CHAPTER 48
Oesophageal Atresia

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Introduction

Little from Africa has been reported or written on the subject of oesophageal atresia (OA). The frequency of diagnosis and especially survival is widely variable, depending on available resources and expertise. In some areas, the incidence of diagnosis and survival is unlikely, whereas in a developing country such as South Africa, where facilities are good and a limited number of dedicated paediatric surgeons accumulate a large amount of experience, results are comparable to those of the developed world.

Demographics

The incidence of oesophageal atresia in Africa is unknown but would appear to be no different from that in other populations. Amongst South Africa's multicultural population, the incidence in the white population seems to be higher; however, this was very likely a spurious impression due to missed diagnoses.

In 2008, Nandi and colleagues reported equivalent incidences of oesophageal atresia/tracheo-oesophageal fistula (OA/TOF; 2.1% of all neonatal admissions) at two linked surgical departments in Europe and Africa: John Radcliffe, Oxford, United Kingdom; and Kilimanjaro Christian Medical Centre, Tanzania. Reports from Nigeria and Zimbabwe show an incidence comparable to that at Great Ormond Street Children's Hospital in London. The incidence in Africa probably corresponds to the 1 per 3,000–4,500 reported across the world in the literature.

Aetiology/Pathophysiology

The mechanisms of embryological development of OA/TOF occurs in the embryo at 3 weeks postfertilisation during the demarcation of the proximal foregut as the oesophagus with a gastric bubble caudally and a ventral lung bud cranially. During the subsequent phase of elongation of the oesophagus and lung bud, there is a further division of the tracheal primordium from the oesophagus.

The mechanism of development of tracheo-oesophageal fistula is the failure of apposition of longitudinal ridges, whereas the mechanism of the development of oesophageal atresia is apposition too posteriorly.

OA/TOF is classified into six types (see Figure 48.1):

A. Isolated oesophageal atresia (8%)

B. Upper pouch fistula with oesophageal atresia (1%)

C. Oesophageal atresia with tracheo-oesophageal fistula (86%)

D. Upper and lower pouch fistula (0.5%)

E. H-type fistula (4%)

F. Oesophagus with tracheal segment (0.5%)

Clinical Presentation

Prenatally, the condition may be suspected from maternal polyhydramnios and absence of a fetal stomach bubble at the 20-week anomaly scan. Prenatal scan diagnosis of OA/TOF is estimated to be less than 42% sensitive with a positive predicted value of 56%. Additional diagnostic clues are provided by associated anomalies, such as trisomy (13, 18, 21); VACTERAL (vertebral, anorectal, cardiac, tracheo-oesophageal, renal, limbs) sequence; and CHARGE (coloboma, heart defects, atresia choanae, retarded development, genital hypoplasia, ear abnormality) association. These associated anomalies are present in more than 50% of cases and worsen the prognosis; thus, prenatal karyotyping is essential. Duodenal atresia may coexist with OA/TOF. The risk of recurrence in subsequent pregnancies for isolated OA/TOF is less than 1%. Delivery is advised to be at a specialised centre with neonatal surgical input.

In the absence of prenatal diagnosis, the presentation may be respiratory distress, cyanotic spells, frothing around the mouth, and arrested passage of a nasogastric tube. Recurrent pneumonia or failure to feed are noted in delayed presentations. Presentation with gastric rupture, especially in low birth weight babies, is known to increase morbidity and mortality.

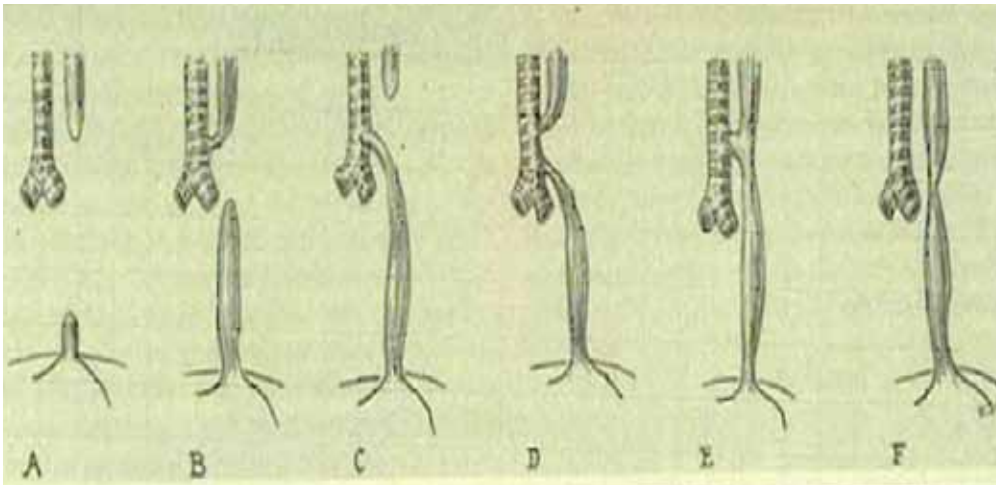


Figure 48.1: Types of oesophageal atresia and tracheo-oesophageal fistula.

