Chapter 37: Diseases of the oesophagus

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Embryology

The oesophagus and trachea first become identifiable as separate structures when the embryo is 22-23 days old, as a median ventral diverticulum in the developing foregut. Shortly thereafter, the primitive stomach appears as a fusiform enlargement immediately caudal to the diverticulum. The oesophagus develops from the short area between the tracheal diverticulum and the stomach. As the trachea and oesophagus elongate, ridges appear in the lateral walls. Fusion in the midline of these ridges separates the trachea from the oesophagus. The separation process commences caudally, proceeds cranially and is complete between days 34 and 36 of gestation. Elongation of the oesophagus also begins in the distal portion and is complete at 7 weeks (that is relative to the rest of the developing fetus).

The circular musculature of the oesophagus appears in the sixth week and by the end of that week innervation by the vagus nerve has commenced. During the seventh and eighth weeks, the epithelium of the oesophagus proliferates to such an extent that the lumen is virtually, but not completely, occluded. Initially, the epithelium is ciliated, but it is gradually replaced by stratified squamous epithelium.

Oesophageal atresia

This was uniformly fatal congenital abnormality until 1939 when the first two survivors were reported independently by Ladd and Levin (Levin, 1941; Ladd, 1944). Both infants required multiple procedures - cervical oesophagostomy, feeding gastrostomy, and subsequent reconstruction. The first success with primary repair of the defect, which paved the way to future developments, was described by Haight in 1941 (Haight and Towsley, 1943). It is now rare for an infant with oesophageal atresia to succumb from the anomaly, unless it is associated with additional complex life-threatening anomalies or extreme prematurity (Rickham, Stauffer and Cheng, 1977; O'Neill, Holcomb and Neblett, 1982; Louhimo and Lindahl, 1983).

Incidence

The incidence of abnormalities in oesophageal development is one in 3000-4000 live births (Myers, 1974). There does not appear to be any standard genetic pattern of inheritance, although the condition has been documented in siblings, in one and very occasionally in both twins and in two generations.

Embryology

The anomaly is thought to arise between the third and sixth weeks of intrauterine development. The precise cause and mechanism are unknown. Failure of complete separation of the foregut from the respiratory tract would appear to be the final common pathway for the development of the various types of defects. Oesophageal atresia may occur as an isolated anomaly but at least 40% of cases have additional malformations.
**Types of anomaly**

The variety and incidence of the different types of tracheo-oesophageal abnormalities are shown. These are as follows:

(1) oesophageal atresia with distal tracheo-oesophageal fistula - 87%;
(2) oesophageal atresia without tracheo-oesophageal fistula - 6-7%;
(3) oesophageal atresia with proximal tracheo-oesophageal fistula - 2%;
(4) oesophageal atresia with proximal and distal tracheo-oesophageal fistula - 1%;
(5) tracheo-oesophageal fistula without oesophageal atresia - 3-4%.

**Associated anomalies**

Additional congenital malformations are found in about one-half of infants with tracheo-oesophageal anomalies. The various systems affected are as follows (multiple defects occur in many patients) (Holder et al, 1964; German, Mahour and Woolley, 1976):

(1) cardiovascular defects - 34%;
(2) gastrointestinal (excluding anorectal) anomalies - 14%;
(3) genitourinary anomalies - 12%;
(4) anorectal malformations - 11%;
(5) skeletal defects - 11%;
(6) respiratory anomalies - 6%;
(7) genetic/chromosomal defects - 2%;
(8) miscellaneous anomalies - 10%.

The VATER complex of associated anomalies was described by Quan and Smith (1973). (The acronym stands for V = vertebral, A = anorectal, T-E = tracheo-oesophageal fistula and (o)esophageal atresia, R = radial and renal dysplasia.)

Ventricular septal defects are the single most common cardiac malformation (Greenwood and Rosenthal, 1976). Of the gastrointestinal anomalies, malrotation of the midgut occurs most frequently, followed by Meckel's diverticulum and duodenal atresia or stenosis. A variety of genitourinary anomalies occurs in association with oesophageal atresia, the most serious being renal agenesis (Potter's syndrome) which is incompatible with survival. The anorectal anomalies are equally divided between the suprlevator (high) and translevator (low) defects.
Diagnosis

