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Case Report

A Case Report of Congenital Oesophageal Atresia with Tracheo-Oesophageal Fistula and Review of the Literature

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ABSTRACT

Introduction
Congenital oesophageal atresia (COA) refers to a congenitally interrupted oesophagus. It is commonly referred to in the literature as oesophageal atresia (OA) with or without tracheo oesophageal fistula (TOF) but acquired TOF is a different entity.

Case Report
We present a case of OA with TOF which was not suspected antenatally despite the presence of polyhydramnios. The baby presented with respiratory distress and excessive oral secretions at the age of 3-hours following initiation of breastfeeding. Thoracoscopic repair was performed on the second day of life. The pre-, intra- and post-operative course was smooth with no major challenges. There was no associated anomalies in our case following thorough systemic evaluation including brain ultrasound, abdominal ultrasound, skeletal survey, ophthalmic assessment and echocardiography.

Conclusion
This case demonstrates the importance of maintaining a high index of suspicion for OA when faced with a combination of respiratory distress and persistent frothy oral secretions in a newborn. The antenatal and postnatal diagnostic approaches are discussed with highlights of associated anomalies and pre-operative assessment and management

Keywords
Congenital oesophageal atresia (COA); Tracheo oesophageal fistula (TOF); Oesophageal atresia (OA); Vertebral defects, anal atresia, cardiac defects, renal anomalies and limb abnormalities (VACTERL); Choanal atresia, retardation of growth, genital abnormalities, and ear abnormalities (CHARGE).

INTRODUCTION

Congenital oesophageal atresia (COA) refers to a congenitally interrupted oesophagus. It is commonly referred to in the literature as oesophageal atresia (OA) with or without tracheo oesophageal fistula (TOF) but acquired TOF is a different entity. The congenital lack of the oesophageal connection with the stomach in OA prevents swallowing, and this in turn prevents normal feeding and may cause the baby to aspirate accumulated saliva or milk leading to aspiration pneumonia. The incidence of COA is 1 case in 3000-4500 births. Maternal polyhydramnios occurs in approximately 33% of mothers with foetuses with OA and distal TOF and in virtually 100% of mothers with foetuses with OA without TOF.

The characteristic presentation of babies with OA is with drooling and excessive oral secretions with or without overt respiratory distress. The baby will typically choke upon starting breast or bottle feeding. This may progress to significant respiratory distress. The diagnosis of OA should be highly suspected if a large bore gastric tube appearance on a plain chest radiograph shows the tube to coil back in the proximal oesophageal pouch. OA occurs in association with other significant congenital anomalies in 30-60% of babies. The associated congenital anomalies are the major source of morbidity and mortality associated with OA. Early identification and management of OA prevents respiratory compromise and improves the outcome.

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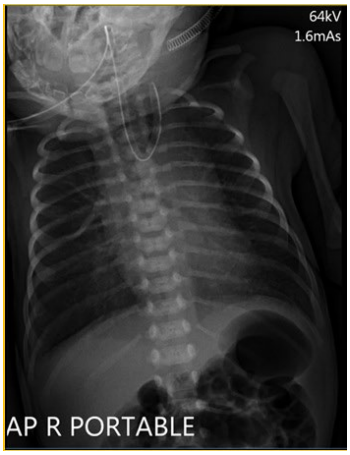
CASE PRESENTATION

A term male baby presented at the age of 3-hours of life with respiratory distress in the form of intermittent oxygen desaturations accompanied with tachypnoea and mild chest retractions. The lowest recorded oxygen saturation was 90% with no pre-post ductal oxygen saturations difference. Drooling and excessive thick oral secretions were noted. The secretions continued despite suction with the bulb syringe. The respiratory rate was 90-100 breath per minute. The baby was breastfed once prior to the onset of the respiratory distress. There was no reported choking. He passed meconium once. The 1st X-ray following admission showed the orogastric feeding tube to coil back inside the oesophagus at the level of the 4th thoracic vertebra (Figure 1). The diagnosis of OA was suspected. Furthermore, the presence of gastrointestinal aeration in the plain X-ray suggested the existence of a TOF. The respiratory distress progressed to mild respiratory acidosis which was managed by heated humidified high-flow nasal cannula. Intravenous antibiotics therapy was commenced following the septic screen. Total parenteral nutrition through peripheral intravenous central catheter (PICC) was initiated as the baby was kept nil by mouth.

