

## **C J Mieny, U Mennen: Principles of Surgical Patient Care - Volume II**

### **Chapter 15: Paediatric Surgery**

#### **Chapter 15.1: Diaphragmatic Hernia**

**M R Q Davies**

##### **Introduction**

The developing diaphragm should be regarded to be a single structure, the septum transversum (His), with five hiatuses and certain blended regional subdivisions. Three of the hiatuses are permanent (caval, oesophageal and aortic), and two temporary (right and left pleuroperitoneal canals). They form between the eighth and tenth week of foetal life, a period during which two other events require consideration. The lung buds invaginate into the primitive pleural cavities, and the midgut loop commences its return to the peritoneal cavity.

*During foetal and early life, herniation of abdominal viscera into the mediastinum or pleural cavities can occur.*

##### **Mediastinum**

- > Through the oesophageal hiatus into the posterior mediastinum. Sliding, rolling or mixed hernias occur that usually involve the stomach.
- > Through the central tendon into the pericardial sac.
- > Through the foramen of Morgagni into the anterior mediastinum.

##### **Pleural Cavities**

- > Through pleuroperitoneal canals.
- > Through lumbocostal trigones (Bochdalek).

Both of these weaknesses are situated posterolaterally in the diaphragm.

An abnormal elevation of all or part of an intact diaphragm is known as an eventration. In the absence of a definable neck and sac to the hernia, a partial eventration cannot be distinguished from a diaphragmatic hernia with sac.

##### **Congenital Posterolateral Diaphragmatic Hernias**

Incidence: 1 in 4000 live births.

Diaphragmatic hernias are a well-recognised cause of foetal death. When not detected prenatally only 50% of cases born alive with this condition reach help.

Two type determined by the site of the defect in the diaphragm are recognised; this distinction is not important:

Pleuroperitoneal Hernia

Kidney uninvolved  
Posterior muscular rim present  
Defect lateral to triangular ligament of liver.

Lumbocostal hernia (true Bochdalek)

Kidney one of herniated viscera  
No posterior muscular rim  
Defect medial to triangular ligament of liver

Overall incidence of right-sided to left-sided hernias is 1 to 10; 10% of the hernias will have a sac.

### **Pathophysiology**

*The herniated viscera produce a mass effect within the chest cavity:*

--> During foetal life, depending upon the time of the prolapse in relation to the growth of the invaginating lung buds and its size, this impedes lung development, causing varying degrees of pulmonary hypoplasia which is most severe on the side of the hernia.

--> After birth, depending upon the degree of intestinal distension by air, this produces further lung compression, impeding respiration.

The pulmonary hypoplasia, if bilateral, might be of a degree incompatible with life. If the patient survives treatment, chronic respiratory insufficiency is seldom encountered in later life. During the immediate postbirth period, pulmonary hypoplasia is associated with abnormal pulmonary vasoconstriction, which gives rise to persistence of foetal or higher pulmonary arterial pressures. Alveolar hypoxia and the presence of an acidosis are two major triggering factors. Persistent pulmonary hypertension precipitates right to left shunting of blood, usually at the level of the ductus arteriosus, rarely within the lung or more proximally in the heart (this is known, as a persistent transitional (foetal) circulation). An already compromised respiratory situation is worsened by the event which, once well-established, may be refractory to treatment.

The defect within the wall of the peritoneal cavity, which is present during the return of the midgut loop from the extra embryonic coelom, causes not only a hernia, but varying degrees of imperfect fixation and rotation of the colon. Duodenal compression or in exceptional instances an intestinal volvulus are due to this anomaly. The hernia may complicate. Strangulation of viscera may occur. Entrapment of a portion of liver within a hernia, is a well recognised cause of an unexplained right-sided hydrothorax.

### **Clinical**

In the neonate and during infancy, symptoms reflect compromised respiratory function. In the child gastrointestinal complaints often predominate.

*A diaphragmatic hernia is a cause of:*

### **Polyhydramnios**

The hernia can be demonstrated sonographically prenatally. As these babies do not

transfer well, referral to a tertiary paediatric care centre prior to the delivery of the case is a priority.

### **Respiratory Distress Syndrome of the Newborn**

The symptoms, cyanosis and dyspnoea, do not help to distinguish this condition from the more common "paediatric" causes.

#### **The astute observer will note the following signs:**

- > An apparent dextrocardia. Dextroposition of the heart due to a left-sided hernia.
- > The abdomen is not distended or, more rarely, empty and scaphoid in appearance.
- > The oxygen test - if anything helps, the respiratory symptoms improve on the administration of oxygen.

### **First Aid Management**

- > Correct the hypoxia by increasing the  $FiO_2$ .
- > Headbox or, preferably, endotracheal intubation and ventilation.
- > Correct the hypercarbia; blow-off  $CO_2$ .
- > Prevent air entering the stomach and decompress the upper intestinal tract by tube.

### **Diagnosis**

Order an urgent X-ray of the chest plus **abdomen** (on one photograph = **babygram**).

Fluid-filled and/or gas filled cyst-like shadows forming a mass will be seen with shift of the mediastinum to the uninvolved side. The infra-diaphragmatic intestinal gas pattern is not normal as most of the bowel is in the chest. Right-sided hernias are more difficult to diagnose:

- > the liver appears to be partially dislocated into the chest
- > in most instances intestinal gas shadows are seen in a supradiaphragmatic position.

### **Differential Diagnosis**

- > Congenital cystic adenomatous lesion of the lung.
- > Acquired cysts - pneumatocoeles (post-staphylococcal pneumonia).

In both of these conditions the stomach and intestines are seen beneath the diaphragm on a chest X-ray.

## **Treatment**

*The degree of respiratory distress can be crudely graded using preoperative arterial blood-gas results:*

- > Normal
- > Abnormal - correctable
- > Abnormal - uncorrectable.

Historically, the time lapse from birth to time of presentation (under six hours = predicted 50% + mortality) was used to estimate the degree of lung hypoplasia. Improved perinatal care has resulted in an ever increasing number of very early detections. Babies with very marginal capabilities, enabling them to maintain a separate existence, are now being seen by the surgeon.

*Newborn cases are now placed into one of the following two groups:*

### **Group A**

#### **Immediate Detection and Correct Resuscitation**

The intrathoracic mass has retained its foetal status; it has not increased in size. The degree of respiratory distress is directly related to the degree of pulmonary hypoplasia present. The operative reduction of the hernia is elective and is only preceded with once the case has been stabilised.

#### **In an ICU Setting**

--> IPPV + respiratory paralysis. Mechanical respiratory support is the most important aspect of treatment. Use rapid rate (panting) ventilation, with ventilator settings that do not markedly impede lung perfusion and without an excessive risk of causing alveolar rupture (pressures exceeding 45 mm Hg or 60 cm H<sub>2</sub>O).

The FiO<sub>2</sub> is regulated to maintain:

- > Preductal PaO<sub>2</sub> greater than 75 and less than 100 torr.
- > Postductal PaO<sub>2</sub> greater than 40 torr.
- > Preductal blood samples are taken via umbilical arterial catheter situated just above the aortic bifurcation.

Please note - do not attempt to catheterize these vessels unless you are well acquainted with the procedure, as unsuccessful puncture could preclude their later use. Transcutaneous monitors can be used.

--> CVP lines can be inserted. Right atrial pressure of  $\pm 2$  cm of water is aimed at. Infuse 5% dextrose/0.45 saline as required.

--> Correct the metabolic acidosis by improving the circulation and hypoxaemia. Every effort must be made to correct hypoxia, hypercarbia and acidosis (aim at a pH 7.35-7.45).

If, in spite of the above measures, respiratory insufficiency incompatible with life cannot be reversed, there is no indication to operate upon the patient.

beware of the undetected pneumothorax. Any sudden change in respiratory status could indicate its presence. Chest-wall transillumination and immediate needle thoracocentesis are life-saving.

## **Group B**

### **Late Detection or Incorrect Early Management**

A post natal increase in size of the herniated viscera, produces additional lung compression, that can be corrected surgically. A history of respiratory function adequate to maintain a separate existence for any length of time indicates a salvageable situation.

### **Repair**

A transabdominal approach is used. Through a transverse incision, the contents of the hernia are carefully reduced. A hernial sac if present, is excised or any posterior rolled up rim of diaphragmatic muscle is found and dissected free. The defect is sutured using interrupted stitches of nonabsorbable material. In 90% of cases, a simple repair can be achieved. A Gortex patch is used where this is not possible.

Any significant degree of malrotation is corrected and the abdomen closed. This is often the most difficult part of the operation.

*If it is necessary to enlarge the peritoneal cavity, carry out the following manoeuvres:*

- > Manually stretch the anterior abdominal wall.
- > Empty the colon and stomach of their contents.
- > An anal dilatation, before commencing the operation is of benefit.
- > Patch the defect, restoring the natural contour of the diaphragmatic leaf. Prosthetic material is used here in preference to placing it in the laparotomy wound.
- > Close the laparotomy wound with skin only (create ventral hernia).

**Note:** Pleural drainage tubes are not used or placed routinely.

### **Postoperative Management**

- > Maintain preductal PaO<sub>2</sub>: avoid abrupt changes. Use respiratory paralysis (pavulon or curare).
- > Maintain PaCO<sub>2</sub> under 55, using whatever ventilator pressures and rates needed.
- > Correct "third space" losses to maintain adequate CVP + Urine output; when enhanced cardiac output is required dopamine 300 microg per kg per h is used.

Vasoactive drugs, ie, priscoline, are of questionable value where persistent pulmonary hypertension and significant shunt are present (preductal PaO<sub>2</sub> 75 to 100 torr at FiO<sub>2</sub> greater than 40% and postductal PaO<sub>2</sub> less than 50 torr).

## **Prognosis**

"The disease that promises everything, but gives nothing!" The survival of the fittest - the degree of pulmonary hypoplasia determines the outcome; when severe it causes a respiratory death; when marginal it could precipitate a cardiac death because of pulmonary hypertension. Although prenatal repair of diaphragmatic hernias has proved feasible in animals, foetal surgery of this magnitude in the human is still experimental.

Terminating the pregnancy at the earliest opportunity assuring a viable patient, brings with it the added risks of lung immaturity. The lecithin/sphingomyelin ratio (bubbles test) is a poor guide in these patients, compounding the problem.

## **Complications**

- > Pneumothorax - significantly worsens the prognosis.
- > Chylothorax - as a result of the operation.
- > Bowel obstruction.
- > Midgut volvulus.
- > Retinopathy of prematurity.

## **Comment**

### **Diaphragmatic Hernia**

#### **J H Louw**

In the pathophysiology of diaphragmatic hernia it should be stressed that the effect of pulmonary hypoplasia are a decrease in the total size of the lung, a reduction in the number of vessels per unit area of lung and increased muscularisation of the pulmonary arterial tree.

In the management of the condition, I believe that pleural drainage tubes should not be placed but clamped until a pneumothorax or hydrothorax should develop.

In an effort to improve the prognosis extracorporeal membrane, oxygenation to support infants with reversible respiratory failure has been used, but its role in diaphragmatic hernia is currently uncertain.

In an endeavour to tide these babies over the critical period of lung hypoplasia and poor oxygenation, some units have used extracorporeal oxygenation, but this has not improved survival.

## Chapter 15.2: Oesophageal Atresia and Tracheo-Oesophageal Fistula (OA/TOF)

M R Q Davies

### Definition

An atresia (an all or none situation), by definition means *no hole*. When present in the oesophagus, the congenital form occurs at a level between D1 and D3; some 10 cm by measurement from the anterior nares.

### Anatomy

In practical terms three morphological types occur:

**Oesophageal atresia (OA)/Tracheo-oesophageal fistula (TOF)** - common type; 85% of cases. Proximal oesophagus atretic at D2; distal oesophagus connected to the trachea via a fistula which opens near the carina.

**Oesophageal atresia without fistula;** 10% of cases. Proximal oesophagus atretic at or below D2; distal oesophagus ends short of the carina.

- > With an intrathoracic portion
- > Ends at or beneath the diaphragm

**Tracheo-oesophageal fistula without oesophageal atresia;** 5% of cases. Cervical oesophagus communicates with the trachea at or above the level of the thoracic inlet.

### Associated Anatomical Abnormalities

Abnormal development of the arch of the aorta

- > Aberrant right subclavian artery - common
- > Right-sided aorta - rare

Airway abnormalities

- > Laryngo-oesophageal (tracheo-oesophageal) cleft - rare
- > Deformation of the trachea due to compression by an enlarged obstructed proximal oesophageal pouch ("tracheo-malacia") - degrees always present.

The VATER association. This is a conglomerate of defects; the most important from a prognostic point of view is left out by this acronym, ie, cardiac anomaly which could be life threatening.

V = vertebral; anomaly usually unimportant

A = anorectal anomaly; in the male usually supralevator and major; in the female the enteric opening can usually be visualized

T & E = Oesophageal atresia/tracheo-oesophageal fistula

R = radial hypoplasia (clubbing) of the upper extremity, or renal anomalies which are usually of a minor nature.

## Oesophageal abnormalities

The functional are more important than the anatomical and affect the postrepair long-term prognosis. Reflux, aspiration and dysphagia occur, due to dysmotility.

Intestinal abnormalities - uncommon but must be looked for

- > Duodenal atresia or stenosis
- > Malrotation of midgut loop
- > Hypertrophic pyloric stenosis.

## Aetiology

Unknown and probably multifactorial. Seen in siblings and progeny of surviving cases. The embryology of the oesophagus and trachea is complex. The lesion occurs during the early organogenetic phase of development, 18-55 days after conception. As foetal deglutition is affected, the normal foetus amniotic-fluid dynamics are disturbed:

- > OA/TOF - 1/3 associated with polyhydramnios
- > OA without fistula - 100% associated with polyhydramnios.

Polyhydramnios therefore is a significant prenatal marker and oesophageal atresia is a probable cause when the foetal stomach cannot be demonstrated by sonar.

*Incidence:* 1 in 4500 live births.

## Clinical Features

Polyhydramnios, preterm delivery and respiratory distress are 3 interrelated pointers to the presence of an OA: when present without an obvious cause an underlying atresia of the oesophagus must be excluded.

"The presence of an oesophageal atresia is excluded when a catheter is passed via the nostril into the stomach."

This test is used routinely by many obstetrical services.

## Total Dysphagia

--> A failure to swallow its own saliva - "blowing bubbles"; requires repeated pharyngeal suctioning.

--> A failure to feed - refusal or when forced, choking occurs.

--> The complication caused by aspiration are present. Typically these follow attempts to feed a baby with an isolated TOF without atresia. *In these instances remember that:*

- Oesophageal motility is abnormal: the baby swallows with difficulty.
- Aspiration occurs during feeding.
- A significant degree of tracheomalacia is usually present which accentuates the respiratory symptoms.



*Note:* The pulmonary pathology due to aspiration is often more severe on the right and is seen as atelectasis (upper lobe) or a combined picture of collapse and bronchopneumonia.

### **Diagnosis**

The biggest catheter that can be introduced without causing damage is passed into the proximal oesophagus via the nostril. A complete obstruction is demonstrated  $\pm$  10 cm from its tip. An X-ray (babygram) determines whether intestinal gas is present. This establishes the presence or absence of a patent TOF. It also diagnoses pulmonary complications, associated congenital anomalies and the presence or absence of intestinal obstruction.

### **Differential Diagnosis**

Flimsy thin-walled feeding tubes will often curl up in the pharynx following their introduction. A catheter that allows a measure of tip control is required to diagnose an atresia (10-12F).

A posterior pharyngeal laceration may complicate endotracheal intubation, give rise to extravasation and the formation of a pseudo diverticulum. This is a rare cause of acquired oesophageal obstruction in the newborn. The history of stormy resuscitation attempts and pharyngeal bleeding suggest this diagnosis, as does the presence of an obstruction lower down than expected in the midthoracic oesophagus. Endoscopic assessment of the pharynx will establish its presence.

If the presence of an isolated TOF is suggested clinically it must be aggressively sought by contrast radiology and/or diagnostic endoscopy.

### **First-Aid Management**

The presence of OA has been confirmed by catheterisation. No contrast radiology is ordered as it is not required. The presence of a TOF is assumed when intestinal gas is present.

### **Prevent Aspiration**

Proximal pouch decompression by catheter - intermittent by hand-held syringe is preferred. Use a Replogle catheter (10F) if continuous suction is to be used.

### **Prevent Gastro-Oesophageal Reflux (The Acid "Burp")**

Causes aspiration via the TOF which is of far greater danger to the patient than pharyngeal spill.

## **Remember**

- The ability of the newborn's stomach to produce acid.
- Babies are "physiologic" refluxers.
- Gaseous distention of the upper intestinal tract impedes respiration by causing diaphragmatic splintage.

A 35° head-up prone position may partially prevent contamination of the airway by gastric content.

## **Treat Atelectasis and Pneumonia If Present**

### **Treatment**

It is emphasised that no contrast radiology is used to clarify the oesophageal pathology unless a seldom-seen variant is thought to be present.

The surgical repair aims to reestablish gastrointestinal continuity using the oesophagus preferentially. This may not be possible when saving life takes precedent.

### **Timing the Operation**

The case is stabilised, appropriate anti-microbial prophylaxis is commenced with, and correctable complications are aggressively managed (hyaline membrane disease; bronchopneumonia and/or atelectasis).

Following on the institution of these measures the surgical options are considered.

Non-correctable associated defects could exclude further treatment (trisomy, cardiac).

## **Oesophageal Atresia with TOF of the Common Type**

### **No Acquired Complications Are Present.**

Smallness in size does not preclude a primary oesophageal repair. This is undertaken as an elective emergency via a right extrapleural thoracotomy approach to the mediastinal structures involved. The TOF is identified, clearly defined, divided and the tracheal end securely closed. In a similar fashion the proximal oesophagus is dissected free from the trachea, opened and an end-to-end axial anastomosis (watertight) is performed between the two oesophageal ends.

--> Routine gastrostomy is not performed.

--> The finest suture material (6-0 PDS; Prolene; braided polyester) appropriate for both patient and surgeon is used.

--> The anaesthetic must be given by an experienced anaesthetist as its administration has as important an influence on the outcome as the operation.

--> The retropleural space is drained by an underwater sealed system.

The patient is managed postoperatively in an intensive care unit. The case is kept intubated and on respiratory support for the first 36 hours from which time weaning is commenced. This is done to protect the anastomosis. Careful pharyngeal suctioning is carried out making sure the catheter never reaches the site of the anastomosis. Routine nasogastric intubation is not used as it is not required. The patient is fed intravenously for 7 days to 10 days at which point a carefully performed contrast study is used to assess the anastomosis. If this is patent and intact, oral intake of a graded type is ordered and the retropleural drain removed. Routine dilatation of the oesophagus is not used.

In the unlikely event of the demonstration during surgery of an abnormally long gap between the two oesophageal ends, every effort is made to obtain an end-to-end anastomosis. An upper pouch myotomy (Livaditis) may achieve the desired aim. If not, the procedure is abandoned, the distal oesophagus closed, the proximal pouch brought out in the neck as an oesophagostomy and a Stamm gastrostomy constructed. An oesophageal replacement procedure of choice is performed at a year.

### **OA/TOF of the Common Type Complicated by Severe Respiratory Distress Syndrome**

This is either due to hyaline membrane disease (rare) or bronchopneumonia. The patient is placed on appropriate respiratory support and is treated actively for the complication in an ICU setting. The proximal oesophageal obstruction is controlled by catheter suction. Reflux via the TOF can be partially prevented by using CPAP, delivered via an endotracheal tube.

Once primary repair becomes feasible, it is immediately carried out. If however, the patient's condition does not improve or worsens the TOF must be mechanically controlled. This is best done via a laparotomy; the distal intra-abdominal oesophagus is ligated with umbilical tape and a gastrostomy is carried out. This manoeuvre does not preclude the use of the distal oesophagus at a later date, as a site of stenosis left by the oesophageal ligation has proved to be simple to correct by dilatation. A delayed primary repair is performed when indicated.

**Note:** The patient in RDS has noncompliant lungs. A large air leak to the upper GIT is present. Tube gastrostomy will render the patient difficult or impossible to ventilate. The TOF must be controlled in this situation.

