MANAGEMENT OF TRACHEOMALACIA IN ASSOCIATION WITH CONGENITAL TRACHEO-OESOPHAGEAL FISTULA

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Tracheomalacia occurs commonly in association with oesophageal atresia and tracheo-oesophageal fistula (Wailloo and Emery, 1979). Tracheomalacia is a term which describes weakness of the tracheal wall which may cause sufficient narrowing of the lumen to produce ventilatory symptoms. Recurrent chest infections and apnoeic or cyanotic spells leading to cardiac arrest may occur after oesophageal repair. The purpose of this case report is to stress the importance of the association between tracheomalacia and congenital oesophageal atresia—an association which is not well documented in the anaesthetic literature—and to outline the anaesthetic management of severe cases which require surgical treatment by tracheopexy.

CASE REPORT

A 3-kg female infant was delivered vaginally at term. It was noted that a feeding tube failed to pass down the oesophagus and a presumptive diagnosis of oesophageal atresia was made. No other anomalies were present; there were no ventilatory problems, stridor was absent, and the cry was normal. A double lumen Replögle tube was passed into the upper pouch to provide some protection from aspiration, and the infant was transferred to the Sheffield Neonatal Surgical Unit. Subsequently, a right thoracotomy was performed. Through an extrapleural approach, a low tracheo-oesophageal fistula was ligated and the two ends of the oesophagus were anastomosed with a transanastomotic feeding tube in place.

Immediately before surgery, atropine 0.06 mg was given i.v. and awake intubation of the trachea was performed with a 3.0-mm uncuffed Magill tube. Anaesthesia was maintained with nitrous oxide in oxygen through a Jackson-Rees modification of Ayre’s T-piece at a flow of 3 litre min⁻¹. When it had been ensured that there was no significant leak into the stomach during positive pressure ventilation, atracurium 1 mg was administered i.v.; manual ventilation of the lungs was performed for the duration of the operation with increments of atracurium, given as required. There were no significant anaesthetic or surgical problems and on completion of surgery, the infant resumed adequate spontaneous ventilation without the need for antagonism of residual neuromuscular blockade. Extubation was performed without incident.

The infant remained well until the 4th day after operation, when there was an acute episode of cyanosis. A chest x-ray revealed a left sided pneumothorax and her condition improved following the administration of oxygen, nasopharyngeal suction and insertion of a chest drain. During the following days there were further acute episodes of cyanosis which were often associated with feeding and were preceded by clinical evidence of upper airway obstruction. One episode was associated with severe bradycardia which required external cardiac compression and oxygen by positive pressure ventilation. Between

SUMMARY

Tracheomalacia is commonly associated with oesophageal atresia and tracheo-oesophageal fistula. Severe cases may present with life threatening cyanotic and apnoeic attacks following surgical repair of the oesophageal atresia. The anaesthetic, and surgical management (by tracheopexy), of such a case are described.
episodes the infant appeared normal. Barium studies revealed no evidence of narrowing at the anastomotic site which could lead to pulmonary aspiration, and there was no evidence that the fistula had recurred.

A systolic murmur had been heard at the left sternal edge. Chest x-ray and cardiac ultrasound revealed no abnormalities, and a cardiac lesion was considered unlikely after cardiological consultation. However, a lateral chest x-ray revealed narrowing of the upper portion of the trachea with evidence of compression during barium swallow. A diagnosis of tracheomalacia was suggested.

Provisional arrangements were made to perform tracheopexy and the infant was transferred to theatre after premedication with atropine 0.1 mg i.m. After the application of a precordial stethoscope, ECG electrodes and arterial pressure cuff, the insertion of a rectal thermometer probe and cannulation of a peripheral vein, anaesthesia was induced with nitrous oxide and halothane in oxygen. Bronchoscopy was performed using a Storz bronchoscope (internal diameter 2.5 mm) while anaesthesia was maintained by attaching an Ayre’s T-piece system to the side arm of the bronchoscope and insufflating oxygen and halothane. The findings were that there was intermittent collapse at the lower end of the trachea and the diagnosis of tracheomalacia was confirmed.

After removal of the bronchoscope, the trachea was intubated with an uncuffed, 3.0-mm reinforced silicon oral tube (Cory Brothers Ltd). Anaesthesia was maintained with nitrous oxide in oxygen delivered through a Jackson—Rees modification of Ayre’s T-piece as above. Again, manual ventilation of the lungs was performed for the duration of surgery with increments of atracurium, as necessary.

Tracheopexy was performed through a cervical approach. Through a low transverse cervical incision the strap muscles were divided and the manubrium split. The edges of the manubrium were retracted laterally and the isthmus of the thymus divided. The trachea was then revealed passing caudally and posteriorly deep to the innominate vein and right innominate artery (fig. 1). Three silk sutures were placed through the trachea into the posterior edge of the manubrium and tied sufficiently tightly to support its anterior wall. Additional supporting sutures were placed into the adventitia of the aortic arch and innominate artery. Thus the anterior wall of the trachea was drawn forwards towards the sternum. On completion of surgery the infant resumed spontaneous ventilation and extubation was performed. She was then returned to the surgical unit where she made an uneventful recovery and had no further cyanotic episodes.