Polyhydramnios occurs in approximately 30% of mothers of infants with oesophageal atresia. Antenatal ultrasound scan may be diagnostic of oesophageal atresia without fistula as a result of failure to demonstrate the presence of intragastric fluid. The infant, at birth, is 'excessively mucusy' and requires repeated suction as it is unable to swallow saliva. Failure to recognize the anomaly at this stage will expose the infant to choking episodes and aspiration pneumonitis with the first feed. The diagnosis is confirmed by passing a large calibre (no 10 French) firm catheter through the mouth and into the stomach. The position of arrest of the tube is confirmed on a chest X-ray. If the tube enters the stomach, there is no oesophageal atresia. In the majority of cases with this condition, the tube cannot be advanced more than 10 cm beyond the lower gum margin. It is important to include the abdomen on the original X-ray in order to assess the presence of intestinal gas shadows. Gas within the gastrointestinal tract implies the presence of a distal tracheo-oesophageal fistula while the distribution of the gas may indicate an additional intestinal anomaly, for example duodenal atresia or malrotation. The chest radiograph should be assessed for pulmonary pathology and the configuration of the heart shadow may be indicative of cardiac defects, for example, Fallot's tetralogy.

Management

Treatment of infants with oesophageal atresia should be concentrated in large centres where the surgical expertise, supportive services (anaesthesia, radiology, pathology) and specialized nursing care are available. Transfer to such centres should be effected promptly without exposing the infant to the risks of aspiration pneumonitis. The infant is transported in a portable incubator either in the 45° upright or in the prone position in order to discourage reflux of gastric juice in the distal tracheo-oesophageal fistula, while continuous suction of the upper pouch prevents aspiration of saliva (Spitz, Wallis and Graves, 1984). Definitive repair may have to be postponed in the presence of aspiration pneumonitis which generally responds very rapidly to broad-spectrum antibiotics and vigorous chest physiotherapy (Koop, Schnaufer and Broennie, 1974; Randolph, Altman and Anderson, 1977; Grosfeld and Ballantine, 1978; Ito, Sugito and Nagaya, 1984). In the infant with severe respiratory distress requiring mechanical ventilatory support, consideration should be given to emergency ligation of the distal tracheo-oesophageal fistula to facilitate respiratory support and to prevent overdistension of the stomach and intestine (Jones et al, 1980; Filston et al, 1982).

The operative procedure is carried out under general endotracheal anaesthesia. Access is ideally via a right posterloateral extrapleural approach through the fourth or fifth intercostal space (deLorimier and Harrison, 1985). After dividing the azygos vein, the distal oesophagus is identified and traced proximally to the fistulous entrance into the trachea. Mobilization of the distal oesophagus should be kept to a minimum as the blood supply to it is segmentally derived directly from small aortic branches and excessive devascularization would expose the anastomosis to impaired healing. The fistula is divided and the tracheal defect closed with fine interrupted non-absorbable sutures (5.0 Prolene). The proximal blind end of the oesophagus is identified and mobilized in the apex of the chest as extensively as required to effect an anastomosis with as little tension as possible. The end-to-end anastomosis is performed using a single layer of full thickness 5.0 or 6.0 polyglycolic acid or Prolene sutures. A Livaditis
oesophagomyotomy and/or the use of mechanical ventilation for a few days postoperatively may be effective in reducing the incidence of anastomotic complications.

The passage through the nose of a fine silastic transanastomotic tube into the stomach or the fashioning of a gastrostomy will allow early enteral feeding postoperatively.

A contrast oesophagogram performed between 7 and 10 days postoperatively will determine the anastomotic integrity following which oral feeds may be introduced.

Patients with a long-gap between the proximal and distal segment (particularly those with an isolated oesophageal atresia) require special attention. The alternative approaches available are to delay repair pending differential growth of the oesophageal segments towards each other (3-6 months), or to perform a cervical oesophagostomy and carry out an oesophageal substitution (deLorimier and Harrison, 1986) at a later stage - colonic interposition (Waterston, 1969), gastric tube (Anderson and Randolph, 1978), or gastric transposition (Spitz, 1984).

**Results and prognosis**

A total of 148 infants with tracheo-oesophageal anomalies were treated at The Hospital for Sick Children, London in the period 1980-1985. The infants were allocated into risk groups according to the definitions of Waterston, Bonham-Carter and Aberdeen (1962, 1963) (Table 37.1). Group C infants were subdivided into those with associated anomalies compatible or incompatible with survival. The survival rates in this series are shown in Table 37.2.