Figure 1. *X-ray Chest and Abdomen Showing the Oro-gastric Feeding Coiling Back Inside the Oesophagus at the Level of the 4th Thoracic Vertebra*



The primigravida mother had a normal prenatal anomaly scan at 19-weeks of gestation. The foetal stomach bubble was seen during the anomaly scan with reported normal liquor volume. At 31-weeks of gestation the mother was diagnosed with gestational diabetes and polyhydramnios. A neonatal alert to the presence of polyhydramnios was not triggered to the neonatal team. There was no consanguinity and no family history of similar or other congenital anomalies.

The male baby, weighing 2361 grams, was born at 39+6-weeks of gestation *via* emergency caesarean due to foetal heart decelerations. He was born at Mediclinic Alnoor Hospital, Abu Dhabi, United Arab Emirates. He did not require active resuscitation at birth. The Apgar score was 9(1) and 9(5). The baby initial assessment was normal apart from the observed small for gestational age, low birth weight and mild ankyloglossia. There were no dysmorphic features pointing towards a recognisable malformation syndrome or pattern. The genitalia were normal with bilaterally descended testicles and the anus was patent.

Following the suspicion of tracheoesophageal fistula a large bore orogastric tube (size 10 F) was inserted to a predetermined length above the point of previous coiling inside the upper oesophageal pouch just above the level of the 4th thoracic vertebra. Low pressure continuous suction was applied to allow drainage and the head of bed was elevated to 450 to minimize the risk of aspiration. The paediatric surgeon was notified and the surgery was planned. It was decided not to use Riley double lumen suctioning tube as the surgery was planned for the following morning

Pre-operative evaluation was normal including cranial and renal ultrasounds. Thorough pre-operative assessment showed no signs of associated congenital malformations. The echocardiogram showed the aortic arch to be left-sided. There was non-significant moderate patent ductus arteriosus measuring 4 mm with left to right flow and a patent foramen ovale. Ophthalmic referral was arranged but there was no apparent iris coloboma. The skeletal survey did not show vertebral anomalies or radial dysplasia.

Thorascopic Repair

The baby was operated successfully on day 2 of life where the congenital defect was corrected with end-to-end anastomosis and separation of the fistula. This was done using thorascopic approach for the first time at our institute. The approach was right sided. There was no intra-operative complications and the baby did not require blood transfusion. Following the operation the baby required a short term mechanical ventilatory support. Analgesia was provided as required using the neonatal pain score. He remained nil by mouth for 48-hours following the operation. The baby was commenced on enteral Omeprazole in anticipation of the gastro-oesophageal reflux which is a universal finding in babies with OA. Enteral feeding using continuous milk pump was commenced with exclusive expressed breast milk and gradually increased till the total daily intake was achieved. On the 7th post-operative day, fluoroscopic guided contrast was done, followed by full reinsertion of the gastric tube under fluoroscopic guidance. The contrast study was satisfactory with no post-surgical leak or stenosis at the oesophageal anastomotic junction. Furthermore, the study showed normal caliber and mucosal pattern of the rest of the oesophagus. It is of note there was minimal reflux aspiration which slightly delineated the upper trachea yet it was not through any tracheo-oesophageal connections. No gastric hiatus hernia could be identified.

Bolus enteral feeding was commenced, on day 10 of life, following the contrast study. Recurrent desaturations and occasional bradycardias were noted, but they were all self-correcting. Both disappeared after commencement of infant Gaviscon. The bolus feeding was progressed steadily with no further challenges. Breast feeding was commenced on day 11 of life and the baby was discharged home on day 15 of life. The follow-up in the clinic after one and 2-months showed a well-growing baby with normal growth and development.

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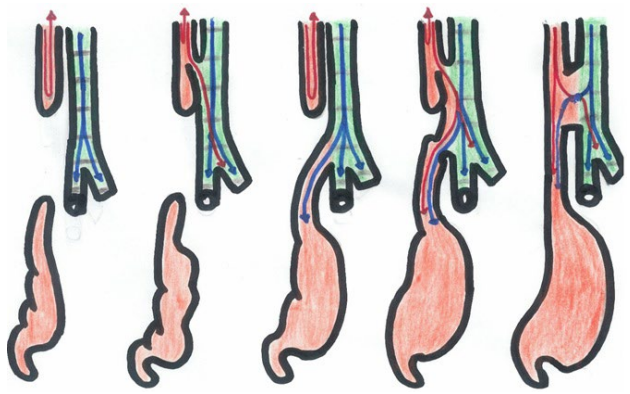
DISCUSSION

Congenital oesophageal atresia incorporates a variety of congenital anomalies with primary congenital interruption of the continuity of the oesophagus with or without a persistent communication with the trachea.[1] William Durston, in 1670, made the first english description of COA in a pair of conjoined twins.[2] In 1697, Thomas Gibson described OA and TOF with the most frequently encountered combination of trache-oesophageal anomalies, a proximal oesophageal atresia with a distal tracheoesophageal fistula.[3] His thorough description clearly described the symptoms and anatomy.