### **OA Without TOF**

A long anatomic gap between the oesophageal segments is always present. Primary repair is not possible shortly after birth. The proximal oesophageal obstruction is controlled by catheter. An elective gastrostomy is performed for feeding. At 6-8 weeks of age the ends of the oesophagus are intubated and the gap is estimated radiologically. It has been shown that due to normal growth, these two ends approach one another, but as the spine grows disproportionately more rapidly after  $\pm$  8 weeks, successful axial anastomosis becomes less likely. If it appears to be feasible the oesophagus is approached via a right extrapleural thoracotomy. Myotomies are usually required and in many instances the two ends can be anastomosed. Severe GOR often complicates this procedure.

## **Isolated TOF**

The patient is tube fed until an elective repair is indicated. This is carried out via a cervical incision and is preceded by a bronchoscopy and intubation of the fistula with a ureteric catheter.

## **Prognosis**

As our ability to manage premature patients has dramatically improved, mortality is linked to life-threatening infective complications and major associated congenital defects. An OA should be repaired without mortality today.

## **Complications**

### **Early**

### **Oesophageal**

### **Anastomotic Leak**

--> Major within 36 hours of repair - managed by reexploration and reanastomosis if feasible.

--> Major after day 2: If intrapleural or if it is not possible to prevent lung collapse, due to the rapid accumulation of intrathoracic gas, the oesophagus is abandoned and an oesophagostomy plus gastrostomy, as life-saving measures, are performed.

--> Minor: Any leak contained within the retropleural space can be managed conservatively with an expected satisfactory outcome.

## **Recurrent TOF**

It is difficult to confirm this diagnosis; it often complicates a recognised anastomotic leak and was associated with the use of silk suture material in the past. It is closed electively using routine surgical principles.

## **Anastomotic Stenosis/Stricture**

This is a common problem and occurs in at least 10% of all cases. Its presence must be excluded when:

--> Feeding difficulties are present.

--> The patient develops new or recurrent respiratory complaints.

The stenosis is confirmed by a contrast swallow. Most stenoses yield to 1 or 2 dilatations.

A fibrotic stricture is best managed by excision and reanastomosis.

## **Oesophagitis Due to Pathological GOR**

Simple gastro-oesophageal reflux is commonly encountered in these patients. It rarely gives rise to severe oesophagitis but has been incriminated in the development of recurrent anastomotic strictures. It is managed medically in the first instance.

Reflux seen in association with a major degree of tracheomalacia has a sinister reputation and is linked to sudden infant death syndrome. Apnoeic episodes and recurrent aspiration are best treated by an urgent fundoplication.

### **Respiratory Complications**

#### **Tracheomalacia**

The flaccid trachea is the cause of the TOF cough. This tracheal anomaly gradually corrects itself and in most cases is only of minor importance. It is a cause of dyspnoea during feeding which may be mistakenly attributed to oesophageal pathology. The symptoms, however, usually start a month or two after successful primary repair. Exclude other correctable lesions, if no improvement is noted, bronchoscopic assessment of the tracheal deformity is done and an aortopexy performed if indicated.

#### **Chronic Airways Symptomatology**

A certain percentage of patients continue with repeated episodes of respiratory infection that cannot be directly explained by any of the above described lesions. It usually settles down in time and is supposedly a summation of minor degrees of all the above anomalies and their complications.

#### **Late**

A degree of dysphagia due to dysmotility of a functional nature is always found in these patients. Repeated episodes of bolus oesophageal obstruction, aspiration and oesophageal pain can occur. Known causes are excluded before the patient is branded an oesophageal cripple, a state seldom encountered following the successful primary repair of an oesophageal atresia.

#### **Comment**

### **Oesophageal Atresia and Tracheo-Oesophageal Fistula**

#### **J H Louw**

I agree that contrast radiology is not required as part of the first-aid management of oesophageal atresia, however, injection of air may delineate an upper blind pouch in the neck.

When intestinal gas is present a tracheo-oesophageal fistula may be assumed but should be confirmed first by tracheoscopy which has the added advantages of locating the exact site of the fistula, detecting the presence of tracheomalacia and visualising

movement of the vocal cords.

Contrast radiology may be considered if seldom-seen abnormalities such as proximal fistula, proximal and distal fistulas, minute fistulae, stenosis without a fistula are thought to be present. Diagnostic endoscopy is less dangerous and more useful for the reasons given above.

Although routine nasogastric intubation is not required after repair of the oesophagus, a fine silastic tube may be guided through the anastomosis just before its completion for subsequent tube feeding.

### **Chapter 15.3: Exomphalos/Gastroschisis**

**M R Q Davies**

**Ex = out**

**Omphalos = navel**

**Gastro = belly**

**Schisis = split**

An exomphalos (omphalocele = umbilical hernia) is present where there is herniation of abdominal viscera into the base of the umbilical cord.

#### **Anatomy**

*As with any hernia the following anatomical parts are identified:*

--> **The sac:** formed by the parietal peritoneum.

--> **The coverings of the sac:** a middle incomplete layer of Wharton's jelly plus the 2 umbilical arteries and a single umbilical vein, are contained within the outer lining, which is part of the amnion.

--> **The neck of the sac** is a defect in the midline aponeurotic layer of the anterior abdominal wall.

Based upon the location of this fascial opening, a simple classification has been proposed - the prefixes minor, major, giant representing less than 2.5 cm; 2.5 to 5 cm; greater than 5 cm, are added to indicate its size.

#### **Upper Midline Anomaly**

When 5 structural defects are present, this is called the Pentalogy of Cantrell:

- > Cardiac anomaly - commonly a ventricular aneurysm.
- > Pericardial defect or opening.
- > Diaphragmatic defect or opening.
- > Sternal defect or opening.
- > A defect of the linea alba.

Clinically, a large hernia with a sessile base and containing a major part of the liver, is found.

### **An Umbilical Anomaly**

The neck of this hernia is contained within the umbilical ring. The ring is abnormally large in these instances. Clinically a small hernia often based on a pedicle or a large hernia is found. The sac is filled with bowel and may or may not contain part of the liver. When a narrow-necked hernia is identified containing bowel, it is called a hernia of the umbilical cord, which possibly represents a physiological aberration of failure of return of the midgut loop following its extrusion into the base of the body stalk.

*Two important variants have been identified in this group:*

### **EMG**

The exomphalos may be one component of the Beckwith Wiedemann syndrome, an obscure grouping of anomalies of uncertain cause. This is best remembered by the acronym EMG:

- E = exomphalos
- M = macroglossia
- G = gigantism

*The large baby with a big tongue and an umbilical hernia.*

### **Gastroschisis**

When prenatal rupture of the sac of an exomphalos has occurred, the baby is born with gastroschisis.

#### ***Morphological Features of Gastroschisis***

--> A full thickness defect in the abdominal wall, lying to the right of the umbilical cord which is attached to the left side of its perimeter by the umbilical vessels that traverse the same fascial defect.

--> Prolapse of abdominal viscera through the defect - the midgut loop based on a universal mesentery with or without the bladder, testis, internal female genitalia, but never the liver, are encountered outside the abdominal cavity. Crowding of these structures as they traverse the fascial ring is common. Not surprisingly, venous and lymphatic insufficiency occur and in rare instances gangrene and atresia formation are seen.

Depending upon the length of the direct exposure of the intestines to the liquor amnii, various degrees of secondary peritonitis are found with the production of a serosal surface peel or exudate. These secondary changes have a detrimental effect on peristaltic activity with a delay in onward movement of intestinal contents and inspissation of meconium, which may further traumatise the bowel wall. The midgut loop is noticeably foreshortened in these cases.

--> The absence of a hernial sac. If sac tissue can be identified the lesion is deemed to be an exomphalos. This distinction is of no importance.

### **Lower Midline Abnormality**

The lesion lies below the insertion of the cord in the hypogastrium and is associated with:

- > Extrophy of the bladder.
- > Cloacal extrophy (Vesico-intestinal fissure).

The exomphalos is usually minor and simple to treat. An associated meningomyelocoele is often present.

### **Aetiology**

Unknown - probably multifactorial.

Overall incidence 1:7500 live births with a worldwide increasing prevalence of gastroschisis. The baby with a gastroschisis is often born preterm and may be small for gestational age. Gastroschisis is seldom associated with other major congenital anomalies and is not encountered in the EMG syndrome.

This contrasts with exomphalos where in many instances systems other than the body wall are affected by the dysmorphogenesis and chromosomal aberrations are encountered.

Prenatal sonography has demonstrated that:

--> A large liver containing exomphalos can consolidate so that at birth only a minor and easily treatable variant is present.

--> An obvious exomphalos can rupture during foetal life so that at birth a classic gastroschisis is found.

Whether all cases of gastroschisis represent examples of prenatal rupture of an exomphalos (minor form), is still debatable. Defects in the abdominal wall lateral to the midline and cord insertion are difficult to explain on this basis.

### **First-Aid Management**

"These babies always drop their core temperature significantly". Every effort should be directed at preventing this happening. Following the birth of the baby, the umbilical cord is clamped and divided, the baby is inspected and then placed into a surgically clean plastic bag with its head protruding (baby in a Jiffy Bag). The gastrointestinal tract is kept decompressed by nasogastric tube. In the EMG syndrome inappropriate insulin production occurs. Blood sugar levels are checked and hypoglycaemia which may be difficult to manage, prevented by intravenous dextrose.



## Treatment

Lethal chromosomal aberrations, ie, Trisomy 13, may preclude any form of therapy. *From the surgeon's viewpoint the problem is one of sizes:*

- 1) Volume/capacity of the abdominal cavity.
- 2) Volume/capacity of the extra-abdominal viscera that have lost their right of domicile.

*Number 1* is directly influenced by the size of the baby - the bigger the baby, the easier it is to enlarge the peritoneal cavity.

*Number 2* - reducing the size of the viscera so that they can be accommodated within the receiving cavity, is only possible with those that are hollow and can be emptied of their contents. In most cases of non-liver containing exomphalos and gastroschisis, a reduction of the prolapse and a primary closure of the abdominal wall is possible.

This is undertaken as an emergency operation.

## Operative Treatment

--> Prevent hypothermia.

--> Empty the intestinal tract of its contents via the stomach and rectum. In gastroschisis an anal dilatation and gentle saline irrigation of the colon, introduced via a catheter aids colonic evacuation.

--> Enlarge the opening in the abdominal wall by dividing the linea alba; a sac, if present, is excised; the insertion of a normal cords is left untouched. Inspect the peritoneal cavity and check that the diaphragm is intact: no attempt is made to unravel the bowel in gastroschisis but intestinal atresias should be identified.

--> Stretch the anterior abdominal wall manually thereby enlarging the peritoneal cavity until primary closure is possible. The use of a gastrostomy is optional in these cases.

--> Close the fascia with one layer of interrupted sutures, leaving the umbilical cord attachment *in situ*. With uncomplicated healing the patient will have a normal-appearing navel.

**Note:** Using this technique the intra-abdominal pressure is abnormally raised for the first 48 hours to 72 hours of the postoperative period.

*This implies:*

--> IPPV will be necessary for  $\pm$  48 hours in all gastroschisis and other major cases.

--> 3rd space losses occur when fluid is trapped in the lower extremities as IVC blood flow is impaired.

This form of repair would be inappropriate if the patient has an already compromised respiratory system (hyaline-membrane disease, pulmonary hypoplasia).

When a "safe" closure is not possible a prosthetic patch or bag is used. The liver containing exomphalos is best treated by staging the reduction. This necessitates the use of a silastic pouch or silo (silastic reinforced with dacron material; or if not available dacron mesh material; or if not available dacron mesh is applied onto the sticky side of any surgical drape which acts as a smooth internal lining). This is undertaken as an emergency operation.

--> The body and sac of the patient are prepared as a surgical field. The patient must be kept dry or else hypothermia will be impossible to prevent.

--> The sac is excised and the contents of the peritoneal cavity inspected. The liver which is central in position, will be found to be anatomically abnormal in shape and form and poorly supported by its very flimsy ligamentous attachments. Careful dissection is required to separate the sac from its surface where it is closely adherent. Note the very anterior position of the inferior vena cava at the level of the diaphragm and the easily definable, elongated, hepatic veins, on which the now mobilised liver hangs. The reason behind staging the reduction, other than a simple discrepancy in size becomes obvious. Any attempt to force the liver into the peritoneal cavity behind the right costal margin will impede its venous return by kinking the hepatic veins. Although this may not occur every time it is unpredictable and the acute portal hypervolaemia that results has potentially lethal implications. In gastroschisis the liver is already within the abdominal cavity and poses no problem.

--> An attempt is made to stretch the abdominal wall and a prosthetic bag is tailored to the indicated size. As its contents are sequentially reduced into the peritoneal cavity, the base of the pouch is made wider than its apex. It is attached to the peritoneum and musculofascial layer by interrupted nonabsorbable sutures after mobilising the skin edges. The skin should cover this suture line.

In spite of the inevitable presence of infection, the pouch should remain firmly attached for 10 days to 14 days by which time surgical closure of the abdominal wall is possible.

By applying pressure to the apex of the pouch, reduction of its contents at a "physiological" rate is started as soon as the patient's general state has stabilized. The sooner the prosthesis can be removed, the better.

### **Non-Operative Treatment**

Non-operative treatment for the unruptured liver containing exomphalos is feasible and may be indicated when ICU facilities are not available. Death of the sac commences at the moment of birth as it has no blood supply. If allowed to dry out it desiccates and loses its tensile strength.

The repeated application of a chemical escharotic, killing and welding the surface layer cells together produces a strong protective scab, toughening the sac wall and preventing its rupture. As the sac cicatrises, its volume increases, reducing its contents into the peritoneal cavity that is gradually increasing in capacity. A large ventral hernia forms which, when the sac remnant is shed, leaves an umbilical ulcer that can be skingrafted.

*Numerous complications of this technique are described:*

- > Rupture
- > Sepsis
- > Poisoning: Mercurochrome in alcohol is the most effective escharotic (0.5% in 65%).

Nutritional deprivation secondary to a prolonged period of intestinal ileus.

This form of therapy may still have a small place in the treatment of exomphalos, but should only be embarked upon by those with experience in its use.

## **The Intestinal Tract in the Patient With an Exomphalos or Gastroschisis**

### **Anatomical Anomalies**

--> Vitelline duct remnants, usually a Meckel's diverticulum, are common. There are managed according to individual circumstances.

--> As part of the midgut loop lies outside the anterior abdominal wall, the normal process of orderly intestinal return and fixation has not occurred. Nonrotation of the midgut loop, the presence of a universal non-fixed mesentery, and incomplete rotation of the midgut are variants commonly encountered that require no specific treatment.

--> An intra-abdominal testis, often on its own mesentery, if no contraindications are present, should be guided through the inguinal canal and fixed in its respective scrotal pouch.

### **Acquired Intestinal Pathology**

--> Partial loss of the midgut loop due to strangulation is seen in gastroschisis and could be lethal.

--> A small or large bowel atresia, may be identified in these cases. The primary surgical procedure is unaffected by this finding which is corrected electively at a later date.

--> A prolonged period of ileus follows the primary operation in most cases and complete intravenous feeding techniques have revolutionised the treatment. Return of intestinal peristalsis can be expected by the end of the third postoperative week.

## **Complications**

Septicaemia could complicate any stage of the treatment. Strict asepsis and the use of prophylactic antibiotics do control its occurrence. TPN catheter-related sepsis and pulmonary infection caused by aspiration are ever-present threats that must be actively guarded against.

## **Prognosis**

Recognized criteria (risk factors) determine the outcome. The standard at which tertiary care is practised is of more importance than the technical capabilities of the surgeon.

No case of gastroschisis with an intact midgut loop should succumb and the long-term prognosis in these patients is excellent.

## **Comment**

### **Exomphalos/Gastroschisis**

**J R H Becker**

A very important factor in the prognosis of these babies is that the emergency treatment by the referring doctor should be optimal. Instructions should be conveyed to him by telephone before the baby is transferred so that the baby arrives in as optimal a condition as possible.

What the referring doctor must remember:

--> IV line

Because the intestines are exposed, there is abnormal loss of fluid and the baby can easily go into shock. An IV line is therefore vital.

--> Management of the exposed bowel

I do not agree that the whole baby should be placed in a plastic bag because this is impractical, dangerous and may interfere with the IV infusion. Only the exposed bowel should be placed in a plastic bag. The referring doctor should not use loose sheets of plastic and never wet saline swabs, neither of which protects the bowel from contamination, dehydration and hypothermia.

The bag should be translucent so that the bowel is visible throughout the journey to facilitate the monitoring of perfusion. It is easy to keep the bag in position and the baby can be kept warm while the bag will prevent dehydration and contamination of the bowel and the baby will not become hypothermic or dehydrated.

--> The accompanying person

The referring doctor must ensure that the baby is accompanied by a capable person who will efficiently manage any emergency which might arise and who will be able to monitor the baby *en route*.

### **Comment**

### **Exomphalos/Gastroschisis**

**J H Louw**

When a chemical escharotic is occasionally used, we prefer 1% *aqueous* solution of mercurochrome which is absorbed more slowly, and apply it only on day one.

## **Chapter 15.4: Congenital Intestinal Obstruction**

**M R Q Davies**

### **Introduction**

The baby born with an intestinal obstruction has a chronic lesion as the problem in most instances has been present from weeks to months prior to birth. Secondary anatomical changes, common to all hollow visceral obstructions, are well established. These factors make the diagnosis, in these cases, simple and straightforward.

### **Foetal Intestinal Obstruction**

During the second and third trimesters the foetus swallows ever-increasing volumes of liquor. At term 500 mL of this fluid is ingested per 24 hour period. A high complete small bowel obstruction is associated with polyhydramnios and intrauterine vomiting of bile containing gastric content. Foetal sonography will clearly delineate the grossly dilated proximal intestinal tract. The low small or large bowel obstructions are less likely to be detected, as the foetus normally does not defecate *in utero*.

### **Neonatal Intestinal Obstruction**

Dilated loops of bowel may be detected at birth. Immediate intubation of the stomach and evaluation of its contents may suggest an intestinal obstruction if the visible loops disappear or 15 mL or more of fluid is obtained. The aspirate is replaced by injecting an equivalent volume of air and a single erect x-ray taken. This will establish or refute the diagnosis but not exclude it in all situations.

### **Symptoms and Signs Common to All Types in This Age Group**

**Distension:** Noted even in high bowel obstructions due to secondary dilatation of the obstructed segment. Most severe in the lower large bowel forms.

**Vomiting:** Persistent emesis of significant volumes of bile-stained material characterize this symptom. Over 80% of all causes of neonatal intestinal obstruction, occlude the bowel lumen distal to the ampulla of Vater. For this reason bilious vomiting always requires urgent investigation to elucidate its cause.

**Inadequate or abnormal colonic evacuation:** At term the colon is a reservoir filled with meconium that is ready to be evacuated. Normally, this commences within the first 24 hours after birth.

### **Confirming the Diagnosis**

Physiologic aerophagy is present in neonates. Large volumes of air are swallowed so that it can be normally easily visualised within the lumen of both small and large bowel. For this reason it is not possible to distinguish, with any degree of absolute certainty, small and large bowel loops from one another on a plain x-ray study.

--> Gas is normally seen within the rectum in the pelvis 6 to 8 hours after delivery. Its presence does not exclude an incomplete obstruction. Over-vigorous rectal examination is a described but unlikely explanation for visible rectal gas, distal to a complete proximal obstruction.

--> Gas is used as a contrast agent proximal to the site of a *complete obstruction*. Remember the system is filled with fluid at birth. Empty and fill with air so that the state of the proximal intestinal tract can be assessed.

--> In the presence of what appears to be a *complete intestinal obstruction* a carefully performed barium enema provides a wealth of further information and does not only help to distinguish the large bowel from the small bowel.

--> When an *incomplete small bowel* lesion is suspected on scout X-ray photographs, a carefully performed barium swallow and follow through, could add further information. Aspiration is an ever present threat.

### **Aetiology**

#### **List of causes in rank order - first 48 hours of life**

- > Jejuno-ileal atresias or stenosis
- > Duodenal atresias or stenosis
- > Congenital intestinal aganglionosis
- > Meconium ileus.

#### **Causes present at birth but often not detected until later**

- > Midgut volvulus
- > Congenital intestinal aganglionosis (CIA)
- > Intestinal stenoses.

