to confirm the diagnosis in most patients. More information may be obtained from a barium meal when incomplete obstruction is suspected.

It is often impossible to distinguish between distended loops of small and large bowel on a plain radiograph. A diagnostic contrast enema is an essential aid in defining the level and cause of more distal obstructions.

Management

The principles of successful management of neonates with suspected obstruction are:

- recognise the condition early
- institute appropriate investigations
- assure complete peri-operative resuscitation

- perform the correct surgical procedure for the underlying condition

Laparotomy via a transverse abdominal incision above the umbilicus is preferred. In duodenal obstructions, the atretic area is exposed by mobilising the ascending and transverse colon to the left. In all types of duodenal obstruction, including stenoses and fenestrated diaphragms, the most widely performed operation is a diamond-shaped anastomosis using interrupted fine absorbable sutures. Anastomosis of dilated bowel and collapsed bowel distal to the origin of a 'windsock' diaphragm is a well-described pitfall.

In patients with small bowel atresias, the entire small bowel should be exteriorized and carefully inspected. The viable bowel length should be measured, because it is of prognostic significance. The proximal dilated bulbous blind end is resected and a primary, one layer end-to-end or modified end-to-back anastomosis is used in most patients. Stomas are fashioned only if there is gross peritonitis. Tapering of the dilated proximal bowel rather than resection is performed when total bowel length is deficient.

MECONIUM ILEUS

In meconium ileus, excessively viscous mucus secretion causes the meconium to stick to the intestinal wall and, in 10-15% of newborn infants with cystic fibrosis, this leads to acute intestinal obstruction. The distal ileum is small and contracted, and contains firm, greyish, pebble-like secretions of meconium. The mid-ileum becomes dilated and hypertrophied, and contains greyish-black material with a thick, tar-like consistency.

The small, unused 'microcolon' contains inspissated meconium pellets. Complications (volvulus, perforation and intestinal atresia) may occur.

Diagnosis

Visible abdominal distension with peristalsis and palpable bowel loops are evident. Plain radiographs of the abdomen reveal multiple dilated loops of bowel of varying diameter, but erect radiographs show a paucity of fluid levels. The meconium-filled bowel gives a 'ground glass' appearance, and contrast enema shows a minute, ribbon-like colon with meconium pellets.

Management

Uncomplicated meconium ileus is effectively treated in most patients by gastrografen enema. Gastrografen is a contrast medium that is hygroscopic, stimulates bowel peristalsis, and has a deterrent effect. Adequate intravenous fluid should be given to compensate for intraluminal losses, because gastrografen is hyperosmolar. When obstruction persists after gastrografen enema, or when complications are evident, surgical exploration is necessary. The authors favour placement of a T-tube into the thickened, dilated bowel. The tube is brought out of the abdomen through a stab incision, and the obstruction relieved by irrigation with a mucolytic agent (e.g. acetylcysteine). The tube can be safely removed after about 3 weeks, when bowel function has returned to normal.

In patients with complicated meconium ileus, resection and stoma formation is the preferred treatment; this is either a double-barrelled, or the Bishop-Koop, in which the distal end is exteriorized and the proximal bowel anastomosed end-to-side just within the abdomen. This allows for venting and irrigation in the postoperative period, with only limited surgery being required for stoma closure. Leak and torsion are recognised complications.

MECONIUM PLUG SYNDROME

Meconium plug syndrome typically presents as distal obstruction, caused by a cast or plug of inspissated meconium in the distal colon and

rectum. It may occur in infants of diabetic mothers, premature infants with intestinal dysmotility, hypothyroid babies, and infants of mothers receiving ganglion blocking agents or sedatives.

Management

Most infants respond to rectal washouts with half-strength gastrografen, following diagnostic contrast enema. Decompression may fail; urgent colostomy proximal to the obstruction then becomes necessary. Hirschsprung's disease and cystic fibrosis must be excluded.

HIRSCHSPRUNG'S DISEASE

The cause of Hirschsprung's disease is unknown, but failure of completely successful migration and maturation of the enteric ganglion cells, which develop from neuroblasts derived from the neural crest, is most likely. These cells normally migrate caudally along the path of the vagus nerve from the 6th week and reach the rectum by the 12th week.

