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Oesophageal Atresia—the Melbourne Experience

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INTRODUCTION

Since oesophageal atresia was first successfully treated at the Royal Children's Hospital in Melbourne\(^1,2,3\) by Russell Howard in 1949, it has always attracted considerable attention at our institution. By February 1994, we had treated a total of 656 patients with oesophageal atresia and/or tracheo-oesophageal fistula. This report describes our experience with these patients, identifies current problems, and suggests possible solutions to these problems.

PATIENTS AND METHODS

Data have been collected prospectively by Nate Myers, supplemented by a major review in 1988, and several clinical research projects supported by the Oesophageal Atresia Research Auxiliary.

RESULTS AND DISCUSSION

INCIDENCE

In Victoria, Australia, the incidence of oesophageal atresia has been estimated to be 1/4500 live births, a figure similar to that quoted by Haight in 1957\(^4\). In a more recent 10-year study, there were 175 indexed cases born in Victoria, including 9 stillbirths, representing a total birth incidence of 1/3448 and a live-birth incidence of 1/3570\(^5\).

In the Royal Children's Hospital series from 1948 to 1988 there were 23 twin pairs, and all were discordant for oesophageal atresia. In the 10-year sub-group, there were 11 twin pairs, a frequency of twinning of 7%, compared to the expected rate of 2.3% for our populations. Significantly, more indexed cases were born following the third and fourth pregnancy than expected. The possible confounding factor of a maternal age effect remains to be clarified.

DIAGNOSIS

It is uncommon for oesophageal atresia to be diagnosed on antenatal ultrasound. The recognition of congenital heart disease and other abnormalities antenatally may encourage the ultrasonographer to look carefully at the size of the stomach and swallowing patterns, but it is rare that oesophageal atresia can be diagnosed confidently, even by experienced ultrasonographers. We have only one patient in whom an antenatal diagnosis was made. The observation of polyhydramnios and certain coexistent abnormalities should encourage the ultrasonographer to examine the fetal esophagus and stomach carefully. It must be remembered, however, that polyhydramnios is also present in many other congenital conditions, and while it is seen in 95% of cases with atresia without fis-
tula, it is only evident in 35% of those with a distal tracheo-oesophageal fistula.

We recommend passage of a size 10 catheter in all babies in whom oesophageal atresia is suspected. Any baby drooling excessive saliva should be assumed to have oesophageal atresia, until successful passage of an oro-oesophageal tube. The varying presentations of oesophageal atresia with distal fistula are summarised in Table 1. A history of maternal polyhydramnios in pregnancy in an infant born slightly prematurely heightens the suspicion of oesophageal atresia. We have encouraged our pediatricians to suspect the diagnosis prior to attempted feeds, to avoid cyanosis, choking and aspiration, but despite this the diagnosis is sometimes made only following feeds (see Table).

Confirmation of Diagnosis

Any newborn infant observed to be drooling excessive saliva should be assumed to have oesophageal atresia until successful passage of an oro-oesophageal tube. However, the clinician should be aware that some premature infants may not appear to secrete much saliva.

A relatively stiff 10 gauge nasogastric tube, introduced through the mouth, becomes arrested 8-11 cm from the gums when oesophageal atresia is present. A smaller calibre tube is not used because it may curl up in the upper pouch and give a misleading impression of oesophageal continuity.

Determination of the Anatomical Type

Obstruction to the passage of an orogastric tube indicates atresia of the oesophagus. The present of a distal tracheo-oesophageal fistula can be shown on plain radiography: an x-ray of the chest and abdomen will reveal gas in the bowel below the diaphragm if a distal fistula is present.

The Gasless Abdomen

If there is no gas in the abdomen for more than a few minutes after birth, the patient has either oesophageal atresia without a fistula (44 infants), or atresia with a proximal fistula (12 infants). The rare distal, unopened fistula has been described, but has been encountered only once in our experience. In the infant with a gasless abdomen, the existence or otherwise of a proximal fistula can be determined by performing either an upper oesophageal contrast study, or bronchoscopy. If no upper pouch fistula is found, atresia without a fistula is assumed.