Investigations

A babygram (Figure 48.2) with a radio-opaque nasogastric tube is the most informative imaging tool. This image helps with diagnosis, confirms OA+TOF or isolated OA, diagnoses the associated anomalies of VACTERL, and identifies associated duodenal atresia. Renal ultrasound is helpful in confirming renal anomalies, and a cardiac assessment may confirm the 30% associated cardiac anomalies. If dysmorhphic features are suspected, karyotyping may confirm chromosomal anomalies. However, if life-threatening chromosomal anomalies are suspected, the treatment may be delayed until karyotype results are obtained, which could be within 48 hours. Contrast swallows are discouraged due to the risk of aspiration (Figure 48.3).

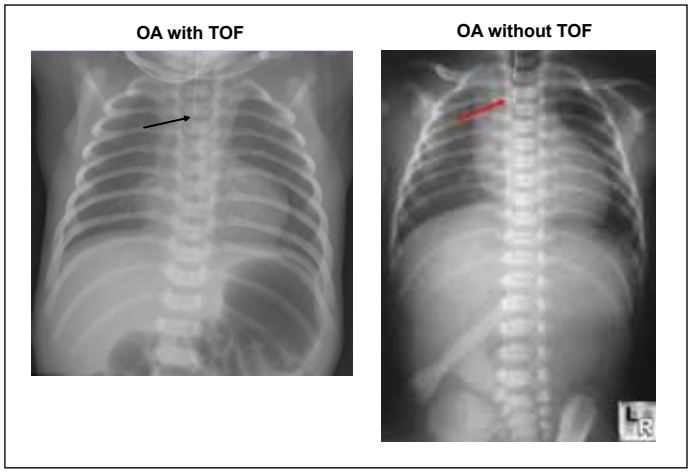


Figure 48.2: A babygram showing OA+TOF on the left and isolated OA on the right.

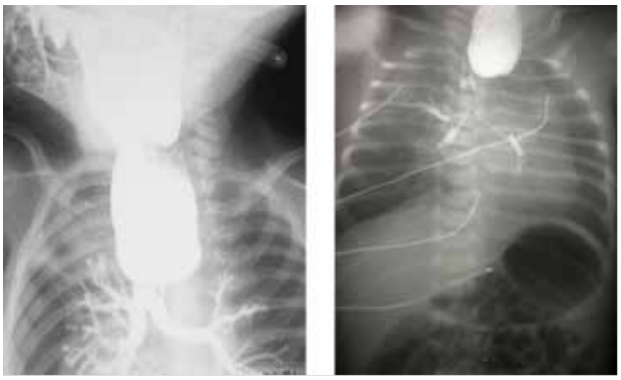


Figure 48.3: Examples of excessive volumes of contrast given to infants with oesophageal atresia, resulting in aspiration.

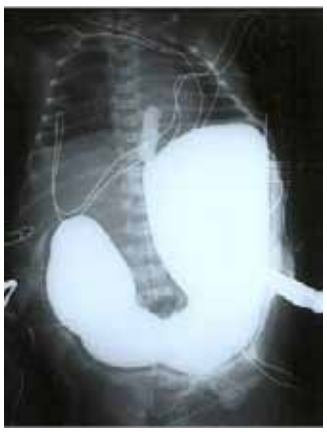


Figure 48.4: An on-table gastrogram in a baby with pure oesophageal atresia and duodenal atresia. A duodenoduodenostomy was performed, followed by oesophageal atresia repair after a 3-day interval.

Cardiac echoes have proven unreliable in terms of identifying right aortic arch; however, this identification hasn't been particularly helpful because a right aortic arch has not been an impediment to repair via a right posterolateral extrapleural access. Routine preoperative bronchoscopy is advocated in some centres, but is not essential, and thus is not practiced by many centres.

In the event of a gasless abdomen and pure atresia, an initial gastrostomy may be required to establish the length of gap between proximal and distal segments of the oesophagus (Figure 48.4).

Management

Surgical intervention is urgent only in the event of abdominal distention causing ventilatory distress, gastric distention, or rupture. Otherwise, oesophageal atresia constitutes an urgent elective case to be repaired within 24 hours, preferably in the light of day, and after acquiring a cardiac echo and convening a suitable team of anaesthesiologist and operating room staff.