*An adynamic ileus may present with features that make the distinction between it and a surgically correctable lesion very difficult.*

--> Intestinal obstruction can be an early feature of both intra-abdominal and extra-abdominal sepsis in the neonate. The lack of hard clinical signs in a baby with an established life-threatening septicaemia compounds this diagnostic quandary.

--> Necrotising enterocolitis also features in the differential diagnosis.

In both of these conditions, the ileus is usually incomplete.

### **Jejuno-Ileal Atresias and Stenoses**

The patient is often preterm and may be small for gestational age (SGA). In the high atresia the presentation is classic and within hours of birth, while the low ileal atresia or rare single sclerosis may be difficult to be certain about both on clinical and radiological grounds.

The patient does not have any other demonstrable extra-abdominal congenital anomaly.

Scout films (erect, supine, prone) with air in proximal bowel show dilatation of more intestine than just duodenum.

*A contrast enema is ordered to:*

- > Demonstrate a patent colon lumen
- > Demonstrate its calibre and contents
- > Detect the presence of a rotational anomaly and to define small bowel from large bowel.

Meconium is formed within the small bowel and stored in the colon until birth. Normally, this fills and stretches the large bowel, giving it its normal size. An early complete small bowel obstruction prevents this happening. The colon is small in calibre (the unused colon incorrectly termed microcolon) and its lumen poorly filled by content.

### **Pathology**

This group of atresia/stenosis have been shown to be due to intestinal strangulation in the foetus, *caused by:*

- > Volvulus
- > Intussusception
- > Placental emboli
- > Hernias (internal and external).

The duration of prenatal intestinal obstruction will vary as will the amount of meconium found distal to it. It is possible in many instances to demonstrate the cause of the intestinal ischaemia, like a rotational anomaly or remnant of an intussusception.

Fibrocystic disease is a well-known underlying precipitating factors.

*The following morphological forms are recognized:*

Type I Atresia	Septum
Type II Atresia	Cord
Type III Atresia.	Gap

In 10% to 15% of cases multiple atresias are found, located usually in the more distal small bowel, but rarely in the colon.

In Type III atresia the intestine is discontinuous and a large mesenteric defect is found. This indicates the disappearance of a significant length of bowel, which may directly influence the long-term prognosis. The term baby has between 250-350 cm of bowel from the pylorus to the ileocaecal valve. Intestinal adaptation, adequate for maintaining body weight and growth, is seen when up to 2/3 of this length is lost - leaving  $\pm$  75 cm of bowel intact. *It is pertinent to point out that:*

--> Bowel proximal to the obstruction is already stretched, partially explaining why unused distal bowel shows a greater ability to adapt centimetre for centimetre. The ileum always outperforms the jejunum and it contains specialised receptor sites (bile salts, vit B12).

--> Recovery is always less than expected if the ileocaecal valve is resected.

In spite of many reports of remarkable recoveries since the advent of effective parenteral feeding techniques, these patients require many months of in-hospital care, and most live a precarious brittle existence.

When the superior mesenteric artery (SMA) is occluded near its origin, most of the mesentery of the small intestine sloughs, but in the foetus the ascending colon and distal ileum can survive on a collateral arterial flow, originating from either the right colic or middle colic vessels. Due to an absent or grossly deficient mesentery the residual distal small bowel spirals around its blood vessel attachments in the characteristic fashion of an apple-peel - the so-called *apple-peel atresia*.

### **Treatment**

The repair is performed as an elective emergency. Gangrene of the grossly distended proximal segment of bowel due to either tension or more likely torsion, can occur.

Aspiration must be prevented at all costs.



*Through an upper transverse laparotomy incision the diagnosis is confirmed and the whole of the intestinal tract inspected:*

--> The bowel proximal to the obstruction is emptied.

--> The bowel immediately distal to this site is opened and saline injected to establish the patency of its lumen. If a contrast enema has been performed, barium in the caecum excludes a colonic lesion. Previously undetected atresias or stenoses are carefully marked.

--> Measure the length of the bowel. Make every effort to save segments where length is a critical factor.

--> Distal obstructing lesions are excised and axial end-to-end anastomoses using PDS, prolene or arterial silk-interrupted sutures are constructed.

--> When it can be sacrificed, the distal bulbous end of the obstructed bowel is resected using the GIA stapling instrument applied obliquely across the bowel axis, so that a simple end-to-end anastomosis can be performed. Where this instrument is not available, tapering the proximal bowel lumen to match the size of the distal bowel is preferred to the classic end-to-back anastomosis.

--> In proximal jejunal atresias a transanastomotic silastic feeding catheter is obligatory. This exits through a tube gastrostomy.

--> Reconstituting the mesentery may be difficult where it is deficient. Concentrating the bowel on the jejunostomy catheter to release the tension on the mesentery, helps to enable closure to be performed. Stabilising the bowel is essential in apple-peel type lesions.

### **Post-Operative Management**

The nutritional care of the patient is most important when a marginal length of bowel is present (less than 100 cm).

### **The Three Phases of Enteral Adaptation**

--> Intolerance to any enteral intake.

--> Partial adaptation - 3 to 6 months post op.

--> Maximal adaptation -  $\pm$  12 months following surgery.

- Complete parenteral nutrition is the main source of caloric intake for the early post-operative recovery phase.

- From day 5, jejunostomy feeding is started.

- 1/2 DD solution once tolerated is changed to 1/2 strength low lactose formula feeds which are gradually increased until full-strength milk feeds are given by day 14. The volume given which is best infused continuously (syringe pump) is gradually increased to

tolerance (the trophic stimulus).

- Prevent enteral overfeeding - it will only cause diarrhoea and fluid and electrolyte deficits.

- The parenteral route will be in use to supplement enteral feeding for weeks to months - preferably use a central venous conduit.

- The operations that have been devised to correct the short bowel syndrome (retarding intestinal transit; recycling of chyme; lengthening the bowel) have not been of any demonstrable benefit.

- H2 receptor blockade, cholestyramine and loperamide have a place in the care of these cases.

### **Duodenal Atresia and Stenosis**

The lesions that are found in the first and second parts of the duodenum are true malformations. The most common site is at the level of the ampulla.

#### **Associated Congenital Anomalies are Common:**

- > Down's syndrome

- > Congenital cardiac anomalies

- > Oesophageal atresia or intestinal rotational anomalies.

A positive family history may be obtained.

The pregnancy is often complicated by polyhydramnios and the diagnosis thus suspected prenatally. Intra-abdominal cyst-like lesions are seen sonographically. Where the kidneys can be visualised and renal pathology excluded in early pregnancy, amniocentesis (chorionic villus biopsy) should be ordered so that a karyotype can be obtained. If appropriate, termination may be indicated where a trisomy is present.

At birth the patient is again preterm and may be SGA. The chronically dilated stomach and duodenum may be visible in the upper abdomen.

In supra-ampullary obstructions nonbilious vomiting occurs. In all other types bile is found in the stomach which characterises these obstructions. The patient, just to confuse the unsuspecting, may pass normally appearing volumes of meconium, where bile has entered the bowel distal to the obstruction.

- > An erect plain x-ray film shows the characteristic double bubble appearance. No gas is seen in the distal bowel when an atresia is present. A distal contrast enema is not indicated as associated small or large bowel atresias are rare.

In duodenal stenosis varying amounts of gas pass into the distal intestine. In the presence of a significant degree of intrinsic stenosis, the obstruction is chronic and the duodenum is dilated. This helps to distinguish this form of duodenal obstruction from the

extrinsic variety.

When an extrinsic obstructing cause cannot be ruled out a contrast study to exclude the presence of a volvulus is obligatory. Where a rotational anomaly is identified an emergency laparotomy is indicated.

### **Associated Anatomical Anomalies**

--> A preduodenal portal vein may accompany the supra-ampullary atresia. The extrinsic pressure of this structure is not the cause of the obstruction, it is only an anatomical curiosity.

--> The atresia may be of any type - I, II or III.

--> The pancreas at the site of an ampullary lesion may partially or more rarely completely surround the duodenum simulating an annular pancreas.

--> The bile duct is bifid when the atresia is at the level of the ampulla. Bile is found in the lumen of the bowel proximal and distal to the obstruction.

--> In duodenal stenosis the lesion may be simple or a fenestrated membrane may be found. This membrane due to peristaltic effort can lengthen, so that it assumes the shape of a wind sock. The bowel distal to the obstruction is dilated by it which could mislead the operator into performing an incorrectly sited anastomosis. Furthermore, in most of these membranes the bile duct or ductules traverse its medial part to open on the edge of the opening in it. Excision of this area with or without suture closure could obstruct the flow of bile.

--> In a significant proportion of cases the presence of a megaduodenum caused by the intrinsic obstruction prevents the normal return of the midgut loop during intestinal rotation. Incomplete rotation of the colon is common in this group.

### **Treatment**

The operation is an elective emergency. Through a right-sided supraumbilical transverse laparotomy incision, the C loop of the duodenum is exposed.

--> The non-obstructed bowel is dissected off the dilated duodenum until the site of the lesion is identified.

--> A side-to-side duodeno-duodenostomy can usually be performed.

--> A transanastomotic feeding tube, carefully tethered by a stitch at the anastomotic site is always used and is left to lie in the upper jejunum.

--> An atresia or stenosis in the third or fourth part of the duodenum could be due to prenatal infarction and may, therefore, be associated with a downstream atresia.

--> Tapering duodenoplasties are used on the obstructed segment where possible to

shorten the period of duodenal ileus which normally persists for 21 days.

### **Congenital Intestinal Aganglionosis**

This is a microscopic anatomical anomaly which affects the nerve supply of the bowel. Absence of ganglia establishes this diagnosis. Histochemical staining techniques have uncovered an ever-increasing number of new anatomical anomalies termed the neuronal intestinal dysplasias - anomalies that in most cases have yet to be married with any certainty to clear-cut clinical entities.

#### **The Length of the Aganglionic Segment**

- > Rectum and most distal sigmoid colon: 70%
- > To descending colon at splenic flexure: 15%
- > + Transverse colon and ascending colon: 10%
- > + Small intestine: 5%.

The clinical presentation is influenced by the length of the anomaly. The bowel immediately proximal to the aganglionic segment, the transitional zone, is also functionally abnormal. In essence there is a derangement or absence of descending inhibition-producing bowel relaxation or a zone of reduced intraluminal pressure, that immediately precedes the peristaltic wave. Incoordinated nonpropagating contractions occur with an increase in mural tone over the whole affected segment. The cause of the anomaly is not yet known.

The patient is classically a full-term male baby the product of an uncomplicated pregnancy.

Excluding one of a twin, this anomaly is not seen in SGA babies. Excluding Down's syndrome, it is not associated with other congenital anomalies. It does, however, occur in families.

Declared to be physically normal in every respect, the patient soon shows clinical signs of intestinal obstruction. No patient with CIA has normal bowel function. Hard clinical signs are often not found. As the most distal bowel is affected, the functional deficit declares itself at this level.

#### **The Sign of the First Meconium Stool**

Normal babies pass meconium during the first day of life.

- > The onset of stooling is delayed.
- > The amount evacuated is always inadequate and abdominal distension occurs.
- > The delay in transit has three important immediate effects.

In the colon, meconium ileus becomes inspissated and meconium constipation follows. This meconium can be seen on a contrast enema as long filling defects which when evacuated, are called meconium plugs. CIA is just one cause of the **stool inspissation syndrome**.

Stasis impedes the natural clearing of bacteria and severe enterocolitis, which is the cause of death when CIA is inappropriately treated, occurs. The ileus that is simply due to obstruction in the beginning, becomes complicated simulating that encountered in a septicemic patient. *Clostridium difficile* has been implicated; pneumatosis intestinalis is seldom seen.

Distal obstruction can cause proximal bowel perforation, usually in the caecum. Appendicular perforations, which are called appendicitis, occur and are also found in total colonic aganglionosis.

Mechanical manoeuvres designed to induce the evacuation of the distal bowel content may be effective but the result is, of course, transitory and will only prolong the period of misdiagnosis. A complete obstruction is changed into an incomplete obstruction, and *the patient develops secondary pathological changes:*

--> The bowel proximal to the abnormal segment undergoes hypertrophy and dilatation.

--> Abdominal distension becomes marked and the chronic intestinal dysfunction causes failure to thrive.

--> Symptoms of hypercontinence are elicited:

- Alternating episodes of diarrhoea and constipation.
- No perianal soiling.
- Difficulty in passing solid motion and also wind and water.

### **Diagnosis**

The presence of a low complete or incomplete bowel obstruction are confirmed on straight abdominal x-ray. Air fluid levels are seen; gas is usually not present in the rectum.

Any form of rectal examination must be avoided until after a contrast enema has been performed.

Under screening, dilute barium is run into the rectum via a catheter in a manner that prevents any distension. Its course is followed until the most distal part of the obstructed proximal bowel is clearly visualized. This establishes the diagnosis. At this point, if constipated meconium is identified the barium is changed for gastrografin and the study becomes a therapeutic enema, which usually successfully decompresses the patient. If no obvious transition is seen, and barium has filled the colon without identifying any pathology, the examination is terminated. Delayed films are taken 6 hours, 12 hours and 24 hours later. The neonate defecates automatically and delayed emptying will be shown. As filling pressures have been allowed to equalise, classic radiological signs may be unmasked. This study has an over 50% accuracy rate. *These radiological signs are not present where:*

- > Any mechanical method to induce stooling has been used.
- > Enterocolitis is present.

### **Treatment**

If the study is diagnostic or suggestive of CIA, treatment of the obstruction is started. This is urgent if diarrhoea (enterocolitis) is present.

A digital dilatation of the anal sphincter and saline washouts using two catheters, of the lower sigmoid colon are done. Satisfactory decompression is usually obtained and steps are then taken to confirm the diagnosis. An absent rectosphincteric reflex is classically seen in CIA. If a normal response is obtained, which is easily reproduceable, CIA can usually be excluded.

The final diagnosis is made on a rectal biopsy specimen.

### **Obtaining the Tissue**

The best developed plexus lies in the intermuscular plain - the plexus of Auerbach. A full thickness specimen is required. Under general anaesthesia a biopsy is taken 2 cm above the dentate line, taking care that the correct muscle tissue is obtained.

Suction biopsy forceps have been designed so that submucosal tissue specimens can be obtained at the bedside. The plexus evaluated by the pathologist in these instances is Meissner's plexus, the deep submucosal plexus. It contains fewer and less well developed ganglia which can be very much more difficult to evaluate. This has been simplified by the development of new staining techniques.

*The following biopsies are done:*

--> **A high biopsy** - mid rectum - on which the anatomy of the plexus is assessed.

--> **A low biopsy** - just above the dentate line - on which the abnormal proliferation of nerves (neuromatosis) a constant finding in aganglionosis, is best visualised by using non-specific cholinesterase, neuron specific enolase, and other staining methods.

On confirmation of the diagnosis an electively performed near enterostomy is constructed (just proximal to the aganglionic segment). Histologic confirmation that the stoma has been raised in ganglionated bowel is essential.

According to the surgeon's preference, a pull-through operation is carried out between six to 12 months after birth.

### **Meconium Ileus**

Quantitatively abnormal meconium obturates the terminal small bowel producing an obstruction early in foetal life. Small bowel secretions do not enter the colon. The small bowel gradually fills up with meconium. At birth a well-established small bowel

obstruction is present. The patient is distended, visible loops of bowel are seen and may be palpable. No stool is passed and on rectal examination the bowel does not contain normal meconium.

### **Aetiology**

Nearly all are due to fibrocystic disease - 10% to 15% of fibrocystics are born with meconium ileus. In 50% of these cases the bowel obstruction is complicated usually by a volvulus due to the torsion of a heavy laden dilated loop of small bowel. Intestinal atresias of prenatal perforation and meconium peritonitis also occur.

In rare instances this type of obstruction is due to the intra-uterine absence of exocrine pancreatic secretion (congenital pancreatic duct obstruction, chronic destructive pancreatitis of congenital syphilis).

Fibrocystic disease is an autosomal recessive disorder. It occurs in people of European descent and is not seen in blacks. In Europe this form of congenital obstruction is very common.

### **Diagnosis**

Scout films confirm what is found on clinical assessment.

--> Grossly distended bowel, filled with air without obvious air/fluid level formation and filled with material with a ground glass appearance which may have trapped gas in areas.

--> The picture is not unlike that seen in low ileal atresias - more solid content is noted. Signs of meconium peritonitis may be present.

--> Barium enema reveals that the colon is unused and is normally rotated. The never-filled "foetal" colon is surprisingly small and is classic of this condition. Half-strength gastrografen is used when this diagnosis is suspected and an attempt is made to reflux the contrast into the terminal ileum. This confirms the diagnosis when pellet-like filling defects are shown and may have a therapeutic effect where the contrast has flowed well into the small bowel. Care must be taken however as this may cause the rapid accumulation of fluid within the bowel lumen with life-threatening third space fluid loss.

### **Treatment**

Transmural necrosis is a significant threat when distension is present. For this reason urgent decompression is required. The therapeutic gastrografen enema should only be carried out by an experienced paediatric radiologist. It is seldom successful when a well-established obstruction is present, but after preparation of the patient and in the absence of clinical evidence of a complication an attempt should be made to overcome the obstruction using this method. Unless success is immediately apparent, it should be abandoned and the patient operated upon.

A supraumbilical transverse laparotomy is performed. The bowel has a classic appearance. Mobile pellets can be felt in the distal ileum proximal to the ileocaecal valve. Above this the bowel is distended by putty-like meconium, and proximally the lumen is filled with fluid and gas.

It is possible to disimpact the meconium by introducing a catheter into the bowel distal to the site of the occlusion. Saline or saline with acetyl cysteine 20% (Parvolex) irrigation is used. This tiresome task is continued until the meconium has been completely freed from the bowel wall and can be milked up and down into the colon. Transmural needle injection of fluid into the meconium at multiple sites has been used, but is not advised. At the completion of the manoeuvre a T tube can be left in the bowel through which further irrigation can be done postoperatively. If the downstream obstruction cannot be cleared a chimneyed ileostomy is established. This is also done when bowel resection is indicated for gangrene. The unnecessary removal of bowel is discouraged, as all of these cases have malassimilation.

The chimneyed ileostomy (Bishop-Koop) is fashioned as follows: the proximal end of the bowel is anastomosed end-to-side to the distal bowel 3 cm from its end which is raised as a stoma on the skin. This acts as a vent until the obstruction is overcome. This is formally closed electively once the patient is stabilized and growing on enteral feeds.

The long-term nutritional care of these patients should be carried out by a paediatric gastroenterologist. Suspected underlying fibrocystic disease is confirmed by a sweat test and lifelong management is necessary. Although the prognosis has improved, long-term outlook remains guarded.

### **Comment**

## **Congenital Intestinal Obstruction**

### **J H Louw**

Type III atresia could be further subdivided into:

Type IIIa - simple blind ends

Type IIIb - apple-peel type

Type IV atresia - multiple atresia/stenosis - all types could also be added.

The presence of an intact ileocaecal valve after resection of obstructed bowel is very important. Currently, a residual intact bowel of  $\pm$  25 cm to 30 cm with the ileocaecal valve and 50 cm without valve seem to be adequate for maintaining body weight and growth.



## Comment

### Congenital Intestinal Obstruction

**J R H Becker**

It is essential that the operator should always test for further atresias in the rest of the bowel. It is commonly recommended that saline should be used for this purpose, but I use air insufflation because the low viscosity of the air allows rapid transit into the rest of the bowel and the air escapes readily from the bowel when the abdomen is closed. I have found it more helpful than saline.

### Chapter 15.5: Midgut Volvulus

**M R Q Davies**

#### Duodenal Obstructions

In early life these are classified into two groups:

**Intrinsic**     - atresia  
                  - stenosis

**Extrinsic**     - volvulus  
                  - bands.

Mixed forms occur and both may need to be corrected in early life it is assumed that when an annular pancreas is present that an intrinsic duodenal stenosis is the primary pathology and any associated anatomical anomaly of the pancreas is of secondary nature. Emphasis is placed on the role "bands" play as a cause of extrinsic duodenal compression in the case with an intestinal rotational anomaly. Practical experience has shown that bands on their own seldom if ever, cause a significant degree of duodenal ileus in the child. Abnormal rotation of the bowel around the superior mesenteric vascular axis gives rise to the commonest type of extrinsic duodenal compression. As strangulation may accompany the volvulus, the immediate detection and correction of the lesion is of paramount importance. The neonate with a midgut volvulus has the most dangerous form of intestinal obstruction.