Misleading diagnosis

We have had two patients in whom a traumatic pseudo-diverticulum has occurred in a premature infant in whom intubation was difficult, giving a misleading impression of oesophageal atresia. Copious drooling of saliva is associated with apparent inability to pass a tube into the stomach. The pseudodiverticulum results from traumatic passage of a suction catheter or endotracheal tube through the mucosa of the posterior wall of the pharynx. If this is suspected, a contrast study of the oesophagus will show that the level of obstruction is lower in the chest than would be expected in oesophageal atresia, and there is little gas in the alimentary tract. A lateral chest x-ray after contrast may show residual barium in the pseudo-diverticulum. In this way, inappropriate thoracotomy can be avoided.

ASSOCIATED ABNORMALITIES

A little over half of infants with oesophageal atresia have significant associated congenital abnormalities (see Table 2), and about half of these have congenital heart

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### TABLE 1 PRESENTATION OF OESOPHAGEAL ATRESIA WITH DISTAL TRACHEO- OESOPHAGEAL FISTULA.

<table>
<thead>
<tr>
<th>Description</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excessively mucousy baby</td>
<td>209</td>
</tr>
<tr>
<td>Respiratory symptoms</td>
<td>79</td>
</tr>
<tr>
<td>Difficulty with attempted feeding</td>
<td>92</td>
</tr>
<tr>
<td>Passage of orogastric tube</td>
<td>92</td>
</tr>
<tr>
<td>*Suspicion because of other abnormalities</td>
<td>44</td>
</tr>
<tr>
<td>*During routine resuscitation</td>
<td>29</td>
</tr>
<tr>
<td>*Because of maternal polyhydramnios</td>
<td>19</td>
</tr>
</tbody>
</table>

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### TABLE 2 INCIDENCE OF ASSOCIATED ANOMALIES IN OESOPHAGEAL ATRESIA

<table>
<thead>
<tr>
<th>Type of Congenital Anomaly</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital heart disease</td>
<td>27</td>
</tr>
<tr>
<td>Urinary tract abnormalities</td>
<td>22</td>
</tr>
<tr>
<td>Orthopaedic (vertebral and limb)</td>
<td>15</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>22</td>
</tr>
<tr>
<td>Chromosomal</td>
<td>5</td>
</tr>
<tr>
<td>TOTAL% OF PATIENTS WITH CONGENITAL ABNORMALITIES</td>
<td>56</td>
</tr>
</tbody>
</table>
disease. In infants with congenital heart disease, early complete repair of the oesophageal atresia with division of the fistula is indicated, except where the cardiac lesion is duct-dependent. In order to recognise the duct-dependent lesions pre-operatively, it is our practice to perform routine echo-cardiography in all infants with oesophageal atresia prior to surgery. If there is a duct-dependent lesion, a prostaglandin E1 infusion is commenced and surgery delayed until the infant is stable.

In addition, recognition that about 3% of infants with oesophageal atresia have a renal abnormality incompatible with long-term survival, has meant that if the infant has not been observed to pass urine, a renal ultrasound is mandatory pre-operatively. If no kidneys can be identified on ultrasound, surgery is delayed until a nec- lear scan of the kidneys has been obtained. It must be remembered that in oesophageal atresia and bilateral renal agenesis, the typical Potter facies is not present.

The availability of geneticists has enabled the diagnosis of major chromosomal abnormalities (the commonest of which are Trisomy 18 and 21) and other abnormalities, eg CHARGE association, to be diagnosed early. Some of these may influence treatment.

Reporting of associated abnormalities

There has been considerable variation in the methods by which associated anomalies have been reported, which makes it difficult to compare experiences between centres. The apparent frequency of associated abnormalities is dependent on the extent to which coexistent lesions have been sought by investigation. We advocate a simple approach whereby:

1. All data on associated anomalies should be collected prospectively;
2. Routine investigations should include renal ultrasound and echocardiography as a minimum requirement;
3. The number of associated abnormalities should be reported as a percentage of the total cohort of patients, rather than as a percentage of the anomalies themselves;
4. True congenital abnormalities should be recorded separately from acquired conditions; and
5. The wide variation in the impact of associated abnormalities on the management and long-term out-

look in oesophageal atresia should be recognised.

