CASE BASED DISCUSSIONS

Case-based discussion from the neonatal intensive care unit: a case of an intentional oesophageal intubation

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ABSTRACT

Tracheal agenesis (TA) is a rare congenital defect consisting of complete or partial absence of the trachea below the larynx. Antenatal diagnosis is challenging, and most cases are detected in the postnatal period. Airway management of such cases, particularly in the absence of antenatal diagnosis, can be challenging. Various methods of management have been described but with limited success, and overall prognosis remains very poor. We present an unexpected case of TA, highlighting management issues and diagnostic methods.

KJ AND SB (TRAINEE PAEDIATRICIANS)

A male infant (baby B), twin one of monochorionic-diamniotic twins, was born at 32+4 weeks' gestation weighing 1410 g (10th centile). Pregnancy was complicated by antenatal diagnosis of tetralogy of Fallot, polyhydramnios and intrauterine growth restriction. The mother presented with spontaneous onset of preterm labour, and placental abruption was suspected. Emergency caesarian under general anaesthesia was performed. Baby B was born with poor respiratory effort; Apgar scores were 3, 5, 6 and 8 at 1, 5, 10 and 15 min, respectively. Resuscitation consisted of immediate non-invasive ventilation via Neopuff t-piece and intubation. Intubation was reported to be technically difficult, with successful ventilation via a 3.0 oral uncuffed endotracheal tube (ETT) placed by a neonatal consultant following three unsuccessful attempts by junior staff, fixed at 8.5 cm at the lips. No audible cry was documented. Chest X-ray showed the ETT tip at the level of T2–T3 vertebral bodies, with bilateral equal lung expansion.

Initial management of baby B was at the local hospital. He remained intubated on minimal ventilatory settings (pressures 12/4 cmH_2O, fractional inspired oxygen 30%) with acceptable blood gas parameters and received one dose of surfactant administered via the ETT. Of note, he experienced several episodes of unprovoked desaturations, requiring suctioning, prolonged periods of intermittent positive airway pressure and minor tube position adjustments. On one occasion, 30 s of cardiopulmonary resuscitation was performed for bradycardia associated with a hypoxic episode.

Prostin (dinoprostone/prostaglandin E2) was commenced at birth due to antenatal diagnosis of congenital cardiac disease but ceased when postnatal echocardiography confirmed non-duct-dependent circulation. The baby became hypotensive at 12 hours of age, requiring inotropic support. A nasogastric tube (NGT) was easily passed, with the tip visualised within the stomach on X-ray. Baby B subsequently developed bilious vomiting and a 'triple bubble' on abdominal X-ray, suggestive of proximal jejunal atresia. Urgent overnight transfer to a tertiary centre was arranged for surgical opinion.

On arrival at the tertiary unit, baby B was stable, with ventilator pressures of 13/4 cmH_2O, a rate of 40/min, in 25% oxygen. Blood gases demonstrated a mild respiratory acidosis (pH 7.24, partial pressure of carbon dioxide 7.6 kPa). The baby was noted to have copious oral secretions requiring regular suctioning. Anteriorly placed anus and abnormal vertebrae on X-ray were noted. Urgent surgical review, as well as investigations including standard bloods, chest and abdominal X-rays, intenstinal contrast studies and echocardiography were requested.

AN (PAEDIATRIC) AND ST (NEONATAL CONSULTANT)

The history of polyhydramnios, in combination with other suspected congenital abnormalities, raises the possibility of tracheal or oesophageal abnormality (most commonly tracheo-oesphageal fistula) as part of a VACTERL spectrum. Passage of an NGT does not exclude rarer variants, such as H-type trachea-oesophageal fistula. Given the initial relative stability of the patient, relevant early but non-urgent investigations should be planned (echocardiography, gastrointestinal contrast studies, genetics and renal and spinal ultrasonography).