--> The volvulus is congenital, ie, it is present at birth.

--> The duodenal obstruction is incomplete and can vary in severity.

--> Delay in the development of the presenting features of an obstruction occurs, as oral maintenance fluid volumes are small during the first 2 to 3 days of life.

Gangrene of the whole of the jejunum and ileum can occur and is a hopeless situation.

## Anatomy

Normally the mesentery of the small bowel is attached posteriorly between the ligament of Treitz and the caecum in the right iliac fossa. Intestinal peristalsis cannot create rotational forces around this as a fulcrum due to its length. Anomalies involving the orientation of the intestines, may alter this by shortening the attachment. The midgut loop enters the umbilical coelom prior to the sixth week of intrauterine life. Orderly return takes place with disappearance of the physiological hernia by the twelfth week. This movement is known as the rotation of the intestines and is arbitrarily divided into 3 stages.

### Stage I: Extrusion

The midgut loop finds in the base of the umbilical cord.

### Stage II: The Return in the Peritoneal Cavity

As this occurs, rotation in an anticlockwise direction takes place within the umbilical sac and during reduction. The caecum is the last portion of bowel to return and at completion of this stage, it lies beneath the liver in the right hypochondrium.

### Stage III: The Descent and Fixation of the Right Colon

At term this phase is still incomplete.

Complete rotation equals  $270^\circ$  in an anticlockwise direction.

*Only two abnormalities are of practical significance:*

--> **"Non"-rotation of the bowel:** Stage I and II completed. Only  $90^\circ$  rotation occurs so that the colon lies to the left of the midline and the small bowel on the right. The caecum lies in the left upper quadrant.

--> **Incomplete rotation of the bowel:** Only stage I and II completed with  $180^\circ$  rotation. The caecum lies above and to the right of the umbilicus beneath the liver where it may be tethered by wild attachments to the duodenum and retroperitoneum in the region of the right kidney (Ladd's bands). Partial fixation is present but the  $90^\circ$  rotation that is part of Stage III is absent.

In the absence of any fixation a universal midgut mesentery is present.

### The Duodenocolic Isthmus

This is the axis of the midgut mesentery around which torsion usually occurs. It is formed when at the two ends of the small intestine's mesentery, the caecum and the duodeno-jejunal junction overlie one another near the origin of the superior mesenteric artery. A duodenocolic isthmus is present in the patient with incomplete rotation of the bowel. Inadequate rotation of the duodenum is an accompanying defect in most cases.

## **The Torsion**

This takes place through 180° to 360° in a clockwise direction. It is thought to be due to normal peristaltic bowel movements and as enteral intake in the form of liquor amni commences from the third month of uterine life, it is not surprising that the volvulus usually occurs prenatally.

## **The Ischaemic Consequences of the Volvulus**

Prenatal intestinal gangrene with repair causes atresia or stenosis. Postnatal intestinal gangrene leads to loss of the midgut loop. Intrinsic duodenal obstructions may be associated with incomplete rotation. Incomplete duodenal intrinsic lesions, or a stenosis, that allow some degree of enteral intake can be complicated by a volvulus.

## **Clinical Presentation**

The greater majority present during the first 7 to 14 days after birth - the time when milk feeds are introduced.

These babies are reluctant to feed normally from the outset and as the volume of fluid intake increases the signs of high bowel obstruction appear:

--> Vomiting of clear or normal gastric content, later tinged with bile. This sign noted during this life period and later requires an urgent explanation (the deadly vomit).

--> The patient evacuates meconium and if the diagnosis is delayed, stools may contain blood.

--> In the presence of significant intestinal ischaemia, clinical anaemia and distension of the affected bowel by ingested air that cannot be passed or absorbed, will be noted.

In rare later presentations, where luminal obstruction to either the bowel or the blood vessels is not significant, the lymphatic return may be compromised causing malassimilation and producing diarrhoea. Chyle in the peritoneal cavity of the processus vaginalis, will guide the informed to its underlying cause (a chronic volvulus).

## **Diagnosis**

The obstruction is high and incomplete. Dilated bowel is not present, but there is delay in emptying of the stomach and the first part of the duodenum. Scout abdominal films are difficult to interpret and little abnormality is seen. A giveaway signal may be an unexpected air fluid level in the proximal duodenum.

*When this diagnosis is suspected a contrast study of the intestinal tract must be ordered.*

## **Barium Swallow and Follow Through**

The configuration of the duodenal C loop is visualized.

--> It is incompletely rotated.

--> Imperfect emptying is seen.

--> As the occlusion is incomplete the presence of a volvulus can be clearly demonstrated; the torsion produces a spiral lumen, which is classic of the lesion.

When the proximal jejunum has filled it is found misplaced to the right of the midline and is not situated in the left upper quadrant where it should be.

The study visualises the site of the obstruction and can demonstrate its cause.

## **Barium Enema**

The whole of the colon must be visualised. Incomplete filling cannot be ruled out unless the caecum can be positively identified when the appendix is seen.

The abnormal findings are:

--> An incompletely rotated colon.

--> The caecum is seen in the mid-abdomen above the umbilicus.

This study does not demonstrate the cause of the obstruction, but may be of more value than the upper GI examination which requires the services of an experienced paediatric radiologist.

## **Treatment**

An emergency laparotomy is carried out. The routine transverse incision is made which must be large enough to allow the whole of the small intestine to be eviscerated, without altering its position. It is immediately apparent that the transverse colon with its omentum cannot be identified in its expected position and careful inspection will demonstrate the volvulus.

*Three definitive steps, each with a separate objective, are required to correct this problem:*

### **Step I: Detorsion of the Volvulus**

Anticlockwise rotation of the midgut loop on its axis corrects the volvulus in nearly all cases.

### **Step II: A Ladd's procedure**

The small bowel's mesentery must be released by dividing all abnormal adhesions that prevent it from being splayed out. The caecum is mobilised off the duodenum and other retroperitoneal structures. Prevent damage to the mesenteric vessels. The distal

duodenum is released and straightened. All the bands that cross it are divided. At the completion of this part of the operation, the duodenum should run down inferiorly, parallel to the spine.

### **Step III: Look for an Intrinsic Duodenal Stenosis**

A fenestrated duodenal web may be easily overlooked.

A large N/G tube is advanced through the pylorus into the duodenum. Saline can be injected to help assess its patency.

Where doubt still remains, a No 10F Foley bag catheter is introduced through the wall of the gastric antrum and advanced through the duodenum. With the balloon inflated it is slowly withdrawn. This excludes the presence of a stenosis.

The appendix can be ligated and removed without stump inversion. The bowels are returned into the peritoneal cavity so that they lie in the position of non-rotation, ie, small bowel on the right, large bowel on the left with the caecum in the left upper quadrant. Suture pexy of the intestines is not indicated.

### **Comment**

#### **Midgut Volvulus**

#### **J H Louw**

While it is true that two thirds of midgut volvulus occurs in young neonates (7 to 14 days), it should be pointed out that 20% of the patients present later in the first year of life and a further 20% after the age of one year.

The pathological anatomy, clinical presentation and diagnosis are beautifully described, but the following should be added:

--> 1. Non-rotation or incomplete rotation of the midgut occurs in approximately 1:500 live births. In the first month of life the male to female ratio is 2:1.

--> 2. The onset of symptoms may be acute, chronic or recurrent and progression to infarction of the midgut may never occur or may be sudden and complete with a high mortality.

--> 3. Although pain is not a prominent feature in patients less than one year of age, it is a major presenting symptom in older children.

--> 4. Three clinical groups can be identified:

--> The neonate presenting as already described.

--> The older infant or child with a longer history and symptoms that are intermittent with stuttering attacks of colic and vomiting. Very often abdominal

examination is normal on each occasion.

--> A group where the previous history is more elusive and may even be absent. The child presents with the sudden onset of symptoms, severe vomiting, not always bile-stained at first, severe pain and diarrhoea. Diagnosis is initially delayed with rapid clinical deterioration.

## **Chapter 15.6: Anorectal Malformations**

**M R Q Davies**

### **Introduction**

In a population with a high birth rate these abnormalities are common as they are not immediately fatal and nearly always noted. In most cases the hindgut communicates with the exterior, albeit via another system. As with most ectopic openings, their calibre is inappropriate, so when the diagnosis is missed at birth, intestinal obstruction becomes the presenting feature.

The anal canal possesses numerous still poorly understood mechanisms affording its owner discriminatory powers over colonic evacuation, seldom present in those with a surgically manufactured copy. The best surgical results are the outcome of a correctly performed first repair. Redo or salvage procedures cannot succeed where the tissues used by the surgeon to construct a muscular sphincter, have been replaced by fibrosis. Failed surgery is usually the initiating event in the lifelong tragedy of noncompetitiveness solely due to anal incontinence.

### **Anatomy**

Concepts regarding the embryology and anatomy of the anal canal and rectum continue to change. The proposals made by Shafik are attractive and are used here.

### **Embryology**

The anal canal is a simple extension of the rectum which reaches the skin. The involuntary sphincter (internal anal sphincter) and the levator cutis ani (the coat tails) are downward modifications of its circular and longitudinal muscular layers. The hindgut is directed to the surface by the muscular pelvic floor (levator ani) which forms a complex canal - the levator tunnel, for this purpose. This structure forms the voluntary or external anal sphincter. Anomalies of the anal canal are presumed to be the direct results of abnormal levator tunnel formation.

As the end of the bowel nears the skin it induces the formation of the proctodeum. An anal membrane forms at the level of the pectinate line and forms a transitory occlusion. Anomalies involving both of these structures are encountered.

**Note:**

--> A visceral defect is associated with a parietal anomaly. An abnormal sacrum (on palpation; pelvic x-ray) indicates a poorly formed pelvic floor, and defective innervation when less than two sacral segments are present.

--> In spite of an absent anal canal, the muscles that would have formed the external sphincteric complex are present, though deficient. The skin in many cases shows changes consistent with those seen during the formation of the proctodeum.

**Anatomy**

To prevent confusion, the traditionally held surgical viewpoint that the rectum ends and the anal canal commences at the level of the levator hiatus (old puborectalis sling) is adhered to.

**Classification**

As the hindgut traverses the muscular pelvic floor it is surrounded by the external sphincteric complex. Where it reaches the proctodeum, the length of the anal canal and its sphincteric mechanism is normal. With shortening or anal dysgenesis, sphincteric deficiency exists.

Anorectal malformations are classified into two major groups:

**Suprlevator Lesions (High)**

--> Anal agenesis is present as the rectum ends above the levator.

--> The blind end of the rectum is connected to the internal genitalia in the female; the urinary tract in the male, by a fistula.

--> Where the rectum is dysgenetic this connection opens in the bladder.

--> The longer the gap between the end of the bowel and the skin, the greater the degree of dysplasia of the pelvic floor muscles.

**Infralevator Lesions (Low)**

--> Part of the anal canal is present (anal dysgenesis).

--> The end of the bowel is connected to the skin of the external genitalia (penis; scrotum; vulva) or perineum, by a fistula.

--> The shorter the gap between the end of the bowel and the skin, the more superficial the fistulous tract becomes.





abdominal distension that could compromise respiration, no urgent treatment is required. The patient is kept NPO and observed for a further 12 hours and then re-evaluated. At this point meconium may be found in the urine. If not, a special X-ray of the pelvis is ordered. A true lateral of the pelvis is taken with the hips flexed so that the ossific centres of the ischia overlap. It is important that the terminal hindgut is filled by gas (the contrast), clearly delineating it at this point:

--> The baby must be at least 6 hours old - time must be given for the swallowed air to reach that point.

--> Meconium impacted in the cul-de-sac may prevent gas from filling the distal bowel simulating a high lesion.

--> This study is of no diagnostic value where the gas-filled rectum can freely decompress itself through a fistula and therefore must not be ordered.

To encourage the gas to rise, babies were held upside down for several minutes before the picture was taken. This is a dangerous practice, for this reason a prone position with the caudal end of the subject elevated by flexing the hips to maintain this posture is now used.

The column of gas is related to the ischium. The most distal anteroinferior point on it is called Kelly's I point. If the gas ends at or above the I point, the lesion is radiologically high; if gas is noted well past this point the lesion is low. Gas is looked for in the bladder; if seen a high lesion is diagnosed.

An A-P view of the sacrum should also be done to exclude any degree of sacral agenesis which could later influence therapy, and the presence of an oesophageal atresia should be excluded clinically.

### **Female Cases**

Due to the interposition of the genitalia between the bowel and the bladder, the ectopic opening of the hindgut can be visualised in many female patients. The hymen (apex of the prostate in the male) divides the vestibule of the vulva from the vaginal canal. It also marks the level at which the vagina and any congenital intestinal fistula pass through the muscular pelvic floor. Armed with this knowledge, the external genitalia are closely examined.

### **The External Genitalia May Be Normal in Appearance**

The urethral and vaginal orifices can be seen. *Anywhere distal to the hymen in the midline the abnormal bowel opening may be detected:*

--> In the fossa navicularis, between the hymen posteriorly and the fourchette.

--> Outside the vulva on the skin surface, either as a fistulous tract (a covered anal canal or an anterior perineal anus).

Where the opening lies in the vulva, the perineum is absent. The skin behind the genitalia is featureless but the muscles that normally form the external anal sphincter are present. In low lesions (anovestibular fistula/vestibular anus) they are well-formed when exposed. The presence of a normal hymen signifies a low abnormality.

### **The External Genitalia are Abnormal**

Varying degrees of foreshortening occur and this involves the labia. The more significant this deformity, the more marked the associated hindgut developmental abnormality.

--> **An open vulva:** There is no labial adhesion and an open introitus is seen. The bowel opens above a deficient hymen in the vagina (anal agenesis with rectovaginal fistula).

--> **The closed vulva:** The labia minora are adherent so that a single opening is present beneath a well-formed clitoris (Cloacal anomaly of which there are different variants).

### **Summary**

Where the bowel opening cannot be seen in the female the lesion is automatically classified as high. X-ray studies are of no additional help.

### **Treatment**

The operation is an elective emergency *in the male*. The sooner the procedure is carried out after birth the less contaminated the large bowel is by bacteria.

### **In Low Lesions**

--> **Simple dilatation** of an inadequate but correctly positioned orifice may suffice (covered anus incomplete; incomplete anal membrane). This also applies to the anterior perineal anus.

--> **An anoplasty** - the excess skin overlying a low abnormality covering the anus is excised. Skinflap procedures are discouraged as is any form of plastic surgical repair. Perianal skin is tacked to the bowel wall. Sutures are purposely tied loose otherwise they will cut out as swelling occurs. The opening is dilated to a no 10 Hegar. If an intracutaneous fistula is present it is laid open (covered anal canal with or without fistula).

An exploration of the deeper tissues in the hope that the bowel may be encountered, a so-called perineal rummage, should never be performed. This can only damage and destroy those tissues that could be used during a correctly carried out procedure later. The diagnosis must be established preoperatively, surgery has no role in making it.

## In High Lesions

--> **A sigmoid colostomy** - a diverting colostomy is performed in the left lower quadrant of the abdomen. It is indicated in high lesions (with or without fistula) and when the precise nature of the anatomy of the abnormality is unclear.

*It is emphasized that:*

--> When a urinary fistula is present the faecal stream must be diverted. Loop colostomies may not be adequate.

--> The distal cul-de-sac must be carefully emptied of its meconium contents; this is best done immediately after the operation is completed while the patient is still anaesthetised. If this is neglected a faecaloma may form which could compromise a later procedure.

Both of these steps are taken to prevent uncontrollable contamination of the urinary tract via a distal fistula.

By placing the stoma in the sigmoid any urine that may enter the bowel via the fistula (recto-vesical) is rapidly evacuated, preventing reabsorption and possible hyperchloraemic acidosis.

*Stress is laid on the following technical points of this operation:*

--> A manhole approach: The wound must not be made too large. If the sigmoid is very distended it can be decompressed via a large needle before it is delivered through the wound.

--> The bowel is carefully sutured circumferentially to the musculofascial layers using nonabsorbable material; the bridge between the stomas is similarly constructed.

If the surgeon is inexperienced with this operation a right transverse colostomy which is much easier to perform is recommended.

In the female patient the initial steps taken in treatment are similar:

--> When the bowel opening is deep to the hymen or cannot be visualised (high lesion) a colostomy is indicated. It must be diverting as the definitive corrective procedure dictates it. The site at which it is raised, sigmoid or transverse colon is optional.

--> As in the male an anoplasty or dilatation, in rare cases, may be all that is required (covered anus; anterior perineal anus).

*--> The vestibular opening poses a problem as the diagnosis could be:*

- A high lesion (*rectovestibular fistula*) which is confirmed during the preoperative EUA when a long tract that cannot be dilated is encountered.

- A low lesion (*anovestibular fistula*) which is a short fistula and is simple to enlarge by dilatation.

A colostomy is indicated as the first step in the treatment of a rectovestibular fistula. In the anovestibular fistula lesions two different treatment pathways can be taken:

- Dilatation of the opening and tract with or without a minor plasty procedure (back cut). A "shot gun" appearance is formed which, although the vaginal and anal orifices tend to grow apart until adult life is reached, is cosmetically, and in most instances, functionally unacceptable. Constipation of an intractable type can complicate this treatment.

- Current opinion suggests that where possible the anovestibular lesion should be treated initially as a high lesion. The patient is given a colostomy and an early "anal" transposition is performed at three months.

### **Postoperative Management**

#### **Early**

--> Where an established orifice has been enlarged dilatations are ordered from the tenth day. These are continued with until an adequate compliant neo anus has been achieved. Ensure that adequate evacuations occur.

--> In the high lesion: place the patient on a low lactose-containing milk feed, to prevent diarrhoeal stools and encourage primary wound healing at the colostomy site. Before discharge exclude the presence of an obstructive uropathy sonographically.

Unless indicated earlier an excretory urogram (EUG) and voiding cystourethrogram (VCU) is ordered at three months.

Check urine regularly especially in male cases.

#### **Late**

--> A posterior sagittal anorectoplasty (PSARP) operation is done when the patient is 8 months or older. Do a distal loop-o-gram (water soluble contrast agent) before the operation. This establishes beyond doubt the type of anomaly that has to be dealt with.

On average the colostomy is closed three months later. This completes the surgery.

The anorectoplasty manufactures an anal canal; it does not establish continence. The child has to learn to cope with its defective continence mechanism. The results of these operations depend on the individual patient's co-operation and an ability to understand what is expected of them. These are functions that closely relate to the patient's age. Most of these unfortunate patients establish "social" continence in time.

## **Comment**

### **Anorectal Malformations**

**J H Louw**

The reasons for making a distinction between high and low lesions are:

--> Associated serious congenital abnormalities are uncommon in low lesions, but frequent in the high, ie, urogenital, vertebral (absent sacral segments) and digestive (oesophageal and duodenal atresia). When a diagnosis of a high anomaly is made, always pass a firm nasogastric tube to exclude oesophageal atresia and order a sonogram of the urinary tract and AP x-ray of the sacrum.

--> The initial treatment differs. In high anomalies a diverting colostomies (done as an elective emergency in males) should be performed as soon as possible. In low anomalies a simple perineal procedure (dilatation or anoplasty) is often sufficient.

--> The corrective procedure is complicated in high anomalies (anorectoplasty) with indifferent results in regard to continence, while in low anomalies it is relatively simple (at most, an early anal transposition) with good results in regard to continence.

Although it is true that an opening in the navicular fossa usually represents a low lesion (anovestibular fistula/vestibular anus) it may be high (rectovestibular fistula).

## **Comment**

### **Anorectal Malformations**

**J H R Becker**

The vital question that needs to be answered in the case of a newborn baby with an anorectal anomaly is "Is anomaly supralevator or infralevator ani?".

Supralevator anomalies will usually need a colostomy; infralevator anomalies can be corrected with an anoplasty except in the case of a female with the anovulvar fistula where the patient needs a colostomy and transposition of the anus later. The supralevator anomalies are corrected by means of the posterior sagittal anorectal plasty (PSARP operation). This operation is performed as soon as the baby is 5 kg in weight or three to four months of age. By the time the baby is six months old, the colostomy is closed and the new anus is dilated daily from then on until about one year of age.