SEVERELY PREMATURE INFANTS WITH HYALINE MEMBRANE DISEASE

The severely premature infant is likely to develop hyaline membrane disease. This is a potentially serious problem in oesophageal atresia, as deterioration and gaseous exchange necessitates increased ventilatory support which may prove ineffective in the presence of a distal tracheo-oesophageal fistula. In the past, this has resulted in gastric dilatation and perforation, pneumoperitoneum, pneumomediastinum, shock and death. Gastrostomy alone is not the solution because it simply encourages air to pass preferentially through the distal fistula. A recent review of our experience makes it clear that the optimal treatment is early surgical division of the tracheo-oesophageal fistula. The more severe the hyaline membrane disease, the more urgently is thoracotomy required. If the infant's condition improves once the fistula is controlled, the oesophagus can often be repaired at the time of thoracotomy. Otherwise, the oesophagus can be joined at a later date.

OPERATIVE PROCEDURE

The infant is placed in the full lateral position with the right side uppermost and a towel folded underneath the body to give lateral flexion. The right arm is raised over the head to facilitate the thoracic approach. A transverse incision is made just below and centred on the angle of the scapula, and after division of the fibres of the latissimus dorsi in the line of the incision, the posterior fibres of the serratus anterior are divided near their origin, as low as possible in the incision, preserving their innervation. The chest is entered through the fourth intercostal space and the pleura swept off the chest wall. The oesophagus is approached in the extra-pleural plane where the aygous vein is ligated and divided. After incision of the fine endothoracic fascia of the posterior mediastinum, the lower oesophagus can be found immediately anterior to the aorta, and is highlighted by vagal fibres running along its surface.

The communication of the oesophagus to the trachea is exposed. A vascular sling may be passed around the fistula where the upper part of the lower oesophageal segment joins the trachea, once the angle between the oesophagus and trachea has been dissected clear. Care is
taken to avoid damage to vagal fibres and the blood supply of the oesophagus. The tracheo-oesophageal fistula is closed with 4/0 or 5/0 polyglycolic acid transfixion sutures, and divided.

The fundus of the upper oesophageal segment can be readily identified within the chest when the anaesthetist passes a catheter into it. A stay suture passed through its lowest part assists its mobilisation and avoids unnecessary handling of (and trauma to) the oesophagus. The upper oesophagus can be mobilised as far as the cri-copharyngeus muscle without devascularising it. Once the upper segment has been mobilised, its most dependent part is opened. In many instances, the gap between the oesophageal ends is such that little further mobilisation is required.

The end-to-end oesophageal anastomosis is constructed by inserting three interrupted 5/0 polyglycolic acid sutures in the posteromedial (furthest away) aspect of the oesophagus, taking in all-layers, with moderately large bites of tissue. It is important that the mucosal layer of both the upper and lower segments is included, because it tends to retract out of view and is easily missed. When the three sutures have been placed, the oesophageal ends are gently apposed and the sutures tied on the mucosal surface. An orosso gastric tube can now be passed through the upper oesophagus into the lower segment, before completion of the anastomosis with a further 4-6 all-layer interrupted sutures, the knots of which may be tied on the outside.

Before closure, the orosso gastric tube is removed, unless gavage feeding is planned, as in the premature infant. The thoracic cavity is irrigated with warm antibiotic saline solution. This fluid allows confirmation that there is no air leakage from the closed fistula with ventilation. A chest-drain is not normally required, unless there is concern about the integrity of the anastomosis.