**DISCUSSION**

Tracheomalacia can be defined as a condition in which there is weakness of the tracheal wall as a result of softening of the supporting cartilage and hypotonia of the myoelastic elements (Baxter and Dunbar, 1963). Tracheomalacia may be divided into two types: primary and secondary.

In primary tracheomalacia the condition is congenital. It may be an isolated finding or be associated with other congenital anomalies—cleft palate, choanal atresia and oesophageal atresia. In secondary tracheomalacia the condition is acquired. The common origin of the trachea and oesophagus from the primitive foregut makes it likely that in individuals in whom the oesophagus is developmentally abnormal, tracheal anomalies will co-exist such as the association between tracheo-oesophageal fistula and oesophageal atresia.

Following surgical correction of tracheo-oeso-
phageal fistula and oesophageal atresia, affected children may have stridor, recurrent cough, wheeziness, dyspnoea and even apnoeic attacks leading to cardiac arrest. Symptoms may be attributed to respiratory tract infection resulting from the inhalation of regurgitated contents, with little attention being paid to the possibility of tracheal anomalies accounting for the symptoms. Early reviews (table I) of surgical and anaesthetic management of oesophageal atresia and tracheo-oesophageal fistula failed to include tracheomalacia as a postoperative problem, although ventilatory complications were usually the commonest cause of morbidity and mortality. Dudley and Phelan (1976), in a follow-up of respiratory complications in long term survivors of oesophageal atresia, concluded that recurrent inhalation of milk and food consequent upon disordered oesophageal motility is the major factor causing respiratory complications. However, Waillloo and Emery (1979) showed an almost invariable association between tracheo-oesophageal fistula and tracheomalacia; 40 children with tracheo-oesophageal fistula were analysed post-mortem; only six tracheae were entirely normal: 75% had a reduction in length of cartilage; 60% showed an increase in the length of transverse muscle and 65% had a wider than average internal diameter. The fistula site could not be used as a marker for tracheal defects. The authors concluded that the effect of these changes is an alteration in the shape of the trachea so that the normal horseshoe shape is lost. This tends to make the trachea soft and floppy, potentiating antero-posterior collapse of the walls during coughing, ventilation and feeding where posterior compression of the trachea by food in the oesophagus may occur.

It has been suggested (Davies and Cywes, 1978) that tracheomalacia is associated with tracheo-oesophageal fistula because, at birth, the proximal oesophageal pouch, in contrast to the distal oesophageal remnant, has become dilated and its muscular wall hypertrophied. The proximal pouch exerts a compressive force on the trachea from early in fetal life, affecting the development of the tracheal wall. A second hypothesis, by the same authors, proposes that the presence of a fistula leads to loss of intratracheal pressure in the fetus causing abnormal development of the trachea. In the normal fetus the tracheobronchial tree is distended by fluid, provided there is a competent glottic mechanism. However, this splinting effect will be lost by drainage of fluid through a tracheo-oesophageal fistula. The tracheal wall then becomes more susceptible to the effects of external pressure.

The cause of cyanotic episodes in children following repair of oesophageal atresia and tracheo-oesophageal fistula can be difficult to determine: oesophageal stricture with aspiration, a recurrent or second fistula, gastro-oesophageal reflux, anastomotic leak, cardiac or neurological defect must all be excluded. A lateral chest x-ray

<table>
<thead>
<tr>
<th>Commonest cause of postoperative complications</th>
<th>Incidence of respiratory complications</th>
<th>Associated tracheomalacia</th>
</tr>
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<tbody>
<tr>
<td>Respiratory complications (including two deaths from convulsions and severe cyanotic episodes)</td>
<td>Not reported</td>
<td>Not reported</td>
</tr>
<tr>
<td>Respiratory complications and associated anomalies</td>
<td>Not reported</td>
<td>Not reported</td>
</tr>
<tr>
<td>Respiratory complications</td>
<td>Approximately 50%</td>
<td>Not reported</td>
</tr>
<tr>
<td>Respiratory complications (including 11 fatal cardiac arrests)</td>
<td>48%</td>
<td>Two cases of laryngomalacia</td>
</tr>
</tbody>
</table>

Table I. Reviews of surgical and anaesthetic management of oesophageal atresia and tracheo-oesophageal fistula
may reveal segmental narrowing of the trachea and screening of the trachea during barium swallow may reveal tracheal compression.