### Table 37.1 Risk groups for children with tracheo-oesophageal anomalies

<table>
<thead>
<tr>
<th>Group</th>
<th>Birthweight (g)</th>
<th>Associated anomaly</th>
<th>Pneumonia</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>&gt; 2500</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>B</td>
<td>1800-2500</td>
<td>None or moderate</td>
<td>±</td>
</tr>
<tr>
<td>C</td>
<td>&lt; 1800</td>
<td>Severe and/or multiple</td>
<td>±</td>
</tr>
</tbody>
</table>

### Table 37.2 Survival rates of 148 children with tracheo-oesophageal anomalies

<table>
<thead>
<tr>
<th>Group</th>
<th>Number</th>
<th>Percentage</th>
<th>Survivors</th>
<th>Percentage</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>66</td>
<td>45</td>
<td>66</td>
<td>100</td>
<td>88.7%</td>
</tr>
<tr>
<td>B</td>
<td>35</td>
<td>24</td>
<td>30</td>
<td>86</td>
<td>88.7%</td>
</tr>
<tr>
<td>C1</td>
<td>41</td>
<td>28</td>
<td>30</td>
<td>73</td>
<td>88.7%</td>
</tr>
<tr>
<td>C2</td>
<td>6</td>
<td>4</td>
<td>0</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

Anastomotic complications were most frequent when silk was used for the repair. Anastomotic leaks occurred in 21% of cases (24 patients) of which all but four healed on conservative treatment. Anastomotic strictures developed in 17% but the vast majority
responded to one or two dilatations. Recurrent tracheo-oesophageal fistulae were identified (Ein et al, 1983) in 12% of infants and all required surgical closure.

Other complications included severe gastro-oesophageal reflux (20%) and tracheomalacia (Wailoo and Emery, 1979) (16%). These were responsible for life-threatening apnoeic attacks or recurrent pneumonia. Nissen fundoplication (Ashcraft et al, 1977; Jolley et al, 1980) and aortopexy (Filler, Rosillo and Lebowitz, 1976; Benjamin, Cohen and Glasson, 1976) respectively were dramatically successful in alleviating the symptoms.

Swallowing difficulties may persist for many years as a result of the inherent oesophageal dysmotility, affecting the distal segment in particular (Laks, Wilkinson and Schuster, 1972). The infant gradually learns to cope with this problem unless there is an associated anatomical defect (anastomotic stricture, gastro-oesophageal reflux, distal oesophageal stenosis). These infants are also prone to recurrent respiratory infections during the first few years of life (Martin and Alexander, 1985).

Mortality in oesophageal atresia is directly related to the severity of associated congenital anomalies particularly cardiac defects (Koop, Schnaufer and Broennie, 1974; Rickham, Stauffer and Cheng, 1977; Myers, 1979; Holder, 1986).

Achalasia

Achalasia is a motility disorder of the oesophagus characterized by an absence of peristalsis and a failure of relaxation of the lower oesophageal sphincter (Henderson, 1966; Payne and King, 1983).

Incidence

Achalasia is rare in children (Azizkhan, Tapper and Eraklis, 1980). The incidence is one per 100,000 and approximately 5% of all patients are symptomatic before the age of 15 years (Herman and Moersch, 1929; Olsen et al, 1951). There are few reports of achalasia occurring in siblings (London et al, 1977; Stoddard and Johnson, 1982) and it has been reported in association with a number of syndromes, for example Riley-Day. Evidence for a familial incidence of the disease is lacking. Both sexes are equally affected.

Aetiology and pathogenesis

The aetiology of achalasia is unknown. Oesophageal dysmotility also occurs in Chagas' disease, scleroderma, oesophageal atresia, diabetes and secondary to gastro-oesophageal reflux, but the unique feature of achalasia is the constantly non-relaxing lower oesophageal sphincter.

Numerous theories exist regarding the pathogenesis of the condition, the primary defect being described variously as neurogenic, myogenic and hormonal. There is evidence suggesting that it is the result of an abnormality of parasympathetic innervation. Absence of ganglion cells in the myenteric plexus in the dilated portion of the oesophagus with normal ganglion cells in the distal non-dilated segment have been described (Gallone, Peri and Galliera, 1982). This, however, is not a constant feature and is reflected in the variable reports of the histopathology of some of the specimens of oesophageal muscle. These range from the
total absence of ganglia, to the presence of normally ganglionated muscle or abnormal ganglion cell morphology. Histochemical staining of acetylcholinesterase may reveal the presence of ganglion cells and nerve trunks in the myenteric plexus, although their numbers are slightly reduced. Recent reports using electron microscopy (Friesen, Henderson and Hanna, 1983) and intestinal polypeptide hormonal assay support the theory that this is a neurogenic disorder (Aggestrup et al, 1983).