LONG GAP OESOPHAGEAL ATRESIA

The long gap in oesophageal atresia still represents one of the greatest challenges to the paediatric thoracic surgeon. Numerous methods have been advocated to overcome this problem, including staged procedures, daily bouginage, circular and spiral myotomies, and oesophageal replacement by stomach, jejunum or colon. It has been our experience that the best long-term results are achieved if the patient's oesophagus is preserved, and it is unusual that oesophageal replacement is required. Even in oesophageal atresia without fistula (the anatomic variant which is associated with the longest gap between the oesophageal segments) the oesophagus can usually be anastomosed successfully, albeit at 2-3 months of age. In our last 18 patients with oesophageal atresia without fistula, successful oesophageal anastomosis has been achieved in patients.

Our policy has been to extensively mobilise the upper pouch as far as the cricopharyngeus, and then the lower pouch as far as the oesophageal hiatus, if required. An end-to-end all-layer oesophageal anastomosis is then constructed, ensuring that the mucosa is included. Some tension on the anastomosis is allowable. Good bites of tissue and meticulous attention to technique with accurate placement of the sutures, leads to a low complication rate. We have found circular myotomy only occasionally helpful. The gap is often greatest when the upper pouch is short, making it difficult to perform a circular myotomy of the upper pouch an adequate distance from the anastomosis. Experimental work in neonatal pigs, which we have performed in association with Watanabe, suggests that a circular lower-segment myotomy may be just as satisfactory.

We no longer use colon for oesophageal replacement. Our preference has been for gastric greater curvature tube replacement. In recent years we have been encouraged to consider the techniques described by Lewis and Searle.

COMPLICATIONS OF REPAIR OF OESOPHAGEAL ATRESIA

A number of problems may occur following repair of oesophageal atresia. Some, such as anastomotic leak, recurrent tracheo-oesophageal fistula and a shelf at the site of anastomosis, are the result of technical inadequacies, whereas others, such as poor oesophageal clearance and gastro-oesophageal reflux causing the late development of an oesophageal stricture, reflect abnormalities more directly related to the oesophageal atresia itself.

Anastomotic Leak

Leakage from an oesophageal anastomosis represents a serious complication of repair of oesophageal atresia. The
likelihood of an anastomotic leak occurring depends on the type of anastomosis employed, and the extent of mobilisation of the oesophagus. An interrupted, all-layers, endo-to-end oesophageal anastomosis using an absorbable suture, appears to have the lowest leakage and stricture rate, making it the anastomosis of choice. Factors which contribute to anastomotic leakage include: (1) incorrectly placed sutures; (2) insecure sutures; (3) excessive tension at the anastomosis; (4) ischaemia of the oesophageal ends; and (5) sepsis. There is a relationship between leakage and subsequent stricture-formation, and recurrent tracheo-oesophageal fistula.

Leakage from an anastomosis may vary enormously in significance and presentation, from a minor radiological leak in an otherwise well infant (for which no treatment is required) to complete anastomotic disruption with mediastinitis, empyema, pneumothorax and septicaemia. The diagnosis is confirmed radiologically, using a contrast study of the oesophagus.

In most infants an anastomotic leak can be managed non-operatively. Safe total parenteral nutrition enables oral feeds to be ceased. Antibiotics are commenced, and the leak will usually close spontaneously. Cervical oesophagostomy is only rarely necessary, when supportive therapy has been unsuccessful and there is ongoing difficult-to-control sepsis. A long-standing leak may require gastrostomy to allow continuation of enteral feeds.

**Recurrent tracheo-oesophageal fistula**

A recurrent tracheo-oesophageal fistula is a severe and potentially dangerous complication. Failure to close the fistula adequately at the time of surgery, and subsequent anastomotic leak with local infection, increase the chance of developing a recurrent fistula. The use of silk appears to be another predisposing factor.

The development of coughing, gagging, choking, apnoea, dying spells and recurrent chest infections, suggest a recurrent tracheo-oesophageal fistula has developed. The typical presentation is that of an infant who coughs and splutters with each feed. The most reliable method of confirming the diagnosis is cineradiographic tube oesophagography with the patient in the prone position. Barium is introduced through a nasogastric tube positioned in the oesophagus as the tube is gradually withdrawn. Bronchoscopy is an alternative method.