The majority of babies (86%) with OA has oesophageal atresia with distal tracheo-oesophageal fistula.[4] Pure OA (without fistula) is much less common occurring in approximately (6%) of total cases.[4] The characteristic feature of pure congenital oesophageal atresia is a gasless abdomen in the presence of oesophageal atresia. Gupta et al[5] in 2017 described an extremely rare case of an isolated membranous atresia causing near-complete obstruction of the esophagus. Isolated TEF is a very rare condition with scant epidemiological data.[6] Lewis Spitz reported cited an incidence of 4%.[1] The five subtypes of oesophageal atresia (Gross and Vogt Classification) are shown in Figure 2. OA with distal tracheoesophageal fistula is the most frequent type. Our case represents the most common type, type C with the frequency of 86% of all cases.[1]

Figure 2. *The Gross and Vogt Classification for Oesophageal Atresia*



The aetiology of OA is not clearly defined. It is very likely to be a multifactorial complex disease, with a combination of mainly genetic factors and added environmental uterine components.[7] There are no known human teratogens causing OA. However, recurrence of OA in families has been reported. Earlier studies have shown the estimated risk of recurrence in a sibling is 2% when an index case is affected. However, Choinitzki et al[8] results in 2013, in contrast to previous studies, suggested a very low recurrence risk for isolated OA/TOF and/or for malformations of the vertebral defects, anal atresia, cardiac defects, renal anomalies and limb abnormalities (VACTERL/VATER) association spectrum among first-degree relatives. The genetic component is further supported by the occasional association of COA with Trisomies 21, 13, and 18.[8] Tal Weissbach (Prenatal Diagnosis, July 2020) reported an increased incidence of oesophageal atresia with/without TOF (EA/TEF) among twins.[9] Nevertheless, genetic causes can be identified in less than 10% of the patients with EA.[10] Yet, it is estimated that 30-60% of the babies with COA with TOF has associated anomalies.[10] It is recognised, that associated anomalies as well as prematurity-related problems significantly affect the morbidity and mortality of COA babies.[11] The described anomalies include but not limited to tracheoesophageal fistula, urogenital system anomalies, VACTERL/VATER association. Coloboma, heart defects, choanal atresia, retardation of growth, genital abnormalities, and ear abnormalities (CHARGE) are also described.[11] Furthermore, neural tube defects, hydrocephalus, tethered cord and holoprosencephaly has been described in association with OA.[11] Other reported associated anomalies include duodenal atresia, ileal atresia, hypertrophic pyloric stenosis, omphalocele, intestinal malrotation, meckel diverticulum, unilateral pulmonary agenesis, diaphragmatic hernia, undescended testicles, ambiguous genitalia and hypospadias.[11] Porcaro F et al[11] reported airway anomalies such as tracheomalacia and tracheobronchial malformations in more than 40.5% of their cohort of 105 consecutive neonates with OA. This contribute to recurrent respiratory exacerbations due to impairment of the mucociliary function. The prognosis is generally good in the absence of these and other significant comorbidities or extreme prematurity. Concomitant laryngotracheal abnormalities are in particular associated with increased morbidity and higher mortality in babies with OA.[11]

The presence of right aortic arch is a rare finding in babies with OA with an incidence of 5%.[12] A recent multi-institutional retrospective study in the United states concluded surgical repair for neonates with right aortic arch is technically feasible *via* thoracotomy from either chest side.[12] However, the same study reported a higher incidence of anastomotic strictures with the right side approach.

The features suggestive of the diagnosis of OA on antenatal ultrasound are small or absent stomach with or without unexplained polyhydramnios.[11] These findings should not be taken as diagnostic as they have a very low positive predictive value. The assumption of the oesophageal atresia based on these findings may erroneously affect the clinical decisions with implications for timing or site of delivery.[11] Caroline Prady and her colleagues in 2019 published[13] a large systematic review and meta-analysis of the prenatal detection of OA. They concluded ultrasound alone is a poor diagnostic tool for identifying OA prenatally, due to the high rate of false positive diagnoses. They recommended magnetic resonance use plus amniotic fluid analysis following the ultrasound suspicion of OA. This recommendation was proposed many years ago by Langer et al.[14]

The post-natal clinical presentation of undiagnosed OA is variable but primarily reflects the effect of inability to swallow milk or the baby's own oral secretions. Frothy secretions which do not clear or recur after suction are typical findings in the majority of cases of OA. Some babies present with respiratory distress, choking and oxygen desaturations due to the reflux of the accumulated secretions from the hypopharynx into the trachea. The H-type fistulas symptoms are related to their caliber.[15] Gastric distension leading to persistent airway secretions in large fistulas presents

with respiratory distress, whereas small ones present with recurrent cyanotic episodes due to saliva and milk aspiration.[15]