**Diagnosis**

**Symptomatology**

The principal symptoms of achalasia in childhood consist of vomiting, dysphagia, chest pain and weight loss. Dysphagia with the sensation of food sticking in the lower oesophagus and postprandial vomiting are the most frequent presenting symptoms. Retrosternal or epigastric pain occurs in one-third of the patients and in a few cases it is the primary presenting symptom. Weight loss of varying extent occurs in one-half of the patients. Nocturnal regurgitation may give rise to respiratory symptoms and recurrent respiratory infections may be experienced. A constant feature is the prolonged delay in establishing the precise diagnosis. The average duration of symptoms prior to diagnosis is 24 months and many children are treated for long period for 'cyclic vomiting' or for 'anorexia nervosa' before achalasia is diagnosed.

**Radiological features**

The plain chest radiograph may show a dilated food-filled oesophagus with an air-fluid level in the distal third. In addition, there may be radiological signs of repeated aspiration pneumonitis. The chief characteristics on barium oesophagogram are a dilated oesophagus, the absence of a stripping wave, uncoordinated oesophageal contractions and obstruction at the oesophagogastric junction with prolonged retention of barium in the oesophagus. Failure of relaxation of the lower oesophageal sphincter leads to classical rat-tail deformity of funnelling and narrowing of the distal oesophagus.

**Endoscopy**

Oesophagoscopy contributes little to the diagnosis, but retained food may be found within the dilated oesophagus. The main value of endoscopy is to exclude organic causes of obstruction in the oesophagus.

**Oesophageal manometry**

The diagnosis of achalasia is best confirmed by oesophageal motility studies using a constantly perfused catheter technique. The criteria for the diagnosis are:

1. a high pressure (> 30 mmHg) lower oesophageal sphincter zone;
2. failure of the lower oesophageal sphincter to relax in response to swallowing;
3. absence of propulsive peristalsis;
4. uncoordinated tertiary contractions in the body of the oesophagus.
Treatment

Three treatment options are available for the management of achalasia - pharmacological manipulation, forceful dilatation and oesophageal myotomy with or without the addition of an antireflux procedure.

Pharmacological treatment

The manipulation of oesophageal motility disorders using pharmacological and dietary measures has been disappointing. Recent reports of the use of isosorbide dinitrate and nifedipine have been more encouraging (Blackwell, Holt and Heading, 1981; Gelfond, Rosen and Gilat, 1982). Nifedipine is a calcium entry blocker and since calcium ions are directly responsible for the activity of myofibrils and consequently the tension generated, their use in reducing the pressure in the lower oesophageal sphincter in achalasia, or for the vigorous oesophageal contractions which occur in diffuse oesophageal spasms, seems logical. Prostaglandin E₂ has also been employed with some success. Its value in the long-term treatment of achalasia remains to be proven.

Forceful dilatation

Good palliation may be obtained by forceful dilatation. The most commonly used dilator consists of a single bag of fixed diameter which is inflated with water (Plummer) or air (Browne-McHardy, Rider-Moeller).

Forceful dilatation has been advocated as the treatment of choice in adults. Fellows, Ogilvie and Atkinson (1983) showed that following pneumatic dilatation in adults only 10% of patients subsequently required cardiomyotomy. In general, the results of pneumatic dilatation in children have been variable. Success rates, ranging from 40 to 60% have been reported in children (Payne, Ellis and Olsen, 1961; Berquist et al, 1983). The aim of forceful dilatation is to disrupt the muscle fibres of the lower oesophageal sphincter. There is, however, no evidence that the muscle fibers tear rather than stretch. Vantrappen and Janssens (1983) were unable to distinguish histologically between sphincter segments in dogs and monkeys subjected to forceful dilatation (Azizkhan, Tapper and Eraklis, 1980). A report of 899 adult patients treated at the Mayo Clinic (Payne and King, 1983) concluded that myotomy was more successful and safer than dilatation, poor results being obtained twice as frequently following dilatation as after myotomy. The incidence of perforation following pneumatic dilatation varies from 1 to 5% (Bennett and Hendrix, 1970; Vantrappen et al, 1971; Fellows, Ogilvie and Atkinson, 1983).