The incidence is 1/5000 live births. Rectosigmoid disease occurs in 85% of cases and is more common in males. Long segment disease is more common in females than males, and where extended segments of bowel are involved. There are few associated anomalies; congenital heart disease and Down's syndrome occur in 5% of patients.

Absence of ganglion cells from the submucosal and myenteric plexus of the bowel, and thickened and disorganised parasympathetic nerve trunks are the typical histological features. In 70% of cases, only the rectosigmoid is affected, but aganglionosis may extend more proximally, affecting the left colon (20% of patients), whole colon (5%) or even small intestine (1-2%). There may be varying lengths of hypoganglionosis (transition zones), or abnormalities of parasympathetic nerve supply with abnormal ganglion

cells (neuronal intestinal dysplasia) in the bowel proximal to the aganglionic segment. Obstruction occurs because the affected segments are unable to transmit a peristaltic wave. If a short segment only is involved, the proximal bowel may hypertrophy and temporarily overcome the obstruction until later decompensation occurs, usually as a result of enterocolitis. In the neonatal period, enterocolitis has a significant mortality.

Diagnosis

Patients present with symptoms from the neonatal period, with failure or delay in passing meconium, abdominal distension and vomiting, in 80% of cases. Infants with longer segment Hirschsprung's disease often present with symptoms suggestive of incomplete small bowel obstruction.

Plain radiographs show dilated bowel with fluid levels. Typically, the small pelvis and rectum do not contain gas-filled loops of bowel, resulting in an 'egg on end' appearance. Contrast enema may be suggestive but can look normal, particularly if a digital examination has been performed in the preceding 24 hours. A delayed radiograph, 24 hours after the enema, will show barium retention. Diagnosis is confirmed by Meier-Ruge histochemical stain (acetylcholinesterase) of a suction rectal biopsy.

Management

Priorities are distal bowel decompression by rectal washouts, fluid resuscitation, and antibiotics for enterocolitis. Once full resuscitation is achieved, a temporary defunctioning colostomy is preferred; this is placed proximal to the aganglionic bowel, guided by frozen section histology of seromuscular biopsies. Definitive surgery is usually postponed until after the neonatal period, though earlier definitive surgery has recently been advocated.

Surgery for Hirschsprung's disease involves placing normal innervated bowel just above the dentate line. An excellent result is obtained in 90% of patients. The definitive procedure is increasingly undertaken in the neonatal period, without colostomy.

INFANTILE HYPERTROPIC PYLORIC STENOSIS

Pyloric stenosis is an abnormality of the pyloric musculature causing gastric outlet obstruction in infancy. Incidence: 3:1000 live births. There is a family history in 5 - 20% of patients. It occurs more commonly in males.

Etiology unknown

Predisposing factors include male sex, first-born child, susceptible race and known family history.

Pathology

The musculature of the pylorus is thickened and hypertrophied. Following pyloromyotomy the pylorus returns to normal over weeks to months.

Clinical Course

Non-bile stained vomiting (often projectile in nature) usually starts at the age of 2-3 weeks and becomes progressively more severe. The infant becomes dehydrated and malnourished with weight loss. Typical biochemical disturbances include hypochloraemia, hyponatraemia, hypokalaemia, metabolic alkalosis and unconjugated hyperbilirubinaemia.

Diagnosis

A history of projectile vomiting, visible gastric peristaltic waves and a palpable pyloric 'tumour' constitutes the diagnostic triad. Radiographic examination shows an enlarged stomach and a barium meal will confirm the long attenuated pyloric canal with stomach outlet obstruction. U/S shows diagnostic elongation of the pyloric canal and thickening of the circular muscle.

Differential Diagnosis

- 1) Gastro oesophageal reflux
- 2) Feeding mismanagement
- 3) Pyloro spasm
- 4) Salt-losing adrenogenital syndrome
- 5) Increased intracranial pressure

Management

When the diagnosis of pyloric stenosis has been established, dehydration and biochemical abnormalities must first be corrected with the infusion of half normal saline with added potassium. This may take 2-3 days.

Surgical Treatment

Pylorogastromyotomy. The avascular area of the pyloric tumour (the antero-superior surface) is identified and the hypertrophied pyloric muscle incised over the full length and is widely separated to expose the submucosa, thus releasing the obstruction.

Should perforation occur during myotomy, it is easily recognised and should be sutured immediately. Haemorrhage from the cut edges usually stops spontaneously.