One should attempt to conclude all operative procedures before six months of age and all anal dilatations by one year. The advantage is that there is a shorter period of time for colostomy problems to occur. Hospitalization and anal manipulations are completed at a stage in the child's development when psychological scarring is less likely.

## Chapter 15.7: Necrotizing Enterocolitis

M R Q Davies

### Introduction

*This disease occurs in two different groups of patients.*

#### The Newborn

The precipitating event may have occurred immediately prenatally but more usually is related to the birth process itself. It is detected within 48 hours of birth, where enteral intake has not been withheld and is by far the most common form of the disorder. Antepartum haemorrhage, maternal sepsis and birth asphyxia are the obstetrical complications associated with it.

#### The Older Baby

The precipitating event is usually severe enterocolitis and shock, caused by fluid loss, secondary to an infection with pathogenic organisms. The patient is usually less than three months old and often nutritionally or immunologically depleted. The disease should be distinguished from ulcerative enterocolitis of intestinal aganglions and pseudomembranous enterocolitis (*Staphylococci*, *C. difficile*).

Since the advent of neonatal care units this disease has been the most common surgical cause of an acute abdomen in the newborn.

The intestinal pathology seen covers the whole spectrum from a minimal and fully recoverable injury to total necrosis of the entire intra-abdominal tract, which is not correctable.

### Aetiology

This is multifactorial and not completely elucidated. In the neonatal form, nonocclusive mesenteric ischaemia is usually the primary insult. A low flow state affecting the mesenteric supply occurs, which has its greatest damaging influence on the mucosal lining of the intestinal tract. This pathology varies from a transitory period of breakdown in antibacterial defences, to a complete and irreversible destruction of the wall. *What happens from this point onwards depends on the presence of bacteria in the bowel.*

This is dependent on:

- whether the patient has been fed
- what type of bacteria are ingested (*Clostridium perfringens* gives rise to a particularly dangerous infection)
- whether substrate on which the bacteria can proliferate is available. This is usually carbohydrate in nature, which is converted into hydrogen that can be detected in

the patient's breath, or seen as pneumatosis intestinalis.

- An accompanying adynamic ileus with stasis of intraluminal contents, further increases bacterial proliferation.

Minor damage to the mucosa may recover rapidly and completely in the absence of bacteria, but in the patient carrying intraluminal potential pathogens, a rapidly fatal disease can occur.

At birth the newborn has literally no secretory immunoglobulin A (IgA) to coat the intestinal tract. Impaired perfusion further damages the ability of the lining layer to combat invasive bacterial infections. *The influence of both these factors is seen in the association between this disease and:*

--> An exchange transfusion performed via the umbilical vein.

--> Bottle feeds in contrast to breast feeding which contains secretory IgA and live macrophages.

--> Umbilical artery catheters.

--> Prematurity - this compounds all known risk factors.

--> Hypertonic feeds - hyperosmolar milk feeds damage the mucosal lining directly or secondarily due to fluid shifts.

*The order in which the three major aetiological factors play their part is:*

--> Mesenteric ischaemia.

--> *Bacteria.*

--> Intraluminal substrate.

--> Bacteria are of major importance in older patients.

This disease is distinguished from the other infective or ischaemic enterocolitides when visible gas is seen in the bowel wall (bacteria + carbohydrate).

### **Pathology**

*The intestinal mucosa bears the main brunt of the damage.*

--> Any part of the tract from the stomach to the anus can be affected, but the ileocaecal region is the most commonly involved.

--> The pathology is patchy in nature and under treatment certain areas will undergo recovery.

--> The resolution may not be complete. Fibrosis, with stricturing of the lumen is common.

*Depending on the stage of the pathology, any of the following may be encountered during a laparotomy:*

--> obviously recoverable lesions

--> obviously nonrecoverable lesions

--> simple sites of perforation with no other stigmata of enterocolitis

--> obviously damaged bowel but its ability to undergo resolution cannot be accurately determined.

The visible presence of pneumatosis intestinalis confirms the diagnosis. Portal bacteraemia may result in a "septic or pyogenic" hepatitis.

### **Clinical**

The patient with an ileus, which is usually incomplete, haematochezia and pneumatosis intestinalis seen roentgenographically, has enterocolitis. In the absence of pneumatosis, confirmation of the disease is not possible, although its presence can be suspected.

*Neonatal cases are divided into two groups:*

#### **"Primary" Enterocolitis**

Although the patient may fall into an "at risk" group, the subsequent development of the disease was not predicted.

#### **"Secondary" Enterocolitis**

The patient is recovering from or is still being treated for a life-threatening illness, when a progressive intestinal tract disease develops.

In both groups the problem could be unmasked by the institution of oral intake.

**A spectrum of clinical presentations occur:**

Type I: Suspected NEC

Type II: Confirmed NEC

Type III: Florid NEC.



## **Symptoms**

--> Ileus

Manifests as increasing gastric residuals or bilious vomiting, with abdominal distension and a failure to evacuate adequately.

--> An invasive bacterial illness

The patient is physiologically unstable (episodes of apnoea, thermolabile); most obvious in the neonate.

It is stressed that the pathology may be biphasic, ie, passes through phases of evolution and then recovery.

### **Minor Diffuse Mucosal Damage**

This precipitates a disaccharide deficiency with lactose intolerance and sugar malassimilation. There are loose, fluid stools, positive for reducing substances with low pH.

The hydrogen breath test is positive, as a result of bacterial fermentation. The above findings are accompanied by haematochezia.

### **Progressive Bacterial Enterocolitis**

This proceeds from an ileus via the translocation of bacteria, to peritonitis; from tenderness on abdominal wall palpation, to abdominal wall cellulitis, eventual septicaemia and a consumptive coagulopathy.

Transmural bowel necrosis presents with a palpable static loop of bowel, a palpable mass (sealed perforation or localised abscess). A free perforation presents with pneumoperitoneum.

### **Incomplete Resolution**

A stenosis (swelling) or stricturing (fibrosis) of the bowel lumen; occurs most often in the colon and presents as an incomplete obstruction with diarrhoea. Time helps to distinguish swelling (reversible) from fibrosis (irreversible), which occurs from the second month onward.

## **Diagnosis**

The clinical diagnosis is confirmed roentgenographically. Scout abdominal photographs show ileus with thickened loops of bowel; significant bowel wall damage is accompanied by a static loop, ascites and extraluminal air. The important feature is pneumatosis intestinalis. Gas is detected in the submucosa (bubbles) or deeper in the muscular or subserosal layers (linear streaks). Portal vein gas is best seen in the liver. The pneumatosis tends to be transitory so that the picture changes continuously. Gas in the

form of bubbles can be demonstrated within the veins of the liver sonographically, where it may not be seen on an X-ray film.

### **Treatment**

Once the diagnosis is suspected the gastrointestinal tract is rested. Confirmation is obtained by detecting hydrogen in the breath; blood in the stool and from positive X-ray studies. Any form of contrast radiology is contraindicated, unless a midgut volvulus must be excluded.

### **Nonoperative Management**

--> Cardiorespiratory care and support in an intensive care unit.

--> Aerobic and anaerobic antimicrobial intravenous drugs are given; oral agents can be added.

#### **Anything that facilitates intraluminal bacterial growth is strictly avoided:**

Enteral feeding causes metabolic stress for the bowel, provides substrate for growth and introduces new organisms. Decompressing the intestinal tract is essential. No oral feeding is allowed for a period of at least 10 days. Complete parenteral nutrition is given.

If the patient's general condition improves it does not necessarily reflect on what is happening within the abdominal cavity. The presence of necrotic bowel must be detected and operated upon before intestinal perforation with spillage of content occurs.

The patient is stabilized and monitored for a period of 12 hours. The abdomen in spite of the presence of significant pathology may settle and the period of observation is extended. The abdominal signs are regularly reassessed, repeated X-ray evaluation of the bowel by cross-table shoot through photographs is done six hourly.

Experience has shown that this is the best approach to these cases.

#### **An emergency laparotomy is necessary when:**

--> pneumoperitoneum is present

--> the patient's general condition, in spite of support, continues in a downward spiral.

If the patient with this disease is dying (non-resuscitable case) an operation performed as a last ditch stand is misspent effort as it is doomed to fail. Decompressing the peritoneal cavity at the bedside where a tense pneumoperitoneum compromises respiration can be a life-saving temporising measure.

#### **An elective laparotomy is necessary when:**

--> progressing pathology is demonstrated (increasing ileus; peritonitis; abdominal wall cellulitis)

--> a mass is palpable.

### **Operative Management**

The whole of the intestinal tract is exposed through a generous laparotomy incision.

**The principles applied are:**

#### **Questionable Non-Recoverable Bowel**

--> Short segment - resection

--> Extensive involvement - proximal defunctioning enterostomy, or second-look laparotomy after 24 hours.

#### **Perforation**

--> Exteriorization, but can be closed if a proximal defunctioning enterostomy is raised.

**Axial anastomoses are not done if:**

--> The general condition of the patient, the extraabdominal pathology, has not been satisfactorily stabilised.

--> Viability of the bowel ends is questionable (dead mucosal surface).

--> General peritonitis is present.

An enterostomy is constructed that can be closed once all preventable risks are eliminated and the patient is in positive nitrogen balance.

The pathology may appear to be patchy but is diffuse and ongoing if an emergency operation has been precipitated by an early perforation. The surgeon may be uncertain at the termination of an operation whether the procedure controlled the situation. Wide radical resections of bowel are inappropriate. Decompression and total rest combined with antibiotic control of the enterocolitis can save severely damaged intestine. Complete parenteral nutrition is given for a period of at least 10 days when the patient's general clinical state indicates it, graded enteral intake can be ordered.

#### **Prognosis**

The role of the intensivist and the ICU team determines the outcome, which in many institutions has reached a remarkable 80 per cent survival rate after surgery. The presence of free air under the diaphragm does not mean that the patient must be operated on immediately. Time is taken to prepare the patient for the procedure. In the presence of adequate tissue levels of oxygen and antibiotics to combat bacterial growth, with adequate urine flow and coagulation factors, the operation is proceeded with, with every expectation

of success.

### **Comment**

#### **Necrotizing Enterocolitis (NEC)**

**H R J Becker**

It is common practice that all babies with NEC who have been treated successfully non-operatively, receive contrast study of the intestinal tract before being discharged from hospital. We do not do this routinely on asymptomatic patients, because even if one does detect a stenosis, one is not necessarily going to react if it is totally asymptomatic. It is essential, though, to be on the lookout for symptoms in those patients who have had successful medical treatment. Special investigations such as a barium meal and follow through or a barium enema should be asked for when any abdominal symptoms develop.

During the medical management of necrotising enterocolitis, it is often very difficult to decide whether a patient needs a laparotomy. The signs and symptoms may be very suspicious but not conclusive. The pulse rate is a good guide under these circumstances. If the baby is adequately resuscitated, a sustained tachycardia out of all proportion to the degree of disease present, would sway the decision in favour of a laparotomy.

### **Chapter 15.8: Hypertrophic Pyloric Stenosis**

**M R Q Davies**

#### **Introduction**

Hypertrophic pyloric stenosis is a common paediatric condition which can be corrected surgically, but is never an emergency. As symptoms rarely present within hours of birth, the condition is correctly called infantile hypertrophic pyloric stenosis. The muscular hypertrophy can be congenital, but need not be so as it is well established that it can develop in a pylorus, macroscopically assessed as normal, shortly after birth. The now outdated, but once commonly practised, method of treating these cases nonoperatively is based on the fact that a degree of gastric outlet obstruction may in time be overcome with spontaneous correction of the disease. For this reason minor forms may not be diagnosed. The operation corrects the obstruction and is only indicated when the vomiting it causes is significant.

#### **Aetiology**

This is unknown. Abnormalities in the nerve supply of the sphincter, both of an anatomic and functional nature can be demonstrated. Spasm of the constricting muscle fibres prevents adequate gastric emptying, in time the pylorus becomes hypertrophied, hard and solid on palpation, and forms a constant narrowing that in exceptional instances may resolve.

## Occurrence

Male preponderance (5:1 - male to female ratio) with a prevalence of 1 per 500 in white races; occurs in families where the reason for this predisposition is not clear.

## Pathology

The sphincteric muscle (Torgersen) enlarges, narrowing and lengthening the pyloric canal. The obstruction results in a compensatory muscular hypertrophy, maximal in the antrum. The stomach undergoes dilatation and is filled with static contents. This causes a gastritis and oedema further compromises the pyloric opening.

*The gastric outlet obstruction leads to vomiting.* Reflux plus emesis causes oesophagitis and possible minor haematemesis. Acute and then chronic, caloric, fluid and electrolyte losses follow.

- The starvation will cause a failure to thrive.
- The fluid loss results in dehydration.
- The electrolyte loss causes a hypochloraemic alkalosis.

## Clinical

*There are three essential points in the diagnosis of this disease.*

- The age group

Day 7 to 8 weeks; average 4 weeks. Cases have been described outside this period, but are exceptional.

- *The main symptom is vomiting.*

It is never bile stained.

It is significant as it causes hunger.

The act is described as forceful.

- *The sign that confirms the diagnosis is a palpable and enlarged pylorus.*

## Diagnosis

*The presence of pyloric hypertrophy can be established in three ways.*

## Abdominal Palpation

This should be the only method of confirming the diagnosis. *The examination is a specific clinical "test" for which the patient must be prepared:*

- The stomach must be empty. Pass an N/G tube or assess immediately after an emesis.

- The anterior abdominal wall must be relaxed; a sweetened saline bottle, or breast feed will achieve this aim.

- The observer must be seated on the left of the patient, using the fingers of the left hand, the pylorus is searched for, beneath the liver, just at or to the right of the midline.

*Its features are:*

- Mobile
- Deep seated
- Well defined, right, upper and lower borders
- As an "olive".

If it cannot be felt, the feed is completed. Gastric peristalsis, seen as definite waves of contraction moving over the left hypochondrium and epigastric regions from the costal margin to the patient's right side are looked for. This sign is not diagnostic, but is suggestive and indicates that further studies are in order.

### **Radiology**

- Straight abdominal x-ray shows an air-filled and enlarged stomach with an inappropriate amount of fluid in it. The intestines appear empty and abnormally airless. Constipated stool may be noted.

- A contrast study of the GI tract. This is ordered to elucidate the cause of persistent vomiting in a baby. *The radiologist is specifically asked to:*

- *demonstrate pathologic gastro-oesophageal reflux*
- *detect pyloric stenosis.*

Vigorous peristaltic waves, a prolonged delay at the pylorus, which when filled by a trickle of contrast shows an elongated, bent canal and a double tract sign. Furthermore the canal is narrow and does not open up. Spasm is not an acceptable diagnosis. Finally the duodenal cap is seen.

Careful positioning of the patient is needed to obtain the correct views which may make this examination difficult for the inexperienced radiologist.

- If the pyloric region is normal *the duodenal C loop and the upper jejunum is visualised*, thereby establishing the possible presence of an extrinsic or intrinsic cause of incomplete duodenal obstruction.

### **Ultrasound**

This method should be used in preference to contrast radiology, where the expertise is available. The muscle mass can be measured and the hypertrophy diagnosed. Overall diameter is 9 in normal and 15 mm in pyloric stenosis, the muscle thickness 1.6 and 5,

and the length is 14 and 17, respectively.

### **Differential Diagnosis**

Conditions that normally fall within the domain of the paediatrician.

### **Feeding Difficulties**

An experienced mother who cannot rectify this in spite of support should alert the practitioner.

### **Foregut Dysmotility Complex**

Aerophagy, burping and regurgitation are part and parcel of the feeding process in early life. The point at which the physiologic becomes pathologic cannot be defined. Simple well-recognised changes to the feeding technique employed, is all that is needed in nearly all cases. Persistent vomiting is usually termed "chalazia" which is again simple to correct, once abnormal gastro-oesophageal reflux is diagnosed. Appropriate posturing and feed thickening is all that is required. Severe oesophago-gastro-antral dysmotility produces projectile vomits that necessitate the exclusion of pyloric stenosis.

### **Uncontrolled Vomiting**

*This is rarely the presenting feature in cases with:*

- adrenogenital syndrome
- CNS disorder
- renal disorder.

It may perpetuate neonatal jaundice. In these cases the vomiting could be due to pyloric stenosis and the hyperbilirubinaemia disappears on its correction.

### **Treatment**

*Preoperative treatment is usually indicated. The word treatment and not preparation is used here.*

- Dehydration

The deficit is corrected using 1/2 strength normal saline in dextrose.

- Hypochloraemic alkalosis

An anaesthetic can be safely administered when:

- the pH is below 7.5
- the chloride is greater than 90 mmol/L

- the standard bicarbonate is less than 30.

The alkalotic patient is hypokalaemic. In the presence of a normal urine output potassium is added to the fluids.

### **Gastric Retention**

Through a large-bore nasogastric tube the stomach is emptied and irrigated clean of any content with normal saline. The stomach is emptied immediately prior to induction of the anaesthetic.

### **The Operation**

A small towel is placed beneath the patient's back at the level of the lower thoracic spine.

### **Incision**

Transverse muscle cutting, midpoint between navel and xiphisternum, through right rectus muscle.

### **Delivering the Pylorus**

Find transverse colon plus greater omentum which will help to identify the greater curvature of the stomach.

### **The Myotomy**

The assistant helps to stabilize the muscle. Through the relatively avascular anterosuperior part an incision is made through the serosa of the pyloric ring to well on to the antrum: shallow at the duodenal end and deeper over the thickset part. The myotomy is completed by fracturing the muscle fibres with the blunt end of the scalpel handle or a MacDonald's dissector until the gastric mucosa is seen. The myotomy must break the most proximal pyloric channel loop of muscular sphincteric fibres. To assure this it must be extended well onto the normal-appearing antral wall.

Distally it supposedly should break the circular fibres completely. This is not possible unless the duodenum is routinely opened, as the operator has no absolute method of establishing this fact. It is preferable to stop short as a complete myotomy at this end is not essential.

The pyloric canal is then splayed open by stretching the edges of the myotomy with a blunt tipped artery forceps. Attention is focused on the most distal part of the pylorus where this manoeuvre is used to further fracture the most distal muscle.

Check that the duodenum has not been opened. Repair with simple sutures and omentum if need be. Haemostasis is secured and the wound is closed carefully with large bites of interrupted all-layer sutures. Wound dehiscence is inevitable where care is not taken.



## **Postoperative Course**

The nasogastric tube is removed on completion of the procedure where the mucosa has not been breached. Feeds are withheld for 12 hours. As the patient is nutritionally depleted and has no reserve stores, a 10% dextrose/electrolyte drip must be maintained if hypoglycaemic episodes are to be prevented. Graduated feeds are then introduced. Vomiting is not unexpected and the parents must be forewarned that it may occur. The next feed is missed and the problem often settles with posturing. In exceptional cases the use of prokinetic drugs may be indicated.

The patient is discharged once he/she is no longer drip dependent.

## **Chapter 15.9: The Acute Scrotum**

**M R Q Davies**

### **Introduction**

This is a surprisingly common paediatric surgical problem. An acute illness involving the scrotum is the presenting complaint.

From a management point of view these patients are best divided by age into 2 groups - infant cases, and preschool and schoolgoing children.

### **Infant Cases**

The mother's attention is drawn to the problem as her son has an obvious anomaly involving his external genitalia. The pathology is usually advanced and is distressingly tender to touch.

On examination it is essential to determine whether the problem lies:

- Within the linings of the scrotal pouch, or
- Involves the viscera, the contents of the pouch.

Careful palpation can determine this in most instances. Although primary pathology of the scrotal viscera can secondarily induce changes in the overlying coverings of the pouch, which could cause misdiagnosis, the reverse does not occur.

### **The Conditions that Arise Within the Structures of the Pouch**

- Bacterial infection of the skin due to:
- Cellulitis - the complications of a "nappy rash" may be localized mainly to the scrotum.
- The result of spread from an anal source which may be present as a scrotal problem.

In the young patient the nature and cause of the pathology is often not clear, hence the lesion is called *acute idiopathic scrotal oedema*.

- Swelling due to generalized oedema that manifests itself maximally in the scrotum (nephrosis, nephritis, allergy).
- Localized trauma or insect bite.

### **Examination**

The pathology is confined to the skin. Erythema and swelling are prominent signs although tenderness is not as severe as these manifestations would suggest. The contents of the pouch and the spermatic cord are not involved by the pathology.