Surgical procedure

Cardiomyotomy as originally described by Heller in 1914 is the basis of all surgical procedures. The controversies concern the length of the oesophageal myotomy, the distance which the myotomy extends onto the stomach and the necessity for including an additional antireflux procedure (Jara et al, 1979; Ballantine, Fitzgerald and Grosfeld, 1980; Buick and Spitz, 1985).
Myotomy may be performed either via an abdominal approach or by a left thoracotomy through the bed of the seventh or eighth rib. Taking care to protect the vagus nerve, a 5-7 cm incision is made through the muscle of the distal oesophagus. The incision is deepened down through the muscle to the mucosa. Particular care must be taken to avoid opening the mucosa. The muscle wall is dissected laterally off the mucosa so that at least half the circumference of the oesophagus is exposed and the mucous pouts freely through the incision. Ellis, Gibb and Crozier (1980) insist that to avoid reflux the incision should not extend for more than a few millimetres onto the stomach. If a longer incision is made onto the stomach wall, an antireflux procedure in the form of a short lax (floppy) Nissen fundoplication should be added to the procedure.

Oesophageal foreign bodies

Over 90% of ingested foreign bodies pass uneventfully through the gastrointestinal tract. Of those that are held up, this mostly occurs in the oesophagus (Spitz, 1971).

Anatomy

There are four sites of anatomical narrowing in the oesophagus where foreign bodies are likely to impact. These are the postcricoid region, the level of the aortic arch, the level of the left main bronchus, and the level of the diaphragmatic hiatus (Slovis, Tyler-Werman and Solightly, 1982). Eighty per cent of impacted foreign bodies are help up at the level of the cricopharyngeus. Impaction may also occur at sites of pathological narrowing, for example strictures secondary to peptic oesophagitis, corrosive strictures, anastomotic strictures of congenital stenosis.

Clinical presentation

In the majority of cases there will be a clear history of ingestion of a foreign object. The child will present with acute symptoms of coughing, choking, excessive salivation, dysphagia or vomiting (Nandi and Ong, 1978; O'Neill, Holcomb and Neblett, 1983). If the foreign body remains impacted, adaptation may occur and the child will select foods which can be managed without producing symptoms (Giordano et al, 1981; O'Neill, Holcomb and Neblett, 1983). In other cases, there is no definite history or acute symptomatology and presentation occurs in an obscure fashion, for example chronic respiratory symptoms (stridor, wheezing, recurrent pneumonia) (Goldsher, Eliacher and Joachims, 1978), anorexia, haematemesis or pyrexia of unknown origin.

Diagnosis

The diagnosis will be confirmed on straight posteroanterior (PA) and lateral X-ray of the chest in those cases with a radiopaque foreign body ingestion. Further investigations are essential in patients in whom a foreign body is suspected but not evident on the plain X-ray. In these cases, a barium oesophagogram (Sharp, 1986), computerized tomography (Crenshaw, 1977) or endoscopy should be undertaken. In particular, it is the radiolucent foreign object which causes diagnostic difficulties. An aluminium can top can easily escape detection on plain X-ray (Spitz and Hirsig, 1982).
Treatment

All foreign bodies lodged in the upper third of the oesophagus as well as sharp objects in the lower oesophagus should be removed using direct endoscopy. Rounded objects may be dislodged by means of a Foley balloon catheter. Rounded objects in the lower half of the oesophagus may be observed for 24-48 hours in anticipation of their spontaneous passage into the stomach following which passage through the rest of the gastrointestinal tract can be confidently predicted (Spitz, 1971). Associated strictures of the oesophagus should be dilated and subsequently subjected to further investigation and definitive treatment.

Complications

Problems usually arise in proportion to the duration of impaction (Clerf, 1975). Possible complications include ulceration, stricture formation, tracheo-oesophageal fistula, erosion through the wall of the oesophagus with mediastinal abscess or penetration into the major blood vessels (Sharp, 1986). Perforation may also occur during attempts at endoscopic removal of a sharp object.

Corrosive injury to the oesophagus

Accidental ingestion of caustic substances has become relatively uncommon as a result of government legislation regulating their use in commercially available drain cleaners (Leape, 1986).