The patient is positioned in the left lateral position with a small strut under the left chest wall. Nasotracheal intubation is encouraged to ensure a stable endotracheal tube above the level of the carina. Surgery necessitates retraction of the right lung, so it is essential that the left lung is ventilated.

Depending on the size of the patient, a 16, 18, or 20 Wishard catheter is introduced into the proximal oesophageal pouch, replacing the Replogle tube, to be advanced by the anaesthetist when required.

An approximate 3-cm incision is made just below the angle of the right scapula, strictly in the skin lines to leave an optimal, almost imperceptible scar. Non–muscle-cutting access to the rib cage is established through the angle of auscultation between the latissimus and trapezius muscles.

The fourth intercostal space is opened on the superior margin of the fifth rib, maintaining an extrapleural plane. This is possible in most cases—a small breach of the pleura can be tied on a mosquito forcep at completion.

The azygos vein is tied and divided and the distal tracheo-oesophageal fistula is identified, isolated, and serially divided with a 1-mm cuff against the trachea, which is approximated with interrupted 6-0 proline or polydiaxonone sutures.

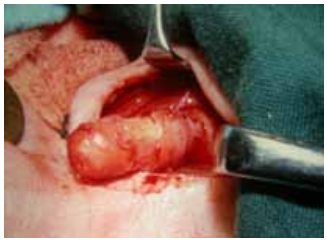
A feeding tube is passed down the distal oesophagus to ensure distal patency and to empty the stomach. The anaesthetist is then asked to advance the Wishard catheter, and the upper pouch is mobilised, if necessary, into the thoracic inlet and neck. This technique greatly facilitates mobilisation and separation in the plane of close adherence to the trachea.

The lower oesophagus is mobilised sufficiently to approximate the two segments. Where limited gap allows, the fistulous proximal end of the distal oesophagus is resected to achieve anastomosis to a better calibre, and to have mild distraction of the two ends.

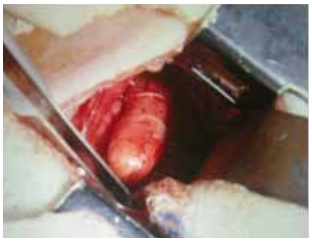
A single transverse incision is made across the end of the upper pouch onto the Wishard catheter, and lateral and medial angle sutures are placed across the segments. The Wishard catheter is withdrawn, and the posterior anastomotic sutures are completed. The angle and posterior layer sutures are then tied. If necessary, the chest wall strut is removed to relieve tension. The anaesthetist is then asked to advance a Replogle or feeding tube, which is guided through the anastomosis. The anterior layer is completed.

The extent of proximal mobilisation overcomes most gaps. In the event of a 2.5cm+ vertical gap, options include the use of several techniques, including circular myotomies (Figure 48.5). If this is required, it is usually in the circumstance of a very high upper pouch, in which case the upper pouch needs to be mobilised out of the neck, myotomised, and placed back in the thoracic inlet. This is done with the Wishard catheter in the lumen prior to opening the apex of the pouch. Other options include an oesophageal flap (Figure 48.6).

Some recent controversy has arisen as to whether to drain the extrapleural para-anastomotic space when one is very confident of the



Circular Myotomy through the neck



Intrathoracic Circular Myotomy through the thoracotomy wound

Figure 48.5: Circular myotomy.

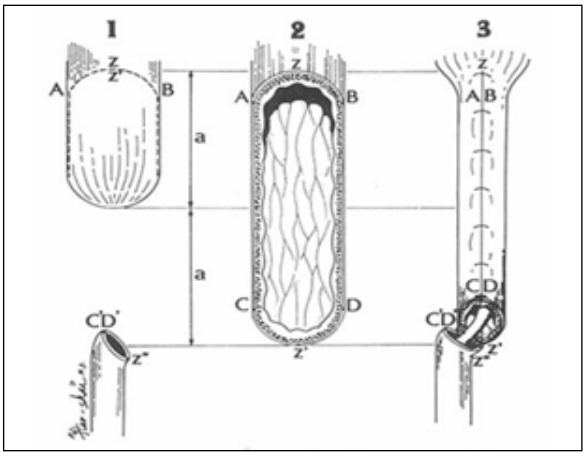


Figure 48.6: Flap technique: (1) the closed upper pouch; (2) the upper pouch is open and its lateral wall reaches the lower segment of the oesophagus; (3) the elongated upper pouch is anastomosed to the lower segment.

integrity of the anastomosis. A drain does no harm and can be removed in 2 or 3 days if one has confidence in the anastomosis. In cases anastomosed under tension, a para-anastomotic drain is retained for a week, and feeds are delayed until a contrast swallow confirms an intact oesophagus.

