Paediatric Respiratory Reviews 19 (2016) 3–9

Contents lists available at ScienceDirect

Paediatric Respiratory Reviews

Mini-symposium: Oesophageal Atresia and Tracheo-oesophageal Fistula

Peri-operative management of neonates with oesophageal atresia and tracheo-oesophageal fistula

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EDUCATIONAL AIMS

The reader will come to appreciate that:

- Oesophageal atresia is often part of a broader sequence and thorough postnatal assessment for associated anomalies is required
- Awareness and appropriate management of peri-operative morbidity is imperative to optimising outcome
- Longer term neurodevelopmental outcomes are currently being explored, and once better understood, may allow improved understanding of the perinatal risk factors that mitigate risk for impaired long term outcome. This may translate into better informed peri-operative management.

ARTICLE INFO

Keywords:
Oesophageal atresia (OA)
VACTERL
Screening
Ventilation
Peri-operative
Outcomes

SUMMARY

Oesphageal atresia is a relatively common congenital anomaly that requires urgent diagnosis, transfer to a neonatal surgical centre and management by a multidisciplinary team. Peri-operative management requires vigilant monitoring for the many possible associated morbidities. There are unique anaesthetic, airway and ventilatory considerations for this group of patients. Beyond the perinatal period, systematic neurodevelopmental follow-up is recommended to better understand the longer term outcomes for these children.

Oesophageal atresia (OA), with or without tracheo-oesophageal fistula (TOF), is a congenital anomaly that occurs with a frequency of between 1:2500 -4500 live births [1,2]. Some families elect to terminate affected pregnancies making estimation of the true incidence imprecise, however Garne *et al* report that 11% of all cases of OA are terminated, with a further 2% dying in utero, and 87% of all cases being live births [3].

Data from 2010-2015 at The Royal Children's Hospital, Melbourne suggest a slight male preponderance to the condition (69:46) with a spread of birth across a wide range of gestational ages (Figure 1). With an average of 70,000 live births per year in the Australian state of Victoria over this six-year period, this gives a frequency of 1:3620. The median (range) gestational age at birth was 38 (25-42) weeks. This data is comparable to published historical cohort data from other centres [4,5]. Antenatal diagnosis may result in earlier delivery unrelated to intrauterine growth restriction or fetal compromise. In a European study fetuses with OA were delivered on average one week earlier if they had been diagnosed antenatally [3]. However the risks of early delivery and complications of prematurity need to be balanced against the safety of timely induced delivery.

Anatomical Classifications: There are a number of anatomical classifications of OA, and several classification systems (Gross [6], Vogt [7] and Ladd [8]) (Table 1). Gross type C (Vogt 3B) is the commonest type of OA, accounting for ~86% of cases.

Co-existent Anomalies: As many as 60% of newborns with OA will have co-existing anomalies [9]. Approximately half of those

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http://dx.doi.org/10.1016/j.prrv.2016.01.002
1526-0542/

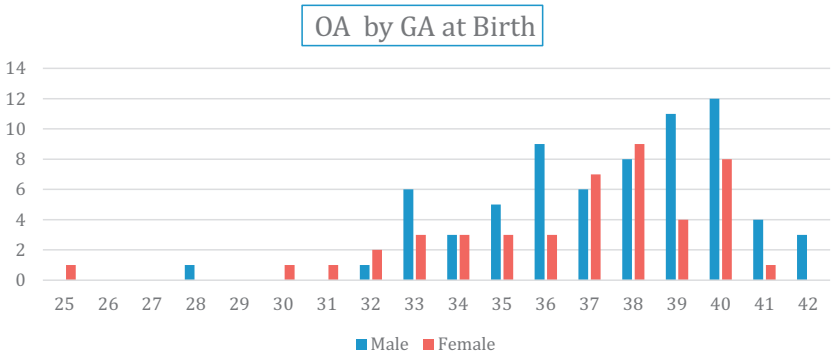

Figure 1. Spread of gestational ages at birth, stratified by gender, for babies with OA presenting to RCH, Melbourne (2010–2015).