Pathophysiology

Caustic soda (sodium hydroxide) ingestion can cause severe injury to the oropharynx, oesophagus or stomach. The strong alkali rapidly penetrates the body tissues producing an intense acute inflammatory reaction and oedema. If the concentration of the solution is high, transmural penetration occurs with resulting destruction of the musculature of the oesophagus, penetration into the perioesophageal tissues with mediastinitis or frank perforation of the oesophagus. The acute phase is followed by sloughing of the necrotic tissue and replacement by granulation tissue. The final outcome varies from complete resolution in the mild case to extensive fibrosis of the entire oesophagus in the severely damaged cases. The extent and severity of the injury are directly related to the concentration of the lye ingested (liquid lye is more damaging than the granular form), the quantity ingested and the duration of contact (Ashcraft and Padula, 1974).

Clinical presentation

There is extensive oedema and swelling of the mouth and lips and the child is unable to swallow. Chest pain is indicative of mediastinitis while abdominal pain will occur if large quantities of lye reach the stomach. There may be haematemesis, dyspnoea, stridor and the other respiratory symptoms develop as a consequence of the resulting oedema or from direct laryngeal injury. Fibrosis of the lips and temporomandibular joint may develop as a result of severe oropharyngeal burns.
Diagnosis

It is important to ascertain whether the lye was actually ingested or whether it entered the oral cavity only. Early endoscopic examination, within 12-24 hours of the injury, should be undertaken to determine whether or not the oesophagus is affected. Assessment of the extent of oesophageal burn is not possible at this early stage and if the endoscopic examination is terminated once evidence of oesophageal injury is encountered, the risk of perforation is minimal (Leape, 1986).

An early contrast oesophagogram will reveal the extent of the injury, determine the presence of a perforation and act as a baseline for evaluating future stricture formation.

Treatment

Emergency

Vomiting should not be induced. The lye should be neutralized by ingestion of milk or, if this is not available, by water. Admission to hospital of all suspected cases is imperative.

Continued management

Initial treatment should consist of broad-spectrum antibiotics, intravenous fluids and prednisone 2 mg/kg per day (Haller and Bachman, 1963). If on endoscopy no oesophageal injury is found, treatment is stopped and the patient is discharged. If a burn is found, antibiotics are continued for 10 days and steroids for 3 weeks. Oral feeds are commenced as soon as the child is able to tolerate fluids. A gastrostomy for feeding purposes may be necessary in cases with severe burns. In these patients the opportunity should be taken to pass a string through the oesophagus to act as a guide for future dilatations.

A repeat oesophagogram is carried out 3 weeks after ingestion of the lye and oesophageal dilatations are commenced if a stricture is found. The dilatations are repeated at regular intervals until the stricture is eliminated. Ninety per cent of oesophageal strictures will respond to dilatation, the remaining cases will require oesophageal replacement.

Gastro-oesophageal reflux

Incompetence of the lower oesophageal sphincter is a common occurrence in newborn infants. In the vast majority of these infants maturation of the sphincter mechanism develops during the first year of life as the infant assumes the sitting and then the upright position (Carre, 1959).

Pathophysiology

Gastro-oesophageal reflux may or may not be associated with an anatomical hiatus hernia. A sliding hiatus hernia is generally associated with reflux while a rolling hernia (paraoesophageal) causes symptoms by virtue of its intrathoracic volume or secondary to complications such as haemorrhage or perforation as a consequence of peptic ulceration or
volvulus of the stomach. A paraoesophageal hernia always requires surgical correction (Ellis, Crozier and Shea, 1986).

Acid-pepsin reflux into the lower oesophagus results in a chemical inflammation of the squamous mucosa which is ill equipped to resist the digestive enzymes. In the early stages there is an inflammatory cell infiltration with erythema of the mucosa. With continuing reflux the mucosa becomes friable and bleeds on contact. Later, ulceration develops which may proceed to stricture formation as fibrous tissue is laid down as a consequence of transmural damage.

Acid-pepsin reflux --> Erythema --> Friability of the mucosa --> Ulcerative oesophagitis --> Stricture formation.

Clinical presentation (Ramenofsky, 1986)

Early infancy

The infant presents with recurrent vomiting which may be projectile and may even mimic pyloric stenosis. The vomitus generally contains ingested milk only, but haematemesis in the form of fresh blood or of 'coffee-grounds' may occur as a result of ulcerative oesophagitis. With repeated vomiting, the infant fails to gain weight at the expected rate and develops severe constipation. Presentation with recurrent respiratory infections or apnoeic attacks will be discussed under aspiration syndromes

Later childhood

Persistent vomiting is still the major symptom in older children, but the problem may only occur in the form of night vomiting of mucus which is noted on the pillow in the morning. Heartburn caused by oesophagitis and dysphagia from ulcerative oesophagitis or stricture formation assume more prominence at this age. Hypochromic microcytic anaemia may develop as a consequence of constant slow blood loss into the oesophagus from ulcerative oesophagitis. Asthma or recurrent respiratory infections may develop from recurrent aspiration of gastric content.