Results

Pyloromyotomy has proven to be a very effective and safe treatment with minimal mortality and morbidity and lasting relief of symptoms.

ANORECTAL MALFORMATIONS

The term 'anorectal malformation' describes a variety of congenital abnormalities of the anus, rectum, urethra, bladder and sacrum in the male and in the female this also includes the genital tract. There is disordered embryogenesis such that the anus or rectum fails to develop and is either stenotic, covered by perineal skin or ectopically sited anteriorly in the urethra or bladder in the male or vagina in the female. The opening is usually vestigial in nature and is often

referred to as a fistula. The muscle sphincter complex, a caudal extension of the levator ani, which normally surrounds the anus and rectum may also be deficient but to a lesser degree. In the past ARM's in which the abnormality ended above the level of the muscle complex were referred to as 'high' and those extending through the complex as 'low'. This has prognostic implications in that infants with 'low' anomalies would be expected to be continent of faeces after reconstructive surgery, whereas those with 'high' anomalies have a more guarded prognosis. About 60% of ARM's are high and 40% low.

Diagnosis

Infants present with abdominal distension, vomiting and failure to pass meconium after birth.

The diagnosis is confirmed by looking at the anal area. In the male the difference between high and low can be established by searching for meconium. If meconium appears anywhere on the perineum or anterior to the site where the anus should be, the lesion is of a low type. Should meconium however appear in the urine, the lesion is of a high type and indicates a recto-urethra fistula. In females the diagnosis easily established by counting visible perineal orifices. The normal female perineum has three orifices visible on inspection. i.e. urethra, vagina and anus. Where only urethra and vagina are visible and meconium is seen in the vagina this would imply a recto vaginal fistula. Where only one orifice is visible this implies that the urinary, genital and gastrointestinal tracts open into a single outlet, which is called a cloaca. This is a 'high' lesion.

If there are three orifices - urethra, vagina and the anal orifice posterior to the introitus - the diagnosis is that of a low ARM.

Low ARM's are treated at presentation by local anoplasty. High ARM's are

given an initial an initial left sigmoid divided colostomy. This is followed by a pull-through procedure after 6 months to a year. The most important feature of the operation is placing the rectum and anus within the muscle sphincter complex en route to the perineum. Most high lesions do not have an internal sphincter thus true continence is rarely attained

EXOMPHALOS

Congenital defects of the anterior abdominal wall are usually severe malformations. The incidence varies from 1 in 3,000 to 1 in 30,000 births. In exomphalos a thin-walled sac, containing abdominal viscera, protrudes through a congenital defect of varying size in the abdominal wall at the level of the umbilical area. The sac consists of amniotic and peritoneal membranes containing liver and usually small bowel. The condition is often associated with other serious cardiac, diaphragmatic or intestinal abnormalities. One of the most serious associations is the Beckwith Wiedeman syndrome, which is associated with persistent hypoglycaemia. The size of the exomphalos is extremely variable. If the parietal defect is > 5cm it is called a major exomphalos. Those with a defect < 5cm are called minor. Three major complications can occur: infection, rupture of the sac and hypoglycaemia.

Management: Exomphalos Major

- Immediate operation and complete repair in 'minor' defects.
- Major defects especially if the infant is premature or has other abnormalities are treated conservatively. The sac is painted with an antiseptic and desiccating agent. Epithelialization of the sac occurs over weeks to months. The ventral hernia is then repaired at a later date.
- Ruptured or large exomphalos when primary closure is impossible, are managed through

fashioning of a "silo" of synthetic material to contain the viscera. This sac is then reduced in size over a period of days until all the contents are accommodated with the abdominal cavity when the silo is removed and the abdomen sutured close.

GASTROSCHISIS

In gastroschisis there is a defect in the anterior abdominal wall through which abdominal contents prolapses during several months of foetal life. The abdominal wall defect is usually to the right of the umbilical cord and only a few centimetres in diameter. Additional malformations apart from intestinal atresias are rare.

Therapy

The treatment is the same as for a ruptured exomphalos. An attempt is made to replace the bowel within the peritoneal cavity and repair the defect. If this cannot be done without compromise of respiration and/or development of abdominal compartment syndrome then a silo is created and reduction then takes place over several days as described for ruptured exomphalos major.



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