Confirmation of the diagnosis of tracheomalacia is made by bronchoscopy, where segmental tracheal narrowing is usually obvious on direct vision. Bronchoscopy is indicated in all children who develop recurrent apnoeic spells after repair of oesophageal atresia when causes other than tracheomalacia have been excluded. Benjamin, Cohen and Glasson (1976) treated 80 infants with congenital tracheo-oesophageal fistula: 21 developed ventilatory symptoms following ligation of fistula and oesophageal anastomosis; in 20 of these patients tracheomalacia was confirmed at bronchoscopy. All 20 infants had a characteristic brassy cough. The cases were classified as severe, moderate or mild. The seven patients in the severe group had upper respiratory tract obstruction with stridor, and cyanotic and apnoeic attacks which occurred in the first 2 months; four patients had one or more cardiac arrests.

The moderate group included three patients who developed symptoms during the first 2 months of life, while five remained symptom free until the 5th month. They developed stridor and wheezing with frequent respiratory infections, during which cyanotic episodes occurred.

The five mild cases had occasional respiratory infections with croup or bronchitis associated with sputum retention. Symptoms did not occur in this group until about the age of 6 months.

Five of the patients in the severe group were successfully treated by tracheopexy via thoracotomy; the patients in the moderate group required regular physiotherapy with home facilities for pharyngeal aspiration; the patients in the mild group were managed by intermittent chest physiotherapy. Filler, Rosello and Lebowitz (1976) also described three cases of life threatening anoxic spells caused by tracheal compression after repair of oesophageal atresia.

The majority of patients with tracheomalacia associated with oesophageal atresia are asymptomatic or have recurrent minor chest infections which improve with age. However, patients who develop life threatening "death attacks" or require emergency and prolonged hospitalization can be successfully treated by tracheopexy. Tracheopexy is a relatively new surgical procedure. Although the operation can be carried out via thoracotomy, the cervical approach (Vaishnev and Mackinnon, 1986) provides the surgeon with easier and more direct access to the trachea. It allows precise insertion of sutures and, as the lung is not retracted, no impairment of ventilation. Five patients have been successfully treated in our hospital over recent years (table II) by this approach. During the procedure anaesthetic problems include kinking of the tracheal tube, obstruction of the great vessels and haemorrhage. The use of a kink-resistant tube, monitoring of breath sounds by precordial stethoscope and manual ventilation of the lungs are essential to the safe management of these patients, and permit the early detection of airway and ventilatory problems during surgery. Careful surgical placement of sutures in the adventitia of the great vessels ensured that blood loss was usually small: of the five patients only one child required blood transfusion. Difficulty with intubation was encountered in one patient. One child developed

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age at tracheopexy</th>
<th>Symptoms</th>
<th>Intraoperative comments</th>
<th>Postoperative course</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>4 years</td>
<td>Recurrent chest infection. Mouth-to-mouth resuscitation at home</td>
<td>No problems</td>
<td>Uneventful</td>
</tr>
<tr>
<td>2</td>
<td>3 months</td>
<td>Cyanotic attacks with feeding. Apnoeic attacks</td>
<td>Difficult intubation</td>
<td>Uneventful</td>
</tr>
<tr>
<td>3</td>
<td>3 months</td>
<td>Apnoeic attacks</td>
<td>Required blood transfusion</td>
<td>Uneventful</td>
</tr>
<tr>
<td>4</td>
<td>15 months</td>
<td>Apnoeic attacks, cyanotic attacks during feeds</td>
<td>No problems</td>
<td>Required 4 days ventilation because of development of pneumonia Uneventful</td>
</tr>
<tr>
<td>5</td>
<td>2 months</td>
<td>Apnoeic attacks</td>
<td>No problems</td>
<td>Uneventful</td>
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pneumonia in the postoperative period and required ventilation for 4 days. In all other patients the trachea was extubated immediately after surgery and the patient nursed in oxygen enriched humidified air. This was an added precaution because of the possible development of airway oedema caused by bronchoscopy before undertaking tracheopexy. In another review, Schwartz and Filler (1980) administered anaesthesia via a ventilating bronchoscope so that relief of tracheal compression could be assessed directly during the course of surgery. However, the advantages of this technique must be outweighed by the trauma to the trachea caused by prolonged instrumentation.

In a recent study, Wiseman, Duncan and Cameron (1985) described the management of tracheomalacia in three neonates using a tracheostomy and long term continuous positive airway pressure (CPAP). Treatment times ranged from 13 to 25 months, each child being eventually weaned from CPAP and decannulated. The authors claim that this method of management is a reasonable alternative to tracheopexy. However, such a prolonged stay in hospital—in addition to the hazards of tracheotomy—would appear to be a major disadvantage when compared with the rapid recovery and lack of postoperative complications in patients managed by tracheopexy.

In conclusion, the diagnosis of tracheomalacia must be considered as a cause of cyanotic and apnoeic attacks in any infant who has undergone surgical repair of oesophageal atresia. Severe cases can be managed successfully by performing tracheopexy. Intraoperative problems are usually few, provided meticulous attention is paid to maintaining patency of the airway and ventilation. The postoperative course is usually uneventful.

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REFERENCES