Oesophageal atresia and tracheo-oesophageal fistula

Spencer W Beasley

Abstract

Oesophageal atresia and tracheo-oesophageal fistula is a congenital structural abnormality that affects 1:4500 live infants. It is due to failure of the primitive foregut tube to separate correctly into the oesophagus and trachea. About 50% have associated abnormalities, of which the VACTERL (Vertebral, Anorectal, Cardiac, Tracheo-oEsophageal, Renal and Limb) association is the most common. Prematurity is common and all have some degree of tracheomalacia. Surgery of the common type can be performed through a fourth interspace thoracotomy or by thoracoscopy. It involves division of the distal tracheo-oesophageal fistula and anastomosing together the two ends of the oesophagus. The absence of a distal fistula reveals itself as a 'gasless abdomen' on plain radiology, and usually indicates a long gap between the blind oesophageal ends: this sometimes necessitates an oesophageal replacement if extensive oesophageal mobilization fails to achieve an end-to-end anastomosis of the oesophagus. Potential postoperative problems include anastomotic leak, anastomotic stricture, recurrence of the fistula, gastro-oesophageal reflux, oesophageal dysmotility, and food impaction. Survival is determined mainly by coexisting congenital abnormalities. The long-term risk of oesophageal malignancy is yet to be established. Isolated tracheo-oesophageal fistula ('H fistula') can occur without atresia, and often presents after feeding has commenced. It is divided through a cervical incision.

Keywords Anastomosis; gastro-oesophageal reflux; oesophageal atresia; oesophageal replacement; prematurity; tracheomalacia; tracheo-oesophageal fistula; VACTERL association

Background

Oesophageal atresia (OA) is a congenital structural abnormality of the foregut that results in a variable length of the mid-portion of the oesophagus to be missing. Typically, there is an abnormal attachment of the lower oesophageal segment to the trachea, called a distal tracheo-oesophageal fistula (TOF). Much less commonly there is a proximal TOF, which is an abnormal connection between the upper oesophageal segment and the trachea. Even more rarely, both proximal distal fistulae are present.

OA with or without TOF, should be diagnosed before or at birth, prior to the first feed being given. In OA, food entering the blind upper oesophageal segment has nowhere to go — except back up, or into the trachea from where it then spills into the lungs.

This article provides an overview of the diagnosis and management of OA and its implications beyond the neonatal period.

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Pathoembryology

Early in gestation, the developing embryo has a single foregut tube. Lung buds sprout from its ventral aspect. Shortly afterwards, and proximal to this, the tube divides into two parallel tubes: the anterior tube becomes the trachea, and the posterior one becomes the oesophagus. This process is orchestrated by the notochord which influences the expression of the Sonic Hedgehog gene, a process which is under strict temporo-spatial control. Any abnormality in the timing or location of this expression leads to abnormalities of tracheo-oesophageal division, as well as other abnormalities seen as part of the VACTERL (Vertebral, Anorectal, Cardiac, Tracheo-oEsophageal, Renal and Limb) association.

Classification

The variety of different anatomical configurations of OA and TOF has led to a number of classification systems, but the most widely used is shown in Figure 1. By far the most common type (about 84%) is OA with a distal TOF, termed a type C in the Gross classification. The most difficult to treat are types A and B, where there is no distal fistula and the gap between the blind oesophageal ends tends to be substantial, so-called 'long gap oesophageal atresia'. Due to the wide range of variations in OA, many surgeons prefer to avoid any confusion by being specific in their description of each case rather than adhering to one of the classifications e.g. pure OA with a long gap etc.

While not strictly OA at all, an isolated tracheo-oesophageal fistula ('H-fistula') is usually included in discussion of OA (see Management of Specific Situations below).

Incidence

Although there are some geographical variations in incidence of OA, the overall incidence is believed to be about 1:4500 live births. It occurs in all regions and all ethnic groups.

Antenatal diagnosis

Sometimes OA can be diagnosed on antenatal ultrasonography. Clues to its existence include a dilated upper oesophagus, small or absent stomach, abnormal swallowing, maternal polyhydramnios, and recognition of other abnormalities that are known to coexist with OA, such as those of the VACTERL association. These features raise the suspicion of OA, such that as soon as the baby is born, the definitive diagnosis still needs to be confirmed.

Clinical presentation

The classical feature at birth is of an excessively mucousy drooling infant that results from excessive saliva accumulating in the blind upper oesophageal pouch. Any infant with these features should not be fed until the diagnosis of OA has been excluded. Other clinical signs include tachypnoea, coughing and choking.