Associated anatomical defects

Gastro-oesophageal reflux is more common in patients with corrected oesophageal atresia, diaphragmatic hernia or defects of the anterior abdominal wall, for example exomphalos. Precipitating causes or malrotation of the midgut and other causes of gastric outlet obstruction, for example pyloric stenosis or antral dysmotility.

Neurological abnormalities

The association between gastro-oesophageal reflux and severe mental retardation and other neurological abnormalities has only recently been appreciated. An estimated 10% of retarded children in institutional care manifest vomiting as a major problem. The tendency is to ascribe the vomiting to psychological causes and this has led to prolonged delay in diagnosis exposing the child to the development of complications from the reflux. Severe
failure to thrive, iron deficiency anaemia, recurrent bouts of pneumonia and strictures frequently occur in these children (Abrahams and Burkitt, 1970; Sondheimer and Morris, 1979; Spitz, 1982).

**Aspiration syndromes**


**Unusual presentation**

Abnormal head and neck contortions in association with ulcerative oesophagitis were first described by Sandifer in 1964 (Kinsbourne, 1964). The range of symptoms has been extended to torticollis, tics, dystonia and irritability. Protein-losing enteropathy may occur as a result of ulcerative oesophagitis.

**Diagnosis**

A range of investigations has been applied to establish the presence and severity of gastro-oesophageal reflux (Darling, 1975; McCauley, Darling and Leonidas, 1979).

**Barium oesophagogram**

The patient should be kept warm and comfortable during the examination. No attempt should be made to induce reflux by means of abdominal compression or other provocative manoeuvres. The patient is screened in the lateral, supine and prone positions. Particular attention is paid to the following details:

1. anatomy of the oesophagus and the presence of strictures, ulcerative oesophagitis, abnormal narrowing or displacement should be noted;

2. peristaltic activity: primary contractions of the oesophagus occur during deglutition, secondary waves clear any residual content and may be normal, while tertiary waves are feeble attempts at contraction which are generally incoordinated and are always abnormal;

3. hiatus hernia: the presence of a sliding or paraoesophageal hernia will become evident during the examination;

4. the degree of gastro-oesophageal reflux is graded according to the highest level of the refluxed barium content:
grade 1 - distal oesophagus;
grade 2 - proximal/thoracic oesophagus;
grade 3 - cervical oesophagus;
grade 4 - continuous gastro-oesophageal reflux;
grade 5 - aspiration of barium into the tracheobronchial tree;
grade 6 - delayed reflux on the 30 minute X-ray film.

The rate of clearance of refluxed barium should also be noted;

(5) evidence of gastric outlet obstruction or duodenal malrotation.

**Oesophageal pH monitoring**

Measurement of the pH in the distal oesophagus by means of an intraluminal pH probe is currently the most accurate method of documenting reflux (Jolley et al, 1978, 1979; Boix-Ochoa, Lafuente and Gil-Vernet, 1980; Sondheimer, 1980; Ramenofsky and Leape, 1981). The recordings are monitored continuously over a 24-hour period. pH levels below 4 are regarded as significant and during the 24-hour recording the following parameters are examined:

1. number of episodes when pH falls below 4;
2. duration of each reflux episode;
3. number of reflux episodes lasting more than 5 minutes;
4. total duration of reflux expressed as a percentage of recording time.

**Oesophageal manometry**

This is best measured by continuous infusion open-tipped catheters. A high pressure zone is normally present at the lower oesophageal sphincter. There is good correlation between reflux and decreased lower oesophageal pressure in adults, but measurements of the sphincter pressure in children is much more variable. Euler and Ament (1977) suggested that lower oesophageal pressure values may be useful in predicting those cases which would eventually require surgical treatment.