In most cases the lesion is not strictly confined to a hemiscrotum as the changes usually cross the midline raphe and often extend to the groin and perineum.

General symptoms and signs are usually absent; the problem is a localized one.

### **Conditions that Involve the Testis, Epididymis and Spermatic Cord**

These are grouped into mechanical and non-mechanical categories.

#### **Mechanically Caused Lesions with Strangulation of Tissue**

##### **Torsion of the Spermatic Cord**

The point of rotation can be within or above the tunica vaginalis. Strangulation of the structures involved in the volvulus occurs.

- Extravaginal torsion of the spermatic cord

This is an uncommon form and is only possible during the perinatal period and very early life when testicular descent is taking place. Due to imperfect fixation to the subcutaneous tissues the testis, epididymis and tunica vaginalis are captured in the torsion.

- Intravaginal torsion of the spermatic cord

This is due to a congenital anatomical anomaly. The most distal portion of the cord and with it the epididymis are completely invested by the visceral layer of the tunica vaginalis. The scrotal viscera hang free within an abnormally capacious serosal sac like the clapper of a bell.

The abnormality usually involves both pouches and is the cause of the usual type of what is termed torsion of the testis. This variety of torsion also occurs in association with testicular maldescent.

### **The Complicated Indirect Inguinal Hernia**

Between 15% to 20% of indirect inguinal hernias in the patient under 3 months old are irreducible when first seen. Strangulation of tissue is always present as the visceral prolapse is held incarcerated by the external inguinal ring. Due to compression by this rigid fascial structure the blood supply to the contents of the sac and also the gonad in the

male is compromised.

### **Thrombosis of Vessels in the Spermatic Cord**

This condition is uncommon and is included for completeness sake. Its true nature has yet to be elucidated.

#### **An Acute Hydrocoele**

##### **Examination**

The mechanical nature of the lesion is expressed by the rapid onset of symptoms and the extreme tenderness to palpation of the involved tissues. In time the pain ameliorates and the signs of inflammation become more apparent.

It is essential to determine whether the swelling is confined to the scrotum or involves both pouch structures and the more proximal spermatic cord.

#### **Non-Mechanical Lesions**

##### **Acute Epididymo Orchitis**

This is an inflammatory process of the epididymis alone in most cases due to:

- Haematogenous dissemination of bacteria from a primary focus elsewhere (bacterial infection).

Reflux of urine (chemical irritation). When encountered in this age group an underlying congenital abnormality of the lower urinary tract (obstruction: reflux) may be the reason for the problem, or the cause of urinary tract infection that has spread to the scrotum.

##### **Examination**

Tissue swelling is a prominent finding. The epididymis is large and tender. Secondary changes, a vaginal hydrocoele and skin pathology are common.

#### **Preschool and Schoolgoing Children**

Symptoms and signs are basically confined to the scrotum though remote complaints (abdominal pain) are more prominent than in the infant. A history that trauma could be a causative factor is often obtained. In nearly all instances the problem lies within the scrotal pouch.

## **Mechanical Lesions**

- Torsion of the spermatic cord (intravaginal).
- Torsion of a vestigial remnant found in the scrotum. Although appendages to the testis, epididymis and cord occur, the problem is confined to the appendix's testis, which is prone to enlarge (Hydatid of Morgagni) making it most liable to undergo torsion.
- A complicated inguinal hernia.

## **Non-Mechanical Lesions**

- Acute epididymitis.
- Viral orchitis.
- Henoch-Schönlein purpura.

## **Establishing a Working Diagnosis**

Everything hinges on the fact that:

- in early life (first decade) mechanical lesions are the most common (70/30)
- the diagnosis must remain uncertain in nearly all instances until the scrotal sac is opened and its contents inspected
- torsion of the spermatic cord is associated with infertility
- torsion of the testis requires urgent correction - a delay of over 6 hours from the onset of symptoms to repair, represents management failure.

## **Intravaginal Torsion of the Spermatic Cord**

Associated clinical signs to look for in these patients are:

- an absent cremasteric reflex
- a shortened spermatic cord (a high testis)
- the lie of the testis is abnormal - there may be an alteration in the normal position of the testicles in the scrotum in both the involved or the normal side. The long axis of the testis is horizontal instead of vertical (superior inversion - the "bell clapper" testis)
- the involved testis may be obviously dystopic (imperfect descent)

When these additional pointers are present the patient has a clinical torsion; in their absence the case should be one of epididymitis. The effects of urgent nonoperative

manipulative detorsion or assessment by sonar or nuclear scan, are usually inconclusive and cause unnecessary delay in instituting effective management.

When there is an incontestable urinary tract infection or recent manipulation of the urethra has preceded the onset of symptoms, an epididymitis is assumed to be present and appropriate special investigation and medical management is ordered. In their absence an urgent exploration of the scrotum will establish the diagnosis.

### **Remember**

- The torsion of the cord is usually greater than 180° and labile.
- Tissue hypoxia and early symptoms are due to venous or outflow obstruction.
- The length or duration of the symptoms do not necessarily correlate absolutely with the degree of infarction (venous gangrene).
- Only the obviously gangrenous gonad is excised - although the blood testis barrier could be damaged by hypoxia, in the under 10 year old patient it would be folly to remove a potentially viable gonad.
- Pexy both testicles in all cases of torsion by placing them into subdartos pouches, holding them in this position by non-absorbable sutures.

If the exploration reveals an epididymis or a nonstrangulating lesion the surgeon leaves the operating room with the knowledge that the correct course has been taken, as torsion of the testis has been indisputably excluded.

### **An Irreducible Indirect Inguinal Hernia**

Urgent reduction of the contents of the pouch is the aim of treatment. When the patient is an infant, patient-related factors are implicated in preventing reduction from occurring. In a vast majority these can be controlled by adequate sedation. The patient is admitted to hospital and consent is obtained for operative reduction and repair of the hernia. When:

- Urgent reduction of the contents of the pouch is the aim of treatment. When the patient is an infant, patient-related factors are implicated in preventing reduction from occurring. In a vast majority these can be controlled by adequate sedation. The patient is admitted to hospital and consent is obtained for operative reduction and repair of the hernia. When:

- overt clinical signs of intestinal obstruction are present with diaphragmatic splinting due to distension; or

- clinical findings on local examination indicate that the contents of the sac are infarcting or dead.

The patient is resuscitated and operative reduction proceeded with.

In the absence of the above two contraindications, a trial of nonsurgical reduction is initiated:

- sedation is ordered. An adequate premedication chosen by an anaesthetist is preferred.

- an infant is placed in Gallows traction. Simple reduction is facilitated by the effect of gravity and by flexing the hip to open the external ring.

With the patient well sedated the hernia is manipulated. In 85% of cases complete reduction of the contents of the pouch is achieved and the hernia can be electively repaired. Where a significant degree of oedema complicates the prolapse it is preferred to delay this operation by 24 hours.

Note:

- All cases in whom nonoperative reduction was successful are kept in hospital until a herniotomy has been performed.

- It must be stressed that taxis, forceful manipulative reduction of the contents of a hernia, should never be used in a child.

If in spite of sedation the hernia remains unreduced, urgent operative repair is proceeded with.

### **Comment**

### **The Acute Scrotum**

**J H Louw**

The following conditions should be added to non-mechanical causes of the acute scrotum:

- Leukaemic infiltration

- Degenerating neoplasm. 60% of paediatric testicular tumours occur in the under 3 year old age group and 80% of them are malignant tumours (yolk-sac tumours).

## **Chapter 15.10: The Paediatric Acute Abdomen**

**M R Q Davies**

### **Introduction**

Acute abdominal is the usual presenting symptom. Its cause, especially if the patient is young, may be difficult to establish. In nearly every instance it revolves around the single question, is it or isn't it appendicitis? For if it is not, then should the case come to surgery, it is probable that the operation was not necessary. The patient thought to have

appendicitis on whom a "negative laparotomy" has been carried out remains the most common misdiagnosis that leads to unnecessary major surgery.

Recurrent headaches, recurrent abdominal pain, and recurrent limb pains are the common complaints of children. For this reason the correct approach to it is essential.

### **Abdominal Pain in Children**

The importance placed on the history in these cases has been over-emphasised. Before the age of communicative speech is required, a third person, usually a parent, supplies the history. Tenderness to palpation is easily assessed by the observer where established pathology is present. However, in early cases physical signs may not be so evident. Later in life communication may still remain a problem, and it may be the patient's first encounter with pain of visceral origin. These facts compound the difficulties in arriving at the correct diagnosis. *There are two categories of pathology that give rise to the symptom.*

#### **The Organic Group**

- A definite cause is found and specific treatment is given.

#### **A Functional Intra-Abdominal Anomaly**

The term psychogenic is attached to this group, but should not be used as it labels the patient inappropriately.

- The symptoms are essentially similar to those seen in the organic group, but a convincing explanation for the symptomatology is difficult to find. This is the more important group from a surgeon's view-point. Unexplained abdominal pain is by far the most common reason for an emergency admission to a surgical unit serving a developed community. In First World countries the prevalence of appendicitis appears to be on the decline.

#### **Acute Non-Specific Abdominal Pain (ANSAP) vs. Appendicitis**

It is the diagnostic see-saw of paediatric gastro-enterological practice.

Note:

- Appendicitis is the commonest cause of abdominal pain that requires to be operated upon.
- Only 1/3 of children admitted to hospital with abdominal pain will require an operation.
- Of the causes of abdominal pain that are correctly treated by surgery, appendicitis and its complications constitute more than 90% of cases.
- In the differential diagnosis of organic diseases that cause abdominal pain, no

single entity stands out, making it consistently second in line.

- If it isn't appendicitis, an operation is probably not necessary.

## **The Examination**

### **General**

How ill is the child? A general assessment is mandatory. Commence it by taking the patient's temperature. This is the most effective way of starting an examination of an acutely sick child, as a measure of confidence is established in this manner.

Look for:

- A rash (Henoch-Schönlein purpura; measles).
- Upper respiratory tract infection.
- Chest pathology.
- A lymphotropic viral infection (palpable lymph glands).

A normal temperature (37.4 °C) in appendicitis:

- may be present in late childhood
- is against advanced disease in the preschool group.

In appendicitis, the pyrexia reflects on and parallels the pathology; it seldom exceeds 39 °C where the diagnosis of a surgical abdomen is in doubt.

### **Local**

Appendicitis is a clinical diagnosis. It is made by palpating the patient's abdomen.

*It follows that misdiagnosis* is the direct result of an inadequate abdominal examination. Defer making any statement until an adequate assessment, after repeated attempts, if necessary, has been made. The use of a sedative may be necessary. If the diagnosis is still in doubt, be sure to examine the external genitalia for pathology and perform a rectal examination. This manoeuvre is never obligatory and is unnecessary when a diagnosis has already been made. It is of most value in the older child and when hard signs (mass, an abnormal stool) are looked for.



## **The Investigations**

Side-room screening:

- Haemoglobin
- White cell count
- Urine dipstick plus microscopy
- Occult blood in stool.

Special investigations:

These are seldom required. They are most useful when a surgical problem other than appendicitis is present.

## **The Working Diagnosis**

- ANSAP (acute non-specific abdominal pain)
- Appendicitis
- Intraperitoneal and extraperitoneal medical and surgical causes of abdominal pain (a long list of uncommon conditions).

## **Acute Non-Specific Abdominal Pain**

### **The Symptoms**

In childhood these are real and not just in the mind. A supratentorial influence in cases with recurrent abdominal pain is not disputed.

- Pain

Described in vague terms; not incapacitating; may be different in nature on different occasions; intensity varies.

- Duration

Short in terms of time (settles within an hour or two).

- No marked influence on GIT function
- Site

Felt peri-umbilically.

## **The Signs**

Tenderness with or without guarding (voluntary spasm) is found; this can closely mimic the signs of early appendicitis.

## **The Diagnosis**

Is usually uncertain. When appendicitis is possible a period of clinical monitoring is ordered.

Active observation in the management of acute abdominal pain.

- The doctor revisits the patient within a specific period.

- Bedrest

- Nil by mouth - ordered during early monitoring period but not continued with as placing the intestinal tract at rest may delay the appearance of hard signs - free fluid intake is encouraged.

- No antipyretics or analgesics

- Intravenous fluids are withheld unless they are absolutely necessary as they may increase the patient's sense of wellbeing and delay making a definitive diagnosis.

- Nursing instructions

Monitor: temperature; pulse; behaviour.

## **Appendicitis**

There are no pathognomonic findings. Organic pathology localised to the RIF in a child is due to appendicitis in 90% of cases.

*The triad* of abdominal pain, fever and localised abdominal tenderness is appendicitis till proven otherwise.

## **The Pathology**

An occlusion of the lumen of the appendix is present in most schoolgoing children who develop appendicitis. An appendicolith, the faecolith that contains a significant amount of calcium, making it easily visible on X-ray, noted in a child with abdominal pain, signifies the presence of appendicular pathology and is an indication for appendectomy.

There has been a recent increase in the prevalence of phlegmonous or nonobstructive appendicitis.

## **Symptoms and Signs**

### **Preschool Patient**

There is often delay in establishing the diagnosis.

Appendicitis may be guised as:

- a generalised peritonitis
- an inflammatory mass
- an intestinal obstruction
- a gastroenteritis-like picture or a gastrointestinal infection could cause secondary appendicitis
- a urinary tract infection. The presence of white cells in the urine does not make this diagnosis, nor refutes the presence of appendicitis.

### **The Schoolgoing Child**

*The medical history can be of assistance in the diagnosis.*

- A shift of pain from navel (visceral) to iliac fossa (somatic) - 50% of cases.
- Woken or unable to sleep - over 50% of cases.
- Pain aggravated by movement - 75% or more of cases.
- No vomiting - in only 25% of cases.
- Dysuria and frequency - 10% of cases.
- Diarrhoea - 10% of cases.
- Confirm the presence of a pyrexial illness.

The decision to proceed with an operation is made once signs of (localized) peritonitis or a mass are detected. The region of maximal tenderness is found by percussion. Peritonitis produces rigidity which once detected, confirms the presence of organic pathology.

The vermiform appendix is usually located anatomically in the RIF, partially protected from the anterior abdominal wall by the caecum (retrocaecal position).  
*Unexpected variation in this anatomy may be the basis for the "missed appendix".*

Signs not in RF = pelvic position, or  
Interposition of other tissues = Retro-colic or retro-ileal.

An ability to palpate the site of the pathology is fundamental to making the diagnosis. Despite reservations expressed regarding rectal examination, it may be the only way of detecting a pelvic appendicitis. Furthermore it is more correct and accurate to use percussion to produce release tenderness than it is to use the painful, crude and more difficult-to-interpret rebound sign.

### **Causes Other Than Appendicitis or Peritonitis**

#### **Localized to the Right Lower Abdominal Quadrant**

- Mesenteric adenitis
- Primary peritonitis
- Meckel's diverticulitis
- Ruptured ovarian cyst
- An ileitis (typhoid; yersinia)
- A typhlitis (amoebic dysentery; Shigellosis; in the immuno-compromised patient)

#### **Mesenteric Adenitis**

Hyperplastic (enlarged) lymph nodes in the ileocaecal mesentery of a child with acute abdominal symptoms not unlike those of acute Appendicitis, is termed *mesenteric adenitis*.

Specific mesenteric adenitis may be viral or bacterial in origin. Acute nonspecific mesenteric adenitis should be a "dying" diagnosis. As the number of unnecessary appendectomies performed by any surgical unit declines so *pari passu* will the prevalence of this entity. *It is possible to identify these cases and to treat them non-operatively as follows:*

- Signs of or a history of an upper respiratory tract infection: Waldeyer's ring, lymphnode pathology.

The complaints (fever 39 °C+; severe pain) and the investigations (high WCC - lymphocytosis) are out of proportion to the signs (minimal tenderness; poorly localized).

A short period of in-hospital monitoring, plus treatment as the uncommon pyogenic form will become complicated if not treated, is ordered.

## **Primary Peritonitis**

### **Meckel's Diverticulitis**

Meckel's diverticulum is the most commonly encountered Vitelline remnant. It gives rise to symptoms when it is associated with a congenital band or heterotopic tissue in its wall. It is a true diverticulum that usually has a wide neck, stasis of its intraluminal contents with diverticulitis is uncommon. Episodes of mural ulceration and healing where gastric epithelium is present resulting in stenosis alter this situation. An antecedent history of abdominal pain and rectal bleeding, may point to this complication.

#### **Causes Other Than Appendicitis of a Right Lower Quadrant Mass**

- Torsion of an ovarian tumour.
- Deep iliac adenitis.

In appendicitis a palpable swelling localised to the RIF may be one of the following:

- An appendix mass. This is a periappendicular congregation of omentum and bowel that surrounds the inflammatory process. When the clinical signs of infection are localised to the involved site, nonoperative treatment is usually successful in most of these patients. However, this is unpredictable.

*Past experience has shown that:*

- Where the mass is still obviously tender an appendectomy can simply and safely be carried out. If the abdomen is palpated during an earlier phase, the mass may only be felt with the patient under sedation or an anaesthetic. It would not be wise to manage this problem nonoperatively, especially in the young patient, although it is practised by some.

- Where the mass is clearly palpable and slightly tender, it will rapidly heal with appropriate management, even in a child. Whether an interim appendectomy is required in these cases is an issue that still has to be resolved.

- An appendix mass. This is a collection of pus and it is treated surgically. The diagnosis is made sonographically when a cavity is seen lying within an inflammatory collection. Once again experience has shown that in most instances without causing damage the appendix can be removed during the procedure. Interim appendectomies are seldom performed by those children's surgical services that follow the above guidelines.

*In a child it is important to be able to distinguish between an intraperitoneal (complicated appendicitis) and an extraperitoneal mass (pyogenic infection) that involves the external iliac lymph nodes.*

With appendicular or intestinal pathology the swelling is not confluent with the inguinal ligament, which can be felt separate from it.

## **Intussusception**

Although usually encountered in a chubby infant of 3 to 18 months, this condition must always be considered, especially during the preschool years. *Anatomically the individual components of an intussusception are:*

An ensheathing layer = the intussusciens.

The entering layer + The returning layer = the intussusceptum.

- The intussusciens has a clearly definable neck.
- The intussusceptum has a head, the lead point.

Most intussusceptions, due to perverted peristaltic movements, undergo spontaneous reduction as they are short in length and do not have anything to perpetuate them. Where a fixed neck or more usually a fixed lead point is present, ie, an endophytic lesion producing a polyp-like effect (hyperplastic Peyer's patch - infant; invaginated Meckel's diverticulum or juvenile polyp - preschool child; non-Hodgkin's lymphoma mural infiltrate - schoolgoing child), reduction does not take place spontaneously.

The pathology causes the intussusception and localises the level of the lesion.

The prefix indicates the site of origin of the lesion.

Small intestine: Jeuno-ileal; Ileo-ileal; Ileo-colic.

Boundary type: Ileo-caecal.

Large bowel: Caeco-colic; Colo-colic; Sigmoido-rectal.

### **Notes on Symptoms**

- Painless intussusceptions are singularly uncommon.
- The pain experienced is of a periodic nature; cyclical unhappiness = colic.
- It produces neurogenic shock, depleting the patient's reserves which indicates its severity.

The invaginated bowel obturates the intestinal lumen, and like other causes of this type of obstruction (gallstone ileus), the occlusion produces signs of an incomplete holdup. Vomitus, unimpressive in volume - non-bilestained. No abdominal distension until late in the course. Colonic emptying continues as the intussusception migrates distally.

## Notes on Signs

In the boundary type:

- On palpating the abdomen a sausage-shaped mass can be felt.
- On rectal examination:
  - an empty lumen
  - the apex of the lesion is felt
  - the stool contains blood (redcurrant jelly).

In any paediatric patient with abdominal pain and blood in the stool an intussusception could be present.

- Non-ischaemic intussusceptions are singularly uncommon.
- The strangulation produces:
  - Breach in antibacterial defences - a pyrexial illness.
  - Loss of intravascular volume - hypovolaemic shock.

## Special Investigations

- An abdominal scout X-ray study will show an intestinal obstruction.
- An abdominal sonar will confirm the presence of a mass and delineate its nature.
- When the lesion involves the colon a contrast enema will make the diagnosis.

## Treatment

- Prevent aspiration.
- Decompress the upper GIT.
- Correct shock.