Spontaneous closure of recurrent fistulae is unlikely. Most centres wait at least four weeks from the first operation before closing a recurrent fistula. A thoracotomy is performed through the original incision when the child is in optimal respiratory and general condition, following a period of intravenous nutrition. The fistula is divided via a transpleural approach. Some surgeons have found the passage of a fine ureteric catheter through the fistula facilitates its localisation, and some surgeons place mediastinal tissue between the ends of the divided fistula in an attempt to prevent a further recurrence.

**Anastomotic Stricture**

Anastomotic stricture is the most common reason further surgery to the oesophagus is required after repair of oesophageal atresia. Factors which influence the development of an oesophageal stricture include: (1) rough handling of the oesophagus at the time of repair; (2) ischaemia of the oesophageal ends; (3) excessive tension of the oesophageal anastomosis; (4) the choice of suture material [eg silk]; (5) anastomotic leak or dehiscence; (6) the use of a two-layer anastomosis; and (7)

**TABLE 3** SEVERITY OF LEAK AFTER PRIMARY ANASTOMOSIS IN 200 CONSECUTIVE PATIENTS WITH OESOPHAGEAL ATRESIA AND DISTAL FISTULA.

<table>
<thead>
<tr>
<th>PRESENTATION OF LEAKAGE</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>No leak</td>
<td>79.0</td>
</tr>
<tr>
<td>Asymptomatic radiological leak</td>
<td>8.5</td>
</tr>
<tr>
<td>Saliva in chest drain</td>
<td>9.5</td>
</tr>
<tr>
<td>Empyema/mediastinitis</td>
<td>2.0</td>
</tr>
<tr>
<td>Lung abscess</td>
<td>1.0</td>
</tr>
</tbody>
</table>

**TABLE 4** RELATIONSHIP OF COMPLICATIONS TO TYPE OF ANASTOMOSIS

<table>
<thead>
<tr>
<th>COMPLICATION</th>
<th>END-TO-END ANASTOMOSIS</th>
<th>SIDE-TO-END ANASTOMOSIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Major leak</td>
<td>4%</td>
<td>4%</td>
</tr>
<tr>
<td>All leak</td>
<td>12%</td>
<td>25%</td>
</tr>
<tr>
<td>Recurrent fistula</td>
<td>4%</td>
<td>11%</td>
</tr>
<tr>
<td>Stricture requiring resection</td>
<td>2%</td>
<td>13%</td>
</tr>
</tbody>
</table>
gastro-oesophageal reflux. Gastro-oesophageal reflux is the most common cause of late stricture development.

Patients with a stricture develop feeding difficulties and dysphagia. Their onset may be insidious. They may present as "slow feeders" or have excessive regurgitation, with or without cyanotic episodes. Older children present with foreign body impaction of food in the oesophagus. Diagnosis is confirmed by barium swallow or endoscopy.

In patients with mild narrowing of the oesophagus, one or two dilatations may be all that is required. However, in patients with associated gastro-oesophageal reflux, it will usually be necessary to perform an antireflux operation (e.g. Nissen fundoplication), after which the stricture will resolve.

Motility problems

Oesophageal motility is abnormal, both before and after repair of oesophageal atresia. It is likely that vagal fibres are injured during mobilisation of the oesophagus, worsening the already abnormal oesophageal motility. Oesophageal motility tends to improve gradually with age, but children with oesophageal atresia often need to drink with their meals. Abnormal oesophageal motility may contribute to oesophagitis and oesophageal stricture formation in the presence of gastro-oesophageal reflux; poor oesophagus-emptying allows acidic gastric juice to sit in the lower oesophagus for a longer period of time than in patients with normal muscular action.

Oesophageal diverticulum and shelf

A pseudo-diverticulum may occur following leakage from the oesophageal anastomosis. Ballooning at the site of a circular myotomy is common, and may result in a diverticulum. A shelf at the site of the oesophageal anastomosis occurs when the upper oesophageal pouch has been opened eccentrically, or the end-to-end oesophageal anastomosis has not been performed with sufficient precision.