infants with associated malformations will have VACTERL (Vertebral anomalies, anal atresia, tracheo-oesophageal fistula and/or oesophageal atresia, radial anomalies, cardiovascular, renal and limb anomalies) with vertebral and cardiac anomalies being the commonest. Other associated conditions include CHARGE (coloboma, heart defects, anal atresia, retarded growth, genital hypoplasia and ear anomalies), 22q11 deletion, trisomies and other chromosomal abnormalities [10]. In a small study from New Zealand, Kimble *et al* report that 80% of babies born with OA without a TOF had other anomalies [11]. Forty percent of these had cardiac anomalies, 20% had renal anomalies and 30% had a gut atresia or stenosis, most commonly an anorectal malformation. Orthopaedic anomalies also affected 50% of this small group, with lumbosacral or digital/limb anomalies being most common. In a retrospective cohort study of long-gap OA, co-existing anomalies were found in 60%, with cardiovascular anomalies being most frequent (47%) followed by gastrointestinal and genitourinary anomalies (27% each). 20% had VACTERL association [12].

PRE-OPERATIVE CARE

Between 20% and 31% of cases of OA are diagnosed antenatally with absence of a stomach bubble on ultrasound and/or presence of polyhydramnios [3,4]. Where antenatal suspicion is aroused, this is often investigated with fetal karyotype or SNP microarray. With the majority of affected patients being diagnosed postnatally, every baby delivered after a pregnancy complicated by polyhydramnios should have the patency of their oesophagus checked prior to leaving the delivery room, by passage of a nasogastric tube. Some newborns with excessive pooling of saliva may become apnoeic at delivery secondary to vagal stimulation of the vocal cords. This presentation also warrants assessment of patency of the oesophagus. Part of the routine examination of the newborn must include examination of the anus, as anorectal malformations are a known association of OA in the setting of VACTERL, especially in the absence of a TOF [11].

For those cases diagnosed postnatally, presentation is usually one of a "mucousy" baby, unable to cope with their saliva/secretions, and who upon orally feeding develops choking and respiratory distress. Diagnosis of OA is confirmed by the inability to pass an oro- or nasogastric tube more than 9 to 11 cm into the oesophagus, with a CXR showing the tip of the tube curled in the upper pouch at the level of T2-T4 (Figure 2).

It has been suggested by one author that introduction and increasing use of nasal CPAP in the management of preterm lung disease is associated with gaseous distention of the hypopharynx and cervical oesophagus, with potential for misdiagnosis of OA [13]. In practice however, this is rarely an issue.

Having made the diagnosis of OA, immediate management consists of withholding all oral feeds and medication, siting an intravenous line and providing maintenance fluids to ensure euvolaemia and normoglycaemia, and transferring the infant to a surgical neonatal intensive care unit (NICU). Placing the infant prone and elevating the cranial end of the cot may minimise the risk of pulmonary aspiration. Administration of antibiotic is not mandatory, but may be indicated where there is a strong suspicion of aspiration, or where there are infective perinatal risk factors (eg. group B streptococcal carriage in the mother, prolonged rupture of membranes or clinical suspicion of chorioamnionitis).

On arrival in the NICU, confirmation of the diagnosis should be undertaken by attempting to site a gastric tube. In a neonate with OA, a 10 French oro-oesophageal tube should arrest between 9-11 cm from the gums. With clinical certainty of the diagnosis, operative repair is arranged as soon as practicable.

Prevention of aspiration may be achieved either by regular intermittent oropharyngeal suction, or placement of a Replogle tube, allowing continuous suction of secretions. The latter option is preferred as it prevents repeated handling of the neonate, and potentially allows the neonate the opportunity to establish more physiological sleep patterns.

Neonates with a distal TOF are at a small risk of significant gastric distention that may compromise ventilation through diaphragmatic embarrassment, or gastric rupture causing pneumoperitoneum.

Table 1
Classifications of OA with their relative frequency.

Gross[3]	Vogt[3]	Ladd[3]	Name(s)	Frequency
-	Type 1	-	Oesophageal Agenesis	N/A
Type A	Type 2	I	"Pure", "Isolated" or "Long gap" Oesophageal Atresia	7%
Type B	Type 3A	II	Oesophageal Atresia with proximal TOF	1%
Type C	Type 3B	III, IV	Oesophageal Atresia with distal TOF	86%
Type D	Type 3C	V	Oesophageal Atresia with both proximal and distal TOFs	2%
Type E	Type 4	-	TOF only with no Oesophageal Atresia, H-Type	4%
Type F	-	-	Congenital Oesophageal Stenosis[6]	N/A