Immediate Management of Suspected Cases

At birth, a 10-12 French gauge gastric tube should be passed through the mouth into the oesophagus for any baby born of a pregnancy complicated by polyhydramnios or if the antenatal scan findings are suggestive of oesophageal atresia. The same should be applied to babies presenting within 24-hours of birth with choking episodes, excessive oral secretions or respiratory distress. For preterm babies size 8 F gastric tube is adequate. Failure to pass the gastric tube beyond 9-10 cm from the lower alveolar ridge has been universally recognized as the classical sign of OA.[1] A plain X-ray of the chest and abdomen would then demonstrates the tip of the gastric tube to coil back within the superior mediastinum. The presence of gas in the stomach and intestine indicates the presence of a distal TOF.[1] However, it should be noted that radiological demonstration of a catheter reaching the stomach does not exclude the diagnosis of OA, as the gastric tube may take an alternative route (through the laryngeal inlet, trachea, tracheoesophageal fistula, and distal oesophagus to reach the stomach), which is a rare but well-known scenario.[16,17]

All babies with suspected OA should be managed within the neonatal intensive care unit as they should be kept nil by mouth with adequate intravenous fluids and nutrition. The baby should be nursed with the head raised around 450. Continuous or frequent intermittent low pressure suction should be applied to prevent the salivary secretions from accumulation as this may lead to aspiration pneumonia. A Replogle tube for continuous drainage of saliva from the upper oesophageal pouch should be considered in cases with very copious secretions or if the surgical repair is likely to be delayed beyond 48-72-hours. It should be positioned 0.5 cm above the distal end of the upper oesophageal pouch.

Broad-spectrum antibiotics (Penicillin or Ampicillin- and Gentamicin) are traditionally administered while preparations are made for surgery. It is advisable to avoid routine endotracheal intubation because of the risk of iatrogenic gastric perforation resulting from stomach distension through ventilation through the TOF. Accurate pre-operative radiological estimation of the gap between the upper and lower oesophageal pouches is essential in order to define the most appropriate surgical plan.[18]

Associated anomalies should be ruled out by thorough clinical examination and radiological assessment as they are mainly responsible for the medium- and long-term prognosis in these patients.[17] Echocardiography is highly advisable prior to the surgical correction. The surgical approach may be modified in the presence of a right-sided aortic arch.

The Surgery and Post-Operative Care

The recent years has seen major advances in the surgical approach for OA and TOF with survival now exceeding 90%.[18] The use of thoracoscopic surgery has minimised the long-term musculoskeletal morbidity associated with open surgery. However, the improved survival in recent decades is most attributable to better neonatal anaesthesia and peri-operative care. Despite this, long-gap OA still poses a number of challenges, and oesophageal replacement still may be required in some cases.[19]

Post-operative ventilation, if required, should be weaned as soon as possible. Adequate analgesia should be provided using neonatal pain score. Total parenteral nutrition is usually required as advancement of feeding may take few days. The trans-anastomotic chest tube may be removed within 48-72-hours if no leak is identified on the X-ray and the output of the chest drain is minimal. It has been customary to start the baby or continue antireflux medications as gastro oesophageal reflux is universal following the anastomosis.

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Nursing Pearls

• Antenatal diagnosis of polyhydramnios should be communicated to the neonatal team.
• Wide pore gastric tube should be inserted in all cases of antenatal suspicion of congenital oesophageal atresia. This includes cases of polyhydramnios.
• The combination of respiratory distress with recurrent excessive oral secretions or choking is highly suggestive of oesophageal atresia.
• Continuous or frequent intermittent suction (every 10-15-minutes) of the upper oesophageal pouch should be applied in all suspected or confirmed cases.
• A Replogle tube should be considered for continuous suction if the surgery is likely to be delayed more than 48-72-hours after presentation of if the secretions are very copious.

CONCLUSION

The diagnosis of COA should be suspected in the antenatal period if there is unexplained polyhydramnios and or if the stomach bubble is small or absent. Excessive frothy oral secretions, in the newly born baby, with or without accompanying respiratory distress are the typical post-natal presenting symptoms. The main modality of treatment is surgical correction which consists of end-to-end anastomosis of the proximal and distal oesophageal pouches and separation of any coexisting TOF and anastomosis of the oesophageal segment postnatally. Thorough pre-operative assessment is required as associated anomalies occurs in 30-60%. The long-term complications are common in surviving cases.

CONSENT

Signed consent is available.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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