KJ AND SB

Shortly after arrival, baby B had a sudden respiratory deterioration, with desaturation and bradycardia. This was presumed secondary to a blocked ETT, which was therefore removed. He was successfully ventilated via Neopuff. Transillumination for pneumothorax was negative. Following adequate oxygenation and cardiovascular stability, rapid sequence induction (fentanyl, atropine and suxamethonium) reintubation was attempted. A grade 1 airway was reported by both senior neonatal medical trainees present, with easy visualisation of arytenoid folds at laryngoscopy; however, there were no readily discernible vocal cords. Although

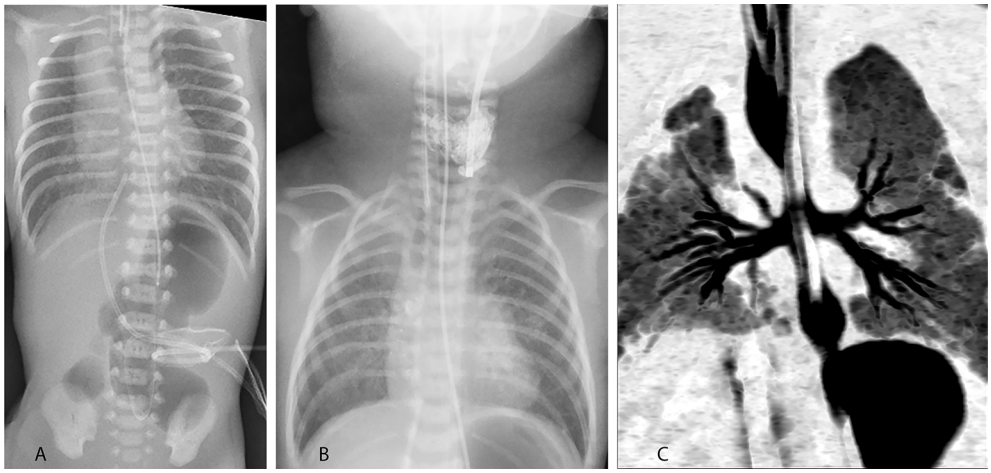

Figure 1 Admission chest X-ray (A) showing absence of tracheal shadow below the tip of the endotracheal tube and very hyperexpanded and hyperlucent lung fields. Subsequent X-ray (B) following instillation of perfluorocarbon radiopaque contrast showing it pooling in the blind ending trachea. CT chest (C) confirming findings of tracheal agenesis; right and left main bronchi appear to arise directly from the oesophagus, from which the stomach also arises below the diaphragm.

the ETT tip could be passed beyond the arytenoid folds, it would only advance approximately 0.5 cm, despite use of smaller ETT (down to size 2.5), gentle corkscrewing and adequate muscle relaxation. Capnography was persistently negative. In recognition of an abnormal airway, ventilation was continued via Neopuff, while urgent contact was made with the non-resident on-call neonatal consultant and on-site anaesthetist. Subsequent review of the admission chest X-ray revealed absence of tracheal shadow below the tip of the ETT, hyperexpanded and hyperlucent lung fields (figure 1A).

AN AND ST

Regardless of the cause, this is an unanticipated emergency airway situation. The incidence for 'cannot intubate, cannot oxygenate' (CICO) specifically within a neonatal unit is not known but is rare. Airway complications in neonates are estimated at 0.6%.[1] Difficulties with face mask ventilation in the *paediatric* population are estimated at 0.02%, while difficult endotracheal (ET) intubation figures are estimated between 0.06% and 1.34%.[2] Of note, there is no standard UK protocol for management of a difficult neonatal airway, and many neonatal units in the UK do not have difficult airway or CICO guidelines nor stock equipment needed for advanced airway techniques.[3]

Appropriate measures were taken in gaining anaesthetic assistance and obtaining GlideScope portable video laryngoscope to optimise further airway examination. The clinical picture, including polyhydramnios, difficult initial intubation and multiple congenital anomalies, made the possibility of tracheal abnormality, including tracheal agenesis (TA), highly possible. In retrospect, it is likely that the ETT had always been sited in the oesophagus with false reassurance from positive capnography due to tracheo-oesophageal fistula.

KJ AND SB

Under the advice of the non-resident neonatal consultant, a size 4.0 ETT was placed into the oesophagus under direct visualisation to the same depth as that inserted at birth, with immediate capnography response, chest movement, tube misting and audible air entry. The tube was secured, and the baby was ventilated with gentle volume-limited ventilation. A large-bore NGT was sited and regularly aspirated.