**Endoscopy and oesophageal biopsy**

Endoscopic examination of the oesophageal mucosa is required to document the degree of oesophagitis and should be supplemented by endoscopic biopsy to provide histopathological grading of inflammatory cell infiltration. Four grades of oesophagitis may be found at endoscopy:

grade I - erythema of the mucosa
grade II - friability of the mucosa
grade III - ulcerative oesophagitis
grade IV - stricture formation.
Scintiscanning

Technetium ($^{99}$Tc) sulphur colloid scans have been used in an attempt to define the presence of gastro-oesophageal reflux over a longer period than during the barium examination (Christie and Rudd, 1978; Jona, Sty and Glickich, 1981). They also provide evidence of pulmonary aspiration in delayed scans taken 24 hours later. Aspiration may also be confirmed by the presence of lipid-laden macrophages in tracheal aspirates.

Management

Medical treatment

Feeds

Small frequent feeds thickened with cornflour or Nestargel should be given to infants with reflux (Herbst, 1986).

Position

The 60° upright position used to be recommended but it was found that the infant slumps forward in this position, thus increasing the intra-abdominal pressure. The position most frequently adopted now is the 30° head elevated prone position (Orenstein and Whittington, 1983).

Antacids

Simple antacids such as magnesium or aluminium hydroxide are more effective in the liquid form. Antacids combined with alginic acid (Gaviscon) form a foam level in the fundus of the stomach which discourages reflux (Cucchiara et al, 1984).

Hydrogen receptor antagonists (cimetidine, ranitidine)

These agents are particularly useful in the presence of severe oesophagitis in promoting healing and in achieving symptomatic relief of heartburn (Cucchiara et al, 19894).

Other drugs

Drugs which increase the lower oesophageal pressure, for example metaclopramide, bethanechol, domperidone etc, also stimulate gastric emptying (McCallum et al, 1983).

Surgical treatment

The indications for a surgical approach are (Spitz and Kirtane, 1985):

(1) established oesophageal stricture;

(2) failure of conservative medical measures:
(a) in the presence of an anatomic anomaly, for example oesophageal atresia, malrotation, exomphalos etc;

(b) in the presence of associated neurological damage. The response to medical treatment is notoriously poor and the additional nursing burden imposed by repeated vomiting adds significantly to the social stress of the family;

(c) apnoeic episodes and repeated respiratory infections which do not respond promptly;

(d) failure to thrive in spite of adequate treatment.

With the exception of near-miss sudden infant death syndrome, failure of medical treatment should not be considered until the treatment has been attempted for at least 3 weeks in hospital or 6-12 weeks at home.

The most widely adopted surgical procedure for the correction of reflux is Nissen fundoplication. This involves wrapping the fundus of the stomach around the distal 2-4 cm of oesophagus. The wrap should be lax and short to allow free passage of food through the oesophagus and to permit the patient to eructate, while at the same time preventing reflux. Postoperative complications are more frequent in patients with established strictures and in debilitated children with severe retardation. Referral for surgery is invariably delayed in those children and earlier surgery may avoid many of the postoperative problems (Spitz, 1982).

**Congenital oesophageal stenosis**

Stenosis of the oesophagus most commonly arises from acquired lesions, for example reflux oesophagitis, corrosive ingestion, foreign body impaction or secondary to surgical resection and anastomosis. Congenital stenosis is a rare anomaly which may be either caused by a membranous web or diaphragm, or may arise as a result of intramural deposits of tracheobronchial cartilaginous tissue. The latter pathology has been most frequently reported in association with oesophageal atresia and/or tracheo-oesophageal fistula (Nishina, Tsuchida and Saito, 1981).

**Clinical features**

In the presence of a complete web, presentation is similar to that of oesophageal atresia with symptoms occurring on the first day of life. In other cases, symptoms may develop at any stage of life through to adulthood but generally arise in early infancy. The symptoms include dysphagia, vomiting with food ingestion, failure to thrive, recurrent respiratory infections and foreign body impaction.

**Diagnosis**

Gastro-oesophageal reflux as a cause of the stenosis should always be excluded. The barium oesophagogram will identify the site of the lesion. Congenital webs are most commonly located in the middle third of the oesophagus and appear as shelf-like projections within the oesophageal lumen. Tracheobronchial remnants are generally located in the lower
third of the oesophagus or at the gastro-oesophageal junction and they cause sharp narrowing
at this point. The precise nature and anatomical location of the lesion should be confirmed
on endoscopic examination.

Treatment

Dilatation alone may be sufficient for many cases of oesophageal webs. Surgical
resection of tracheobronchial cartilaginous tissue is generally recommended (Scherer and
Grosfeld, 1986).