GASTRO-oesophageal REFLUX

Gastro-oesophageal reflux is a major cause of ongoing problems in oesophageal atresia patients, and in some series has been reported in up to 50%. It is a potent cause of oesophageal stricture in oesophageal atresia patients, particularly as oesophageal motility is often abnormal, making the oesophagus more susceptible to stretcher formation. The most effective treatment of oesophageal stricture is fundoplication. Even with oesophageal dilatation, once reflux has been corrected, severe strictures often resolve spontaneously.

LONG-TERM MORBIDITY

Chest-Wall Deformity

Five years ago we reviewed the appearance of the chest wall following thoracotomy and repair of oesophageal atresia. This study revealed that the majority of adults who had neonatal repair of their oesophageal atresia through an intercostal approach, and who had only one thoracotomy, had little or no chest wall deformity. Factors which appeared to increase the likelihood and severity of chest wall deformity included: (i) rib resections; (ii) multiple thoracotomies; and (iii) intra-thoracic complications, e.g. mediastinitis or empyema. The factor which had the greatest bearing on subsequent chest wall deformity and/or scoliosis was the presence of a congenital vertebral abnormality, part of the VATER association.

It would appear that the use of the intercostal approach, avoidance of deliberate staged procedures, and reduction in the incidence of anastomotic dehiscence and its complications, can all be expected to reduce the incidence and severity of later chest wall deformity.

Quality of Adult’s Life

We have performed a comprehensive study of our adult survivors of oesophageal atresia, supported by the Oesophageal Atresia Research Auxiliary. This Auxiliary is a group formed by parents of oesophageal atresia patients, dedicated to supporting research in oesophageal atresia. More than 83% of survivors were reviewed, and it appears that most have a normal life-style. Many have minor dysphagic symptoms, may require a drink with their meals and are prone to respiratory illness, but they differ little from the normal population. Given the recent improvements in surgical and neonatal care, and the lower incidence of complications, I believe we can be very confident in reassuring parents of newborn infants with oesophageal atresia, that the outlook for their child is good in the long term.

MORTALITY

Consistent with the experience in all other major centres, at the Royal Children’s Hospital there has been a
steady decline in the overall mortality of oesophageal atresia with each passing decade\textsuperscript{25}. For more than a decade, mortality in oesophageal atresia has been essentially that of the major associated anomalies. No longer do infants die from complications of the oesophageal atresia itself, or of its treatment. Recognition of specific syndromes and associations, and pre-operative investigation, have enabled the surgeon to recognise major concomitant lesions which do not justify ongoing treatment. Prior to 1972, 6.5\% had surgery withheld, compared with 13.5\% since 1972. Another factor which appears to contribute to this increased non-operative rate in recent years is that infants with multiple anomalies are more often reaching our tertiary institutions\textsuperscript{26}.

\textbf{Causes of Death}

In the early years, most deaths were the result of respiratory failure and inadequate resuscitation, from soiling of the lungs, hyaline membrane disease, and other complications of prematurity. The other major cause of mortality was from complications of the oesophageal surgery itself, particularly those related to dehiscence of the anastomosis and poor nutrition. As neonatology and operative techniques have improved, the main cause of death has become that caused by associated anomalies, and in particular, major chromosomal aberrations.

\textbf{The Waterston Classification}

In 1962, Waterston \textit{et al} recognised the importance of birth weight (and, indirectly, gestation) and soiling of the lungs as factors which influence survival. The simple classification offerd by Waterston\textsuperscript{27} gained widespread acceptance. The dramatic improvement in survival for oesophageal atresia in each of the Waterston groups is such that mortality in Groups A and B is now extremely uncommon. The continued high incidence of mortality in Group C reflects the presence of major associated anomalies, rather than complications of oesophageal surgery or prematurity. Since 1970, prematurity has had little effect on mortality. Our experience would suggest that it is no longer relevant to use the Waterston classification, and that in the future, mortality will be best predicted by the type and severity of concomitant congenital abnormalities alone.

\textbf{REFERENCES}


12) Myers NA, Beasley SW, Auldist AW, Kent M.