Gastrostomies are essentially used only for pure oesophagael atresia cases. The rest are fed via a transanastamotic tube initially, and subsequently are fed orally.

Minimally invasive endoscopic surgery has become the vogue in paediatric surgery, and has excellent application for a considerable range of procedures. Whether this is true for thoracoscopic repair of OA since the first report in 1999 is debatable. The foremost and most enthusiastic endoscopic surgeons promote this procedure and claim that their results and complication rates are at least comparable with open operative procedures. The technique may be used if equipment and expertise are available.

Postoperative Complications

An anastomotic leak may have dire consequences if a major intrapleural disruption occurs. Anastomotic leaks should be uncommon and, if extrapleural and contained, can be treated expectantly by maintaining parenteral nutrition and drainage until the leak seals and heals. A major intrapleural disruption can be life threatening and warrants early re-exploration. If total disruption occurs, the oesophagus may have to be abandoned and the upper pouch exteriorised as a cervical oesophagostomy.

A recurrent tracheo-oesophageal fistula requires operative intervention, closure, and tissue interposition, although endoscopic injection of glue has been described.

Another complication is stridor and cyanotic episodes related to tracheomalacia and a collapsing upper airway. The combination of gastro-oesophageal reflux and tracheomalacia may be particularly sinister and life threatening and require early intervention. Generally, gastro-oesophageal reflux is addressed first. Occasionally, infants cannot be fed at all without immediately refluxing and aspirating, and may be at risk of sudden infant death syndrome (SIDS).

Other postoperative complications include anastomotic stricture; gastro-oesophageal reflux; long-term Barrett's oesophagus due to chronic reflux; and chylothorax.

A well-recognised but uncommon pitfall in the operative management of OA, tracheo-oesophageal fistula occurs in the event of a carinal fistula with tracheal communication into the fork of the carina. In this situation, the relative position of the right main bronchus and distal oesophagus just below the fistula is reversed. The surgeon should always confirm that a structure isolated as the distal oesophagus is correctly identified by atraumatic occlusion and confirmation that the right lung still ventilates before dividing the structure.

Conclusion

In the Western world, current expectations of survival are that all patients with oesophageal atresia will survive unless there are major congenital malformations affecting other systems. There is no place for technical risk or error to compromise survival. In Africa, however, limiting factors are delayed diagnosis and restricted access to a neonatal intensive care unit (NICU). Shortages of medical and nursing personnel demand techniques be selected that limit the NICU requirement, in-hospital stay, and complications. The potential to rescue patients with surgical complications is probably not as good as it is in First World centres.

Evidence-Based Research

Table 48.1 presents a review of a 10-year personal experience with OA and TOF.

Table 48.1: Evidence-based research.

Title	Oesophageal atresia and tracheo-oesophageal fistula: review of a 10-year personal experience.
Authors	Adebo OA
Institution	Department of Surgery, University College Hospital and College of Medicine, University of Ibadan
Reference	West Afr J Med 1990; 9(3):164–169
Problem	Outcomes of TOF and OA repair in the African setting is poor.
Intervention	This study seeks predictors to improve outcome.
Comparison/ control (quality of evidence)	Eleven neonates with oesophageal atresia and distal fistula were managed between July 1977 and January 1987. The male-to-female ratio was 1.2:1. The patients were aged between 1 to 14 days (median of 7 days) and weighed 1.85 to 3.10 kg (mean of 2.6 kg) at presentation. Associated anomalies were present in 5, pneumonia in 4, and uraemia (mean serum urea of 88 mg%) in all patients. A primary repair and simultaneous gastrostomy (omitted in one) were performed for all cases. There were 5 operative deaths. Fifteen postoperative complications occurred in 10 patients, including septicaemia in 3, wound infection in 3, anastomotic leak in 1, and tracheal mucous plug in 1. Statistical analysis indicated no difference between survivors and nonsurvivors on the basis of age, weight, degree of uraemia, or presence of pneumonia. One of the 6 survivors (now 5 years after surgery) required bouginage after 26 months and has remained asymptomatic; the other 5 are well and without symptoms 3 to 11 months postoperative.
Outcome/ effect	High mortality and morbidity for this neonatal condition.
Historical significance/ comments	The most significant determinants of survival are the effectiveness of pre- and postoperative managements of patients.

Key Summary Points

1. The incidence of oesophageal atresia in Africa is comparable to that in the Western world.
2. Prenatal diagnosis of OA is rare.
3. Delayed presentation of OA with pneumonia, dehydration, and failure to feed is the norm in Africa.
4. Diagnosis of PA is confirmed with a nasogastric tube and high index of suspicion.
5. A babygram is the most useful image.
6. Outcomes depend on the availability of resources and technical skill.

Suggested Reading

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