Always present and can be demonstrated if the patient is correctly assessed. In even the compensated case, give colloid 20 mL/kg over 2 hours and then only proceed with any form of treatment.

- Antimicrobial cover is administered.
- *Hydrostatic reduction is the management of choice but is contraindicated:*

- in unresuscitated patients
- when an abdominal catastrophe is present clinically
- if an experienced interventional team is not available.

-Surgery with reduction by manipulation and/or resection completes the list of management alternatives.

*Two further diseases featuring the letter P are added to those of primary peritonitis that have already been discussed, namely pancreatitis and parasites.*

### **Pancreatitis in a Child**

This is caused by:

- Blunt abdominal trauma.
- Idiopathic - an ever increasing reported incidence.
- Infection - viral.
- Drugs.
- Obstruction - worms.
- Metabolic - hyperlipidaemia; - aminoacidurias.

### **Parasites Associated With the Surgical Abdomen**

These are:

- Entamoeba histolytica
- Ascaris lumbricoides.

### **Comment**

#### **The Paediatric Acute Abdomen**

**J H Louw**

I agree that the crucial problem in children presenting with acute abdominal pain revolves around the single question: is it due to acute appendicitis or not? It has become fashionable to concentrate on acute non-specific abdominal pain (ANSAP) as appendicitis, but it must be pointed out that the diagnosis of ANSAP is usually only made in retrospect, and that it can only be a provisional working diagnosis.

In South Africa abdominal tuberculosis should be included in the differential



diagnosis, also when a right lower-quadrant mass is present.

## **Chapter 15.11: Paediatric Trauma**

**M R Q Davies**

### **Introduction**

In developed countries injuries are the commonest cause of death of persons between the ages of 6 months and 15 years.

### **Cause of Death in Ranking Order**

- MVA or related accidents
- Burns
- Drownings.

Paediatric injuries are grouped under one of three major headings:

- Birth trauma
- Child abuse (intentional injuries)
- Trauma to the mobile child.

### **Incidence**

Trauma causes one in 10 deaths in the general population and five in 10 deaths in the age group one to 14 years.

### **The Abused Child**

10% of all injuries in under 5 years olds and 60% in the under 3 year old age group are cases of child abuse. During this period, children are demanding, defenceless and mainly non-verbal. Repetitive attacks of increasing severity usually by a related caretaker (punishment injuries) occur.

- 20% will have a positive radiological bone survey.
- 15% are at risk of dying if the case is sent home without treating the cause of the injury.

### **The Mobile Child**

The Ignorant: "At about 2 years of age the child becomes a runner, a climber and an explorer!" Collisions and falls onto blunt objects.

The Incapable: The child and his bicycle - under 8 years old: cannot control it adequately.

Under 12 years old: has insufficient road sense to use a public road.

An Innocent Bystander: Simple victims of their circumstances.

The child as an occupant of a car - over 50% of unnatural deaths are transport related accidents.

Table 15.11.1. The child as a patient

- A metabolic BONFIRE - O<sub>2</sub> and carbohydrate hungry.
- Built to LOSE heat.

Table 15.11.2. The paediatric airway

It commences high in the neck.

The newborn = obligate nose breather.

The cricoid ring = narrowest part.

Age

Premature	2.5/3.0
Newborn	3.5
1 year	4
4 years	5
8 years	6
12 years	7

Table 15.11.3. Important vital signs

	Baby	Child	Adult
Heart rate	120	100	60
Respiratory rate	37	20	12

Saving life comes first. In a new paediatric trauma case shock is almost always cause by blood loss.

The volume of the intravascular compartment is 80 mL/kg.

Haemodynamically significant losses are:

- 20 mL/kg if rapid

- 40 mL/kg if slow.

Resuscitation can be equated with a therapeutic "poker" game. Observe the response to a bolus of 20 cc/kg of a physiologic electrolyte solution.

Table 15.11.4. Estimating the patient's mass

Infants double their birth mass in 5 months.

Infants treble their birth mass in 1 year.

Between:      1-9 years mass (kg) = (age + 4) x 2  
                    8-12 years = age x 3

### **Head Injuries**

The problem here is delayed deterioration of consciousness after "trivial" injury.

Under one year this is common but usually insignificant. In an over one year old in contrast it may be malignant.

### **The Talk and Die Situation**

For this reason it is important to observe all cases closely. This must be done in hospital when a history of loss of consciousness is obtained, or in lesser instances where the home circumstances are inadequate.

The cause of loss of consciousness is secondary brain swelling (oedema). A relatively larger proportion of the total blood volume of a child is in its head. Think of a child's brain as an erectile organ.

Prevent (prophylaxis) and treat this change by fluid intake restriction and diuretic administration. It is essential to secure the airway.

### **Thoracic Injuries**

These are second to head injuries as a cause of death in traffic-related accidents.

Penetrating thoracic trauma is relatively rare. The problem is the resilient chest wall of the child. Paradoxically significant mortality is seen in patients without rib fractures, but who have sustained significant pulmonary parenchymal injury. Children who were "run over" with few clinical findings on admission (stress petechiae) may develop ultimately fatal pulmonary (or myocardial) contusions.

## **Abdominal Injuries**

The result of blunt impact in 90% of children compared to 50% of adults. A child with an abdominal injury is difficult to examine and it is usually associated with another extra-abdominal injury.

### **Points to Remember**

- Unprotected viscera in the upper abdomen are crushed against the spine.
- Air swallowing with acute gastric distension causes confusing clinical signs.
- The most frequently injured organs are kidney, spleen, liver.
- The commonest cause of death are brain, liver and lung injuries in that order.
- The relative mobility of the kidney obligates a check for the presence of red cells in the urine. If positive a limited (one shot) IVP is required to exclude a renal arterial occlusion.

### **Clinical Assessment (think of 5)**

- Parenchymatous viscera.
- Viscera concerned with the production, transport and storage of urine.
- The intraperitoneal hollow viscera - the intestines.
- The retroperitoneal space
  - Pancreas
  - Great vessels
  - Duodenum
- Diaphragm.

Diagnostic peritoneal lavage is accurate in determining an intraperitoneal injury, but such injuries do not often require laparotomy for repair. Therefore lavage is ordered only in exceptional instances.

The most important parameter used in assessing the magnitude of an intra-abdominal injury is serial abdominal examination.

A raised serum amylase level detected during the phase of resuscitation is noted but not reacted to. An obvious elevation on repeated testing following stabilisation indicates significant pancreatic ductal damage that should be treated surgically.

## Table 15.11.5. State

### **Stable**

Non-operative treatment for: 1. kidney - traditional; 2. spleen - most cases; 3. liver - recent.

### **Unstable**

Emergency laparotomy: 1. continuing haemorrhage; 2. an acute abdomen; 3. ruptured hollow viscus.

### **Trauma of the Spleen**

Historically the mortality of nonoperative management was 100%.

1919: The importance of the spleen in resistance to infection described.

1952: Susceptibility to infection after splenectomy performed in infancy documented.

1973: Post-splenectomy sepsis (OPSI) reviewed.

Overwhelming post-splenectomy infection (OPSI) is a definite threat, well recognised by the paediatric surgeon. This is one form of hyperacute sepsis that can be compared to the *Waterhouse-Friederichsen's syndrome*.

### **Cause**

The spleen plays a major role in host defence. In this context the early elaboration of antibodies in response to any intravenous challenge by particulate antigens is of apparent extreme importance in infection by micro-organisms with polysaccharide capsules, ie, *Streptococcus pneumoniae* (50% of cases), *Meningococcus* and *H. influenza* are responsible for 80% of post-splenectomy septicaemias. The critical function of the spleen in these infections is the clearance of *blood*-borne antigens towards which the plasma antibody concentration is low (less common organisms or serotypes). Following splenectomy the defect is lifelong and most severe in:

- the infant and child
- children with haematologic disorders, malignancy or portal hypertension.

In the patient who has had a splenectomy for trauma the risk factor is 50 x that of unsplenectomised people.

### **Presentation**

- These include chills, fever, and abdominal pain and vomiting.

- This is followed by shock, DIC and coma.
- Death occurs in a matter of hours.

### **Prevention**

- Preserve the maximal amount of splenic tissue with its normal blood supply.
- Non-operative management of splenic rupture is possible in most cases in childhood. Stabilize the patient in a trauma unit, then admit for active observation in high-care unit.

### **Laparotomy is Indicated:**

- for blood loss when a massive haemoperitoneum is present or blood volume replacement greater than 40 mL/kg is necessary for ongoing haemorrhage;
- for organ damage where clinical evidence that the spleen has been shattered (confirmed by scan) exists; or where a left upper quadrant mass (clot) with persistent left upper quadrant signs (peritonism) is present or after a failed trial of conservative management.

The operation in these instances is seldom life saving but is always the best course as it expedites recovery by immediately controlling a potentially dangerous situation.

It is not the laparotomy that is frowned upon but the unnecessary removal of functional splenic tissue.

### **Practice Splenic Conservation**

In the young the spleen is relatively simple to operate upon. After establishing vascular control at the level of the hilum, splenic repair and/or segmental resection can be performed. An adequate mass of retained tissue plus the maintenance of its normal blood supply, not one without the other, is essential. Splenic autotransplantation fails in its aim because normal blood flow to the spleen is not maintained.

### **The Management of the Asplenic Patient**

#### **High Risk of OPSI**

- Give prophylactic penicillin till the patient is no longer "immunologically compromised" by age or disease.
- Administer polyvalent pneumococcal vaccine.
- Treat all infection vigorously medically.

### **Low Risk of OPSI**

- Administer polyvalent pneumococcal vaccine.
- Wear *Medic Alert* bracelet.
- Educate patient and family.
- Treat all infective illnesses with an empirical antimicrobial drug (kept at home - available for immediate use). Always consult a medical practitioner.

#### *Management principles used in the patients treated nonoperatively with suspected splenic rupture*

- Close monitoring till stable and recovering.
- Bed rest until abdominal signs have resolved.
- Confirm diagnosis by scan:
  - sonar
  - isotope.
- Mobilize and discharge.

There is a fear of secondary haemorrhage which is a well-described entity in adults but has not been reported in children. In this age group it is accepted that most cases of so-called delayed rupture are patients in whom there has occurred a delay in recognition of the presence of splenic trauma.

### **Comment**

#### **Paediatric Trauma**

#### **J H Louw**

The value of serial scans (sonar, isotope and CT) should be emphasized in the management of abdominal trauma. It should also be stressed that conservatism is not only applicable to spleen injuries, but also to liver trauma in children.

### **Chapter 15.12: Burn Injuries in Paediatric Patients**

#### **M R Q Davies**

#### **Introduction**

The altered physiology which occurs during the post burn period is of a magnitude surpassing that of other types of trauma and the duration of these changes far exceeds that

known to occur in injuries of other types.

Table 15.12.1. Cause

Hot liquid	70% (immersion or spill scalds)
Fire	28%
Chemical	2%
Electrical	0.5%

Table 15.12.2. The five points considered during the appraisal of the seriousness of a burn injury

- Estimate of percentage of body surface burned.
- Depth of burn.
- Age of the patients.
- Anatomic locations of burns.
- General physical condition of the patient and concomitant injuries.

### **An Outline for Sorting or Disposition of Burned Patients**

Critical burns:

- 2° burns of over 25%
- 3° burns of the face, hands or feet or if over 10%
- Electrical burns
- Burns complicated by:
  - respiratory tract injury
  - major soft tissue injury
  - fractures.

These cases should be admitted to a special burn unit.



## **Major Burns**

- 2° of 10-25%
- 3° of less than 10% except hands, face or feet.

It is accepted practice to treat this group in a special burn ward in a general hospital.

## **Minor Burns**

- 2° less than 5%
- Ambulatory care suffices.

## **First Aid**

- Stop further injury
- Fires are extinguished and clothes are removed. In chemical burns the immediate copious lavage of the wound surface with any available bland solution (water) is mandatory.
- Cool the head damaged surfaces.
- Cooling is applied for about 10 to 15 minutes until all pain has been controlled. Limit this form of therapy to wounds of less than 15% of the body surface to prevent hypothermia.
- Cooling can be used within a 3 hour time lapse. Animal experimentation has showed that tissue destruction is limited by that manoeuvre.

## **The Accident Where Fire is the Energy Source**

Combustion is stopped and the surface is cooled. Signs of an inhalation injury are looked for. The inhalation of heat, carbon monoxide and/or the noxious vaporised products of combustion has taken place. A true lung burn seldom occurs, but the inhalation of hot air, can lead to thermal trauma, involving the mucosal surfaces of the glottis and upper trachea. Clinical signs of major upper airway obstruction are found.

In enclosed space accidents involving fire, "smoke poisoning" commonly occurs. The inhalation of cyanides and other acid products released when plastic materials are vaporised damage the smaller airways. Bronchospasm is often followed by pulmonary oedema and subsequent bronchopneumonia. *The clinical signs to look for are:* the presence of burns involving the face, nostrils, nasal hairs, the mouth and upper pharyngeal areas. Where carbonaceous sputum is detected, gross inhalation has occurred. Tissue damage occurs in the peripheral airways and surrounding parenchymal structures in the lungs.

### Table 15.12.3. Criteria for admission to a paediatric burns unit

Burn wound of 10% or greater in size. Anything less than 10% affecting: face, perineum, hands and feet, flexures, circumferential.

All infected burns

All cases where an inhalation injury is possible

Electrical thermal injuries.

### Table 15.12.4. Principle phases in management

Emergent (1 to 72 hours post burn)

Acute (72 hours to completion of autografting)

Reconstruction.

Bronchoscopy is mandatory for all suspected respiratory burns. The onsite administration of a therapeutic dose of steroids can limit the degree of tissue destruction.

### **Determining the Depth of Tissue Destruction**

Wounds on clinical evaluation are either obviously superficial or deep, or most commonly, of indeterminate depth.

Three levels or depths of tissue death are traditionally described. An epidermal lesion represents the first degree burn; total skin death where damage extends into the subcutaneous fatty layers, the third degree burn. Both degrees are easily recognisable clinical types. Dermal involvement, the second degree burn, is usually encountered in wounds where mixed levels of destruction are present. Accurate clinical assessment within the first 5 days of burning may be impossible. This is the reason why the modern burn physician speaks of the partial thickness or dermal burn, and the full thickness burn wound.

The partial thickness burn is defined as *a wound where the depth of tissue destruction has extended into the dermis, is of mixed depth and nerve endings and dermal vascular plexus have not been totally destroyed.*

In contrast the full thickness lesion has a pigskin appearance, is anaesthetic and appears bloodless, and blistering seldom occurs. The accurate distinction between these two depths which are of prognostic importance may only be possible towards the end of the first week.

The magnitude of the injury is assessed by estimating not only the depth of the wound but its extent. Most medical practitioners are well-acquainted with the rule of 9 but it is not applicable to children. In early life the lower extremities are smaller and the head larger in surface area. Berkow's charts should be consulted. The patient's outstretched palm

represents a surface area of approximately 1 per cent.

### **Management of the Patient**

- Record vital signs
  - temperature
  - pulse
  - blood pressure
  - respiratory rate
- Monitor visceral perfusion
  - level of consciousness
  - urine flow
  - Check for bacterial infection
    - on skin surface
    - involving ENT system (full clinical evaluation).

### **Investigations**

#### **Side-Room**

- Blood
  - Hb
  - Haematocrit
- Urine
  - Specific gravity
  - Haemoglobin products.

#### **Laboratory**

- Blood
  - Full blood count
  - Blood gas determination (CO level)

The halflife of CO is 3 hours and the blood level is only of value if the blood is taken very soon after the injury was sustained.

- Blood typing
- Cross match
- Microbiology specimens
  - oropharynx
  - skin - burn wound and other infected sites
  - sputum
  - stool.

#### **Radiology**

- Chest film.

## **Treatment**

The objectives of treatment in order of importance are:

- Relief of pain
- Prevention or correction of hypovolaemic shock
- Prevention of burn wound infection
- Immediate and continuous biological closure of the areas of skin damage
- The early detection and treatment of burn wound infection
- The early autografting of areas of deep partial and full thickness skin loss
- Nutritional support
- Prevention of contractures and hypertrophic scar formation
- Psychological support

### **Pain Control in the Hospitalized Burn Patient**

The pain experienced is inversely proportional to the depth of the initial thermal injury which represents the degree of receptor irritation or destruction.

### **Analgesics and Sedatives**

#### **Early**

- Keep to a minimum
- Rarely needed regularly beyond 48 hours
- In the shocked patient use titrated IV doses of morphine 0.1-0.25 mg/kg.

#### **Late (Dressing Changes)**

Ponstan - Vallergran

Dosage: 2.5-5 mg/kg/dose

**Beware - do not deeply sedate a child if:**

- clinically shocked
- septicemia or ileus is suspected
- in the presence of a foodfilled stomach.

Regurgitation and aspiration is an ever-present potentially fatal hazard.

### **Control of Shock**

The aim of therapy is to prevent the development of hypovolaemic shock and renal damage.

Resuscitate the following patients:

- Less than 2 years old with a TBSA greater than 5%.
- Greater than 2 years old with a TBSA of 10% or more.
- Special circumstances
  - electrical burn
  - associated trauma.

### **Method**

Monitor (essential during period 0 to 72 hours post burn):

### **Vital Signs**

- Pulse
- Blood pressure
- Respiratory rate.

### **Body Temperature**

- Keep the patient normothermic (core temperature recordings)
- Prevent hyper/hypothermic reactions by controlling environmental temperature.

Environmental temperature should be between 28 °C and 33 °C.

### **Urinary Output**

- Catheterise the bladder to aid monitoring in greater than 20% burn injury
- Aim at a flow rate of 1 mL/kg/hour.

In the presence of haemoglobinuria, aim at greater than double this rate per hour.

*This is a useful indication of visceral perfusion only:*

- During the administration of "thin" fluids
- During the first 48 hours post burn
- In the presence of normal kidney function (urinary SG).

### **Haematocrit - Haemoglobin Values**

- Determine haematocrit 12 hourly: desired level less than 45%
- Determine Hb concentration 24 hourly: desired minimum level 10 dL/L.

### **Respiratory Function - Acid Base Balance**

- Determine blood gases
- Normal cell respiratory function, ie, increase FiO<sub>2</sub> levels (in head box up to 50%)  
in the patient with a burnt surface greater than 20%
- Ensure clear airway - prevent aspiration

- Control oropharyngeal secretions
- Prevent gastric content regurgitation.

Stop oral intake in all patients that require intravenous fluids for at least a 12 hour period.

Pass a nasogastric tube in all patients with a 20% or greater body burn.

Where respiratory injury is suspected humidified oxygen should always be given.

### **The Nasogastric Tube**

It is essential that the upper gastrointestinal tract is decompressed. Statistics show that aspiration of gastric contents is a well-known cause of death in the burnt child. In early life, mild degrees of shock are associated with severe impairment of splanchnic circulation and ileus often follows which, if unrelieved, may give rise to this unnecessary complication.

### **Guidelines to the Replacement Fluid Therapy**

#### **Fluid Type**

In the clinically shocked patient (hypovolaemic situation), or with a full thickness skin loss greater than 10%, or in the presence of haemoglobinuria.

#### **First Fluid**

A balanced salt solution.

Volume: 20% of the estimated blood volume (10-15 mL/kg).

Rate: in 10 minutes to establish urine flow. In the patient that does not fulfil the above criteria, this first fluid is omitted.

#### **Second Fluid**

Plasma (FFP or liquid plasma - preferable to dry product).

Volume: Formula for use as a guide during the replacement plasma volume calculation:

$$3.5 \text{ cc x pts wt in kg x \% TBSA}$$

Formula modification - maximal % TBSA allowed:

- In the 0-2 yr old age group - 20%
- In the 3-7 yr old age group - 25%

- In the 8-15 yr old age group - 30%.

Rate: volume calculated is divided.

1/3 during the first 4 hrs

1/3 during the next 8 hrs

1/3 in the next 24 hrs

2/3 over 12 hrs.

Check the haematocrit twice daily. Should the haematocrit fall below 35% or to below 40% after 36 hours of fluid replacement therapy then:

### **Third Fluid**

Packed cells.

Volume: 20 cc/kg.

Rate: over a period of 6 hours.

### **Maintenance Fluid Therapy**

Fluid volume: determined by body mass in kg.