The paediatric intensive care unit bronchoscopy team were consulted urgently, and bedside bronchoscopy was performed. A laryngeal mask airway with angle adapter was used, allowing examination while ensuring adequate ventilation throughout. An abnormal, hypoplastic larynx was identified, and perfluorocarbon radiopaque contrast instilled via what appeared to be a possible small laryngeal inlet showed caudal pooling (figure 1B).

CT chest demonstrated no patent trachea and oesophageal intubation. Unusually, both the right and left main bronchi appeared to arise directly from the oesophagus, from which the stomach also arose below the diaphragm (figure 1C).

Multidisciplinary discussion concluded that the lesion was not amenable to repair or reconstruction. Care was redirected to palliation with parental consent, and the baby passed away peacefully.

AN AND ST

TA is a rare congenital defect consisting of complete or partial absence of the trachea below the larynx. Estimated incidence is 1 per 50 000 births, with a male-to-female ratio of 2:1.[4] The condition occurs in association with other congenital abnormalities in 90% of cases, and there is an association with prematurity and polyhydramnios,[4] all of which were present in this case. Various factors, genetic and environmental, have been proposed, but no clear causation has been proven.[4]

The condition was first described in 1900 by Payne, with classification systems based on the presence and length of the tracheal remnant proposed independently by both Faro and Flody. Flody's classification system is most commonly used and separates TA into three types[4]:

- Type 1: oesophagus connected to distal trachea via fistula (incidence 11%).
- Type 2: oesophagus connected to carina via fistula (incidence 61%).
- Type 3: two main bronchi arise from oesophagus (incidence 23%).

Antenatal diagnosis is inconsistent, with most cases postnatally diagnosed. The most common presenting features in affected neonates include: respiratory distress, poor air entry despite apparent chest wall movement, absence of an audible cry and failure to pass an ETT beyond the vocal cords.[4]

CT imaging is the diagnostic gold standard. However, bedside bronchoscopy and use of radiopaque contrast may help delineate the defect in settings where an appropriately experienced team is available. Perfluorocarbon is a synthetic liquid fluorinated hydrocarbon with high solubility for oxygen and carbon dioxide. It is chemically and biologically inert, radiopaque, not metabolised in either kidney or liver and is the usual pulmonary and airway contrast agent used in this tertiary centre.[5]

Intentional oesophageal intubation provides a temporary life-saving measure in such babies but is only possible in the presence of tracheo-oesophageal or broncho-oesophageal fistula, with a large bore ETT placed just about the fistula. This procedure facilitates temporary ventilation; however, there are significant limitations to the longer term use of an oesophageal substitute airway: the calibre of the wall is weaker than that of the trachea, increasing the risk of desaturation, trauma, bleeding and even erosion and air leak.[6]

Various strategies, including EXIT procedures, emergency tracheostomy for initial airway management, as well as reconstructive surgery and oesophageal banding in the intermediate term have been described,[4] but with limited success, and overall prognosis remains very poor.

Formulation of consensus guidelines for advanced neonatal airway management, incorporating the key clinical indicators of abnormal airway anatomy, the role and limitations of capnography, as well as expanded use of novel airway imaging techniques, would facilitate more standardised management of newborn infants with unanticipated airway abnormalities, including TA.

Contributors AN conceptualised the case report based on a clinical case for which all authors were closely clinically involved. KJ drafted the initial manuscript. All authors participated in the revision of the initial and subsequent versions of the manuscript and approved the final manuscript as submitted.

Competing interests None declared.

Key messages

- Difficult neonatal intubation is rare but occurs; there is a need for UK consensus guidelines.
- Capnography confirms CO_2 presence and hence alveolar ventilation, not tracheal intubation.
- Intentional oesophageal intubation can be life saving in tracheal agenesis with tracheo-oesophageal fistula. This may be the only solution if presenting in situations with limited support but requires confident recognition of an abnormal airway.
- Radiopaque perfluorocarbon is a useful contrast agent for delineating airway anatomy.

Patient consent Guardian consent obtained.

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