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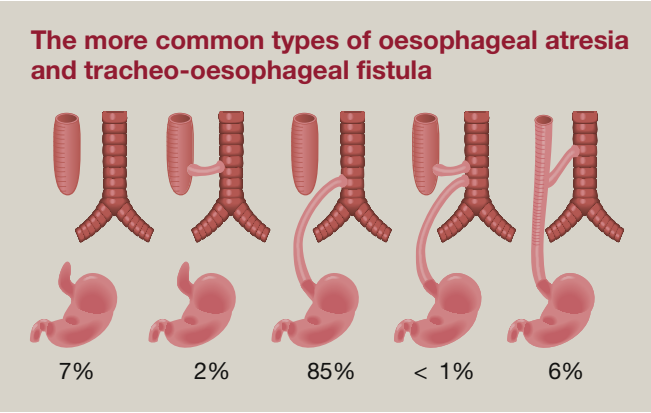

Figure 1

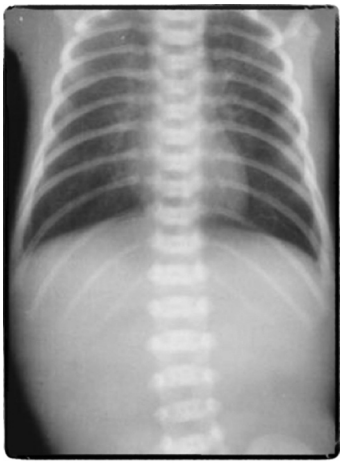

Figure 2 The gasless abdomen signifies absence of a distal tracheo-oesophageal fistula.

About one-third of infants with OA are born prematurely and about 30% have a history of maternal polyhydramnios: this percentage is much higher in those without a distal fistula. Where there are other abnormalities that have a known association with OA, the possibility of OA must be considered. For example, if there has been polyhydramnios, or there is an anorectal malformation, the patency of the oesophagus must be confirmed by passing a tube through it.

Investigations

There are three stages in the investigation of every infant with suspected OA.

1. *Confirm atresia of the oesophagus*. This is done by passing an 10 FG orogastric tube through the mouth into the oesophagus. In OA such a tube cannot be introduced further than 9 −10 cm from the gums. This is the only test required to make the diagnosis. A finer tube is not used because it can curl around in the upper oesophageal pouch giving a false impression of oesophageal continuity.
2. *Determine the type of atresia by establishing whether there is a tracheo-oesophageal fistula*. The presence of a distal TOF is done by taking a plain X-ray of the torso: air below the diaphragm, within the bowel, confirms that there must be a connection between the trachea and stomach (i.e. a distal tracheo-oesophageal fistula). No gas below the level of the diaphragm implies no distal fistula (Figure 2). The further investigation of an infant with a 'gasless abdomen' is described later.
3. *Identify any coexistent abnormalities that may influence the management of the OA*. The key abnormalities are those of the VACTERL association, CHARGE association and major chromosomal abnormalities.

Most significant congenital heart disease is now diagnosed on antenatal ultrasonography, and the passage of urine after birth signifies functioning kidneys. However, most centres still perform echocardiography and obtain a renal ultrasound scan. Examination of the perineum will diagnose an anorectal malformation, and the thoraco-abdominal plain X-ray (already performed to diagnose a distal fistula) also allows assessment of the vertebrae and ribs. Radial and thumb abnormalities are evident on clinical examination. Sometimes these abnormalities influence the immediate management, including the timing and type of surgery. For example, duct-dependent congenital heart disease may require immediate commencement of a prostaglandin E infusion, and surgery to the OA is deferred until the infant is stable. If there are multiple gastrointestinal abnormalities, the correct order of repair is from proximal to distal. This means that closure of the fistula and repair of the oesophagus precedes duodeno-duodenostomy for duodenal atresia, and finally a colostomy for any concomitant high anorectal malformation.

Major chromosomal aberrations occur in about 7% of cases of OA/TOF. The most important of these are trisomy 13, 18 and 21. If suspected, urgent karyotyping is performed because, for example, if trisomy 18 is present, definitive surgery for the OA may not be appropriate.

Initial treatment

The normal care and monitoring of the surgical newborn is initiated. Specific attention is paid to maintaining normothermia at a time where multiple other investigations and interventions are taking place.

In addition to this, it is essential that continual or frequent aspiration of the saliva accumulating in the upper oesophageal pouch is initiated to prevent aspiration. This is achieved by insertion of a suction catheter with frequent aspiration of saliva, or positioning of a Replogle tube with continuous suction is required. This continues until the definitive operative repair has been performed.

The parents should be carefully informed throughout, updating them about what is planned, including details of the purpose and likely outcomes of all procedures. They will need to provide informed consent for the operation.

Surgery

The definitive operation (Figure 3) involves:

- division and closure of the tracheo-oesophageal fistula; and
- joining the two blind ends of the oesophagus together (end-to-end oesophageal anastomosis).

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