Infant: 150 cc/kg

2-5 years 100 cc/kg

5-8 years 80 cc/kg

8-12 years 50 cc/kg.

Fluid type: a maintenance solution in 10% dextrose.

Route of administration: In the presence of or expected intestinal intolerance - intravenously. In all other patients, orally.

### **Prevention and Treatment of Infection**

- Combat bacterial colonization
- Keep the eschar bacteriostatic
- Remove all dead tissue daily
- Promote wound healing.

## **Bacterial Contamination of the Wound**

Minor inoculations with bacteria are unpreventable. "Wild" strain organisms are relatively unimportant. By hospitalizing the patient you expose him to resistant strains.

### **Endogenous Bacterial Sources (Skin - ENT)**

In the presence of an obvious gram positive coccal reservoir give Erythromycin 25-50 mg/kg orally for 5 days.

*Exogenous bacterial sources:* Control the patient's environment.

A "clean" wound demands:

- Clean cubicle
- Clean fellow patients
- Clean dressing room partners
- Clean nursing and medical contacts.

Never transfer organisms between patients.

*To fulfil there requirements, the following is necessary:*

- Accurate bacterial monitoring. Monitoring swabs to be taken twice weekly by the doctor himself. Avoid unnecessary swabs.
- Where available facilities do not allow routine isolation, the patient's bedside in the burn unit (room rotation) is determined by: *wound staging according to age of wound.*

Stage I wound: Less than 10 days old. Eschar kept bacteriostatic = effective topical burn cream applications.

Stage II wound: After surgical debridement and successful complete autograft cover (healing donor sites).

Stage III wound: Greater than 10 day old. Debridement dressing techniques in use, following incomplete autograft cover.

or

New wound but gross bacterial contamination has occurred without clinical evidence of infection.

or

The burn wound that has not been adequately treated for a period of 24 hours or longer.



Stage IV wound: The infected burn.

- quantitative bacterial culture greater than  $10^4$  per gram of burn tissue
- cellulitis present
- Special contaminants present:
  - beta haemolytic streptococci, heavy growth of staphylococcus aureus, pseudomonas pyocyanea.

Maintain similar stages together at all times (dressing room plus ward cubicle). Major burns (25%+) must be managed in intensive care cubicles. Isolate all stage IV patients.

### **Control the Development of Opportunistic Infections**

Amphibiont organisms play an important role in burnt-patient mortality.

- Pseudomonas (gram negative bacillus)
- Moniliasis (yeast).

Control must be kept over this threat = maintain a strict antibiotic policy.

### **The Role of Systemic and Locally Given Antibiotic Substances in the Management of the Burnt Patient**

- Antibiotics are not to be given topically
- Antibiotics are only given systemically when:
  - A quantitative culture of greater than  $10^4$  organisms/gram of burnt tissue is obtained.
  - Clinical burn wound infection is present.
  - A septicaemia is diagnosed or strongly suspected.
  - In the presence of verdoglobinuria.

Pseudomonas organisms produce haemolysins and slime layer toxins which inhibit the conversion of verdoglobin to biliverdin. At both neutral, slightly acid or slightly alkaline pH a urine specimen containing verdoglobin turns an olive green colour when viewed under ultraviolet light.

- In the presence of thermally induced respiratory tract damage.
- Beta haemolytic streptococcal contamination or infection is suspected or diagnosed.

The burn eschar is avascular. A twilight zone exists in the plane between the eschar and the underlying unburnt tissues. During the systemic administration of antibiotics, this zone provides an ideal environment for the development of resistant bacterial mutants.

The preparation ordered is determined by:

- Best guess
  - The wound age and wound stage is a guide to the bacteria present.
  - Early - predominantly coccal gram + (0-72 hrs).
  - Intermediate - mixed coccal and bacillary (72 hrs-10th day).
  - Late - predominantly bacillary gram negative (10th day to end of the 3rd week).
  - The nature of previous topical therapy used.
- A recent bacteriological monitoring swab or blood culture result may prove a useful guide.
- The specific unit's own microbial population, and their known resistance patterns.

### **Length of Administration**

- Until clinical bacteriological growth control has been obtained.
- When used as antibacterial cover for wound surgery - a minimum of three days of administration is indicated in all instances.

### **Antifungal Agents**

Oral antifungal prophylaxis is given to all patients with a TBSA of greater than 15% and to all patients placed on broad-spectrum antibiotic therapy.

Preparations:

Mycostatin 1-4 million units 6 hourly.

Amphotericin B suspension or lozenges.

### **Beta Haemolytic Streptococci and the Burn Wound**

An unwanted organism in the burn unit.

Reason:

- It destroys epithelial cells and increases the depth of the burn.
- Lyses autogenous split thickness skin grafts. A significant percentage of children harbour this organism in their oropharynx. *Take the following steps:*
  - Do a throat swab on admission.
  - If the history of a recent cold or ENT evidence of an infection is found - start prophylactic antibiotic treatment.
  - Control and remove any burn care personnel source if present - isolate and monitor suspects.

*In the presence of suspected or proven contamination or infection, use:*

- Erythromycin 25-50 mg/kg/24 hours orally till bacteriologically clear or
- Cloxacillin 50-100 mg/kg/24 hours orally.

NB: Penicillin is often unable to control the growth of this organism and is *not* indicated.

### **The Control of Bacterial Growth on the Surface of the Burn Wound**

Cheomtherapeutic agents are used to control this bacterial growth. They are given prophylactically and therapeutically to all burnt patients. A variety of successful topical burn creams are available. The following are currently used by burn-care teams.

#### **Betadine Cream (Povidone Iodine) 5%**

Characteristics:

- Effective for 12 hours
- May impair epithelial regeneration
- Broad-spectrum antibacterial, fungal, viral action
- Poor tissue penetration
- Minimal side effects.

Uses

Good prophylactic cream to control bacterial wound contamination. Wound stage 1-2.

#### **Sulphamylon (Mafenide Cream) 10%**

Characteristics

- Effective for 12 hours
- Mainly gram negative bacterial cover
- Good eschar penetration
- Side effects:
  - Stings on application
  - Causes metabolic acidosis.

Monitor the blood pH when this cream is used in the patient with a greater than 30% burn. Do not use in the presence of impaired pulmonary function.

Uses

- To keep thick eschar bacteriostatic.
- In the treatment of gram negative wound infection.

## Silver Sulphadiazine (Flamazine) 1%

### Characteristics

- Effective for 24 hours
- Mainly gram negative cover. Gram + (staphylococcus aureus) infections are poorly controlled by this antibacterial agent.
- Poor eschar penetration

### Uses

Good prophylactic application for the initial treatment of the uncontaminated dermal burn. For the treatment of gram negative infections in dermal burns.

## Mupirocin

This is an excellent agent for gram positive infections.

## The Burn Wound

### The Partial Thickness or Dermal Burn

For a period of up to 10 days after injury, continuing epithelial cell death occurs in the dermal burn which is inappropriately managed.

Due to destruction of the protective action of the covering epidermal layer, the middle zone of potentially viable cells (perinecrotic zone) may be lost to the zone of cell death increasing the depth of the lesion or the eschar thickness. The loss of a vapour pressure barrier-like action of the covering layer, allows the already ill underlying cells to become dehydrated within hours after injury and cell death follows. Traditionally burn wounds are managed in an "open" (without dressings) or "closed" (with dressings) manner. Surgical dressings only partially protect the damaged cell zone.

Table 15.12.5. The three zones of the burn wound

- Central - Tissue coagulation - Cell death
- Middle - Vascular stasis - Cell damage
- Peripheral - Tissue reaction - Cell irritation.

Full protection with sick cell recovery follows *biological closure* of the burn wound.

*This may be achieved by:*

- leaving burn blisters intact
- retaining the destroyed *epidermal layer in situ* on the wound surface
- replacing the dead layer with a split thickness autograft
- the use of skin-substitute dressing techniques
  - homograft

- xenograft - fresh or lyophilized pig skin
- human placental membranes
- synthetic semipermeable barrier drapes (op site; Telfa)
- synthetic skin substitutes (Biobane).

Protection of potentially salvageable tissue is achieved by exclusion of the environment and the re-establishment of the milieu interior. Of these alternatives, the first and second are easily applied but the third and fourth are more difficult. However, none of these dressing methods have an inbuilt antibacterial action. Bacteria must be destroyed before biological closure is attempted. Should this be neglected, bacterially induced tissue destruction will follow which is far more serious than cell death as a result of dehydration.

### **Tangential Excision of the Thermally Destroyed Tissue Surface**

Concept: The surgical excision of lost tissue to limit ongoing and progressive damage cell death.

This is achieved by:

- removing the dead cell layer
- establishing complete control of bacterial inflammation
- replacing the function lost by the burnt skin layer.

### **Patient Selection**

This form of treatment should be offered to all suitable patients.

Method: Horizontal necrectomy is only applicable for use as a debriding technique in the burn wound with incomplete destruction of the dermal layer.

### **Wound Management**

Save the stasis layer.

Prevent:

- wound-cell destruction by occlusive dressing and continuous topical cream cover
- wound-cell inflammation by adequate topical therapy and nontraumatic dressing techniques.

### **Critical Assessment of the Degree of Cellular Destruction**

%TBSA on arrival and on day 2.

Depth of destruction on arrival and on day 2.

*Early wound-depth assessment is difficult*, but delay leads to deepening of the lesion, and an increase in the bacterial population in the dead cell layer with the increased likelihood of infection in the postexcision period.

## **Guidelines for Depth Assessment**

### **Energy Factor**

The magnitude of energy transfer is determined by:

- The nature of its source.
- The duration of transmission.

(Hot liquid injury - second degree; fire injury - third degree).

### **Anatomical Factor**

- Age of patient
- Site of lesion

(Skin thickness/dermal appendage development).

### **Wound Factor**

- Even with experience this clinical evaluation remains problematical.

## **Horizontal Necrectomy**

### **Indications**

- Deep 2° > 5% in size
- Dermal burns of:
  - hands
  - neck
  - flexor sites.

If tangential excision is contemplated, the following additional criteria must be met:

- Areas up to but not exceeding 10%-15% of the body surface may be surgically paired per sitting.
- The dead tissue layer must be bacteriostatic.
- Adequate topical therapy for 48 hours.

Preparation preferred:      Betadine cream  
   Silver sulphadiazine.

- Bacterial monitoring essential from admission.

Throat swab and burn wound surface swab to exclude: beta-haemolytic streptococci, Pseudomonas pyocyanea, resistant Staphylococcus aureus.

A burn eschar biopsy is indicated by day 7 and if tangential excision is used on the deep dermal burn wound. A quantitative bacterial count of organisms greater than  $10^4$  is regarded as an absolute contraindication to surgical incision. (Stage IV wound.)

- Adequate donor sites must be present to provide graft cover for the area surgically debrided.
- The patient must be fully resuscitated and in a clinically stable state.
- Adequate patient support facilities must be available for use during the surgical procedure.
- Blood loss replacement.
- Temperature control.

Table 15.12.6. Difference between superficial (partial thickness) and deep (full thickness) wound

- Wound surface:

Pink - red  
Wet  
Blanching on pressure

Pale  
Dry and flat  
Vessels occluded  
At 2 days - surface translucent revealing thrombosed veins  
Flame burn - brown, charring.

- Blisters:

Large  
Thick-walled  
Will usually increase in size

None  
If present thin-walled  
Will not increase in size

- Sensation:

Test day 2. Applicable in children 4 or more years of age.

Normal or increased sensitivity to pain and temperature

Anaesthetic to pain and temperature

- Texture:

Normal to firm

Firm to leathery.

### **Surgical Procedure**

The wound is not uniform in depth of destruction but the whole of the affected thermally damaged surface must be graft-covered at the completion of the surgical

debridement (total biological closure).

### **Method**

Clean all surgical surfaces with Povidone iodine skin prep solution. Excise the eschar with a Humby knife and start in the central area where the estimated thickness of destruction is greatest and cut to the sides of the wound removing very thin slices of dead tissue until punctate haemorrhages appear. When too much tissue is excised, vigorous bleeding is caused. This results in unnecessary loss of dermal tissue and excessive blood loss, both of which are avoidable.

### **Blood Loss**

In the small child this may be excessive - be prepared. Excessive loss may be curbed by careful dead tissue excision. The use of a tourniquet where applicable and adequate intraoperative help.

### **Graft**

Use meshed graft (for drainage). The whole of the burnt surface is covered with skin in a jigsaw puzzle like manner.

Maintain graft placement with:

- steristrips
- absorbable stitches

### **Dressing**

Vaseline gauze followed by "Kling" gauze bandage soaked in Betadine topical cream is used. Add a further layer of cotton wool or crepe if immobilization is necessary.

### **Post-Surgical Care**

Change surgical dressing on day 4 or 5 and redress with Betadine-soaked "Kling" gauze bandages and crepe cover until full healing has occurred.

### **The Full Thickness or Deep Burn Wound**

As all epithelial elements have been destroyed both open and "closed" wound dressing techniques may be utilised. As the dermal layer and its vascular plexus is also destroyed, the surgical technique of horizontal necrectomy, can no longer be used.

### **Further Differences Need Emphasizing**

- A burn cream that is applied to the surface of a thick eschar, must be able to penetrate through the layer without losing its anti-bacterial effect.



- Surface bacterial swab specimens often prove inadequate. Biopsy methods should preferentially be used.

- The constricting effect of the eschar leads to impairment of the blood flow to and from damaged tissue. Escharotomy is often a tissue-saving manoeuvre and especially in the hand the complete but conservative, excision of eschar, may save tissue loss due to *strangulation*.

- Anaerobic conditions may be present. Tetanus is a known complication of this type of burn wound.

### **Management**

- Adequate topical therapy is essential.

- Prevent nosocomial infection - a protracted period of open-wound management faces these patients.

- Treat burn-wound infection aggressively - diagnose infection before signs develop.

- Prevent tissue strangulation - Doppler assessment of vessel perfusion should be utilised.

- Surgical removal of dead tissue must be carried out - on the 8-10th day in the uninfected patient and during each tubbing session.

- Skin graft cover is only possible once all dead surface tissue has been fully removed.

- When an adequate supply of autogenous skin graft is not available - biological substitute dressing techniques may be used.

- Nutritional depletion, hypertrophic scarring and/or keloid formation and contractures, must be expected and all known methods of preventing these complications employed.

### **Nutritional Support**

#### **The Catabolic Phase of Injury**

Mass loss accompanied by negative nitrogen balance, characterises the early metabolic response in the patient with extensive thermal trauma.

Reason:

- After major burns a significant rise in basal metabolic rate is seen.

- Excessive fluid loss to the exterior follows on thermal destruction of skin

function: 2425 kJ are lost during evaporation of 1 litre of water.

- Failure to obtain early wound closure places a prolonged period of abnormal stress on the patient.
- Infective complications increase body temperature and caloric requirements.

### **Partial Control of the Increase in Metabolic Rate**

May be achieved in the following manner:

Early physiologic closure of the burn wound and donor sites.

- Biological wound closure
- Tangential excision of dermal burns
- "Early" surgical debridement of areas of full thickness skin loss, and "early" autograft closure

The prevention of burn wound infection

The control of the environmental temperature

By increasing this temperature from 22 °C to 32 °C the basal metabolic rate may be *reduced* by:

25% in moderate burns (25% TBSA)

50% in severe burns (50%+ TBSA).

The optimal temperature varies between 28 °C and 29 °C - a temperature slightly above that of the unburned skin, but below that at which the skin begins to sweat.

### **Calories for Repair**

#### **The Maintenance of an Adequate Energy Intake**

A formula for estimating this requirement for burnt children up to twelve years of age per 24 hours is:

Protein: 3 g/kg body mass + 1 g/% burn

Energy: 250 J/kg body mass + 150 J/% burn.

The maximum TBSA value which may be used during this calculation is 50%.

## **Monitor the Caloric Intake and Estimated Losses**

Daily energy intake estimates should be recorded. Only in this manner may shortfalls be detected.

### **Hyperalimentation**

Both oral and intravenous energy supplementation may be necessary.

### **Oral Supplementation**

Early 0-3rd day: Not given.

Intermediate 3rd-10th day: Build up oral intake to tolerance.

Late: Use liquid food additions. Precision high nitrogen diet; Ensure; Caloreen; Alburon; Liproclil.

The hen's egg: 7 g protein + 80 cal/egg.

Vitamins: Vit B Co, C and Vit K should be given to all patients.

Additional zinc is always required.

### **Intravenous Supplementation**

- Early phase 0-3rd day: Patients are intolerant to carbohydrates but not to fats. Give Intralipid 4-6 g/kg/24 hours.

Intermediate 3rd-10th day: Build up to tolerance using:

- carbohydrates
- amino acids
- fat.

Late: Monitor carefully. Use only peripheral vein sites and simultaneous carbohydrates and fat infusion solutions.

### **Nursing Care**

The doctor is just part of a team caring for the burn patient. Good nursing care is the deciding factor on which the outcome hinges.

### **Wound Care**

It is essential, as soon as it is permissible, that the burn patient be tubbed.

## **Hydrotherapy**

- Use a special bath or shower.
- Prevent nosocomial infection.
- Use Chlorox (Jik) 1-120 (or if strong 1-240).
- Rinse off well before new dressings are applied.
- Povidone iodine 1-1000 is an alternative.
- Hair is removed by shaving or clipping over 14 days old where the body has already formed a peripheral inflammatory cell barrier around the wound (early granulation) with demarcation of the slough.

## **Debridement Dressings**

### **The Triple Mix**

Balanced salt solution

Hydrogen peroxide 3%

Acetic acid 25%

*applied as*

- Halve's solution

Balance salt/acetic acid is 50/50, or

balance salt/hydrogen peroxide is 50/50.

- Third's solution

Balance salt/hydrogen peroxide/acetic acid is 33.3:33.3:33.3.

Cotton-gauze wet to dry dressings with the above solutions aid mechanical cleansing of the wound.

## **Splinting**

This is controlled by the physiotherapists.

## **Concluding Notes**

- Definition of invasive burn wound sepsis.

The proliferation and active invasion of the burn wound by micro-organisms in the quantity of 100,000 or more per gram of tissue.

- Between 15% and 30% of all thermal accidents occur during childhood.

- In most First World countries, burns are the third commonest cause of death in children between 1 and 15 years of life.

- Recognise the abused child.

- Triage criteria for major or critical cases.

Transporting a burnt patient over any distance is hazardous. Immediate transfer after accident when travelling time is less than 30 minutes during phase of compensated shock, is permissible but not advised.

Transferring a shocked patient is mismanagement. Resuscitate and treat till stable and refer only after day 3.

- Prognosis is directly related to TBSA.

Get skin cover.

Reduce the wound size.

Lethal area with 50% mortality:

1940 = 35%

1970 = 55-65%

1980 = 70%

Sepsis is the most important cause of death.

- Unburn the burn is the patient's plea.

In full thickness lesions this is unfortunately not possible. Where wounds of this type are sited on cosmetically exposed areas the victims will remain scarred for life. The final phase in the management of the burnt patient, is a rehabilitation period. This has been neglected in the past. The plight of the scarred patients has remained unrecognised by many a burn physician. Those that you have saved may remain disabled. Keep track of all your cases by establishing a strict outpatient follow-up programme and aim to return your patient to their rightful places in their own environment.

## **Comment**

### **Burn Injuries in Paediatric Patients**

**J H Louw**

Current therapeutic regimens all transfuse the calculated fluid during the first 24 hours post burn and not during the first 36 hours. We have adopted this policy. The rate of flow therefore is as follows: half of the calculated fluid volume is given during the first 8 hours post burn, and the other half during the next 16 hours.

Tetanus toxoid should be given to all patients who have not been immunized within the past 5 years or if their immunization programme is not available. In addition, major electrical burns exceeding 30% of total body surface area should also receive tetanus immunoglobulins.

It is debatable whether a heavily infected burn wound (bacterial count greater than  $10^4$  per gram of tissue) is necessarily a contraindication to surgical excision. Indeed, it may be the only way to resolve the invasive bacterial infection.

The current surgical tendency is to excise deep partial-thickness and full-thickness burn wounds as soon as the patient is haemodynamically stable. Sequential layer tangential excision to viable bleeding points, even to fat, while minimising the loss of viable tissue is the generally accepted technique today. The excised areas are then covered with skin grafts and, where indicated, with homografts as well (cadaver skin).