CASE REPORT

Neonatal airway: challenging endotracheal intubation in infants with tracheal malformations at birth

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SUMMARY

Intubating newborn infants can be exacting. We describe two cases of endotracheal intubations in infants born with tracheal malformations. A male infant aged 30 weeks required intubation at birth for respiratory distress. Repeated attempts at intubation failed to achieve an optimal endotracheal tube position as the tube could not advance beyond the vocal cords. Hence ventilation continued with suboptimal air entry in the lungs. Bronchoscopy and CT scan confirmed tracheal stenosis. Slide tracheoplasty was successfully executed on day 78 of life. A female infant aged 33 weeks was intubated at birth for perinatal depression. Attempts at intubation were unsuccessful due to non-visualisation of the laryngeal inlet. Oesophagus was intubated and attempts to inflate showed air entry in the lungs, suggesting a fistulous communication between oesophagus and airway. A contrast oesophagogram showed a fistula connecting oesophagus and carina. With airway patency in question and associated major anomalies, parents were counselled and support was withdrawn.

BACKGROUND

Endotracheal (ET) intubation in preterm infants is often challenging, especially in the presence of airway malformations. Prompt recognition of airway malformations facilitates directed resuscitation and appropriate interventions, the delay of which could be fatal for the baby.

Successful intubation of a newborn infant at birth can be difficult, even for an experienced neonatologist. This may be due to anatomical variations of the airway or mechanical obstructions such as cysts and tumours. We present two cases: (1) congenital tracheal stenosis (CTS) and (2) tracheal agenesis (TA).

CASE PRESENTATION

Case 1

A male baby weighing 1100 g was born at 30⁺⁺ weeks gestation to a 29-year-old primigravida. The parents were non-consanguineous and maternal antenatal scans and serologies were unremarkable. At birth, he required resuscitation using Neopuff (Fisher & Paykel, New Zealand). At 15 min of life, ET intubation was attempted by a neonatal registrar because of increasing respiratory distress, but was unsuccessful. At 20 and 23 min of life, repeated attempts to intubate failed due to an inability to

push the ET (2.5 mm) tube beyond the vocal cords. Leaving the ET in suboptimal position with optimal oxygen saturation, the infant was ventilated and transferred to the neonatal intensive care unit. Reintubation was attempted with a 2 mm ET tube by a consultant. However, he found it impossible to push the ET tube beyond the vocal cords, raising the suspicion of tracheal stenosis. On auscultation, air entry was audible but reduced bilaterally. The baby did not have any dysmorphic features and his general physical and systemic examinations were unremarkable.

Case 2

A female infant weighing 1800 g was born at term by normal vaginal delivery to a 16-year-old single, unmarried mother. Polyhydramnios, renal pelvic dilatation and growth retardation were reported in the antenatal ultrasound scan done at 30 weeks by a private sonologist and was referred to the tertiary centre for detailed imaging. However, due to the fear of loss of confidentiality related to maternal marital status, family did not pursue with the follow-up scan. She was admitted in labour at term to the tertiary hospital. At birth, the infant was cyanosed, gasping and bradycardic. An experienced neonatologist attempted oral ET intubation. The vocal cords were not visualised clearly. Repeated ET intubation attempts failed. Neopuff mask ventilation was continued, showing chest excursions and air entry in the chest and stomach on auscultation. Further attempts to intubate past the cords were unsuccessful, despite using smaller ET tubes.

Full resuscitation was commenced, and intermittent breath sounds could be heard in both lungs. The heart rate increased to 100 beats per minute, but the infant's colour failed to improve and oxygen saturation was below 80%. Ventilation was continued with little success. Attempts by an otolaryngologist and anaesthetist to intubate using an introducer were also unsuccessful. An attempt was made to inflate the lungs by oesophageal intubation and ventilation. Lung air entry was audible, suggesting a fistula connecting the oesophagus and the lower airway.

INVESTIGATIONS

In case 1, the otolaryngologist attempted an urgent rigid bronchoscopy but the scope could not pass beyond the larynx, hence ventilation was continued

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Figure 1 (A) Coronal CT image showing the presence of an endotracheal tube (ETT) within the trachea (short arrow). Just distal to the ETT there is a stricture measuring 6mm in length (long arrow). (B) Three-dimensional reconstructed image showing the stricture (long arrow) just distal to the ETT tube (short arrow).

with suboptimal ET position. Baby was saturating well in moderate conventional ventilator settings. CT scan showed marked tracheal narrowing beyond the vocal cords, consistent with the diagnosis of severe CTS (figure 1A,B). The rest of the tracheal tree was normal. An MRI of the chest and upper airway was performed to exclude external soft tissue compression.

In case 2, the baby showed a rudimentary right ear, absent right thumb and index finger on examination. Anus was patent. An infantogram revealed an absent right radius and sixth dorsal hemivertebra (figure 2A). No significant murmur was noted on auscultation but echocardiogram could not be done, as the infant was very sick. While ventilation through the oesophagus was performed (figure 2B), a fistulous communication between the oesophagus and carina was delineated in the contrast oesophagogram done under digital fluoroscopy. There was retrograde filling of the carinal stump and both bronchi, confirming TA with carinoesophageal fistula.

Treatment

In case 1, the infant with tracheal stenosis remained ventilated until he weighed 2000g. He had episodes of desaturations, requiring adrenaline nebulisation and short courses of steroids. A slide tracheoplasty was done successfully on day 78 of life. Postoperatively he required high-frequency oscillatory ventilation and inhaled nitric oxide followed by continuous positive airway pressure support for few days. He was discharged on day 136 of life on tube feeds and nasal cannula.

Figure 2 (A) Skeletal survey showing the sixth dorsal hemivertebra (black arrow) and an absent right radius (white arrow). (B) Contrast oesophagogram showing an anomalous fistulous communication (black arrow) between the oesophagus and carina.

For case 2, with airway patency in question, and heart rate not sustainable with multiple congenital anomalies, the mother and her parents were counselled and the ventilator support was withdrawn.

OUTCOME AND FOLLOW-UP

Bronchoscopy was used to diagnose tracheal malformations. The infant with CTS recovered well after tracheoplasty. At 2½ years of age, her growth and neurodevelopment are within normal range. The infant with TA died after the life support was withdrawn. Family refused an autopsy on religious grounds.

DISCUSSION

Tracheal malformations, severe CTS and TA are rare, with an incidence of 1 per 50 000–60 000 live births.¹ Cantrell and Guild first noted CTS, and Payne described TA.^{2,3}

CTS develops secondary to absent membranous trachea.² There are three distinct types of CTS: generalised hypoplasia, funnel-like stenosis and segmental stenosis.² Segmental stenosis is seen in case 1. TA stems from the abortion of the lung bud outgrowth, and delayed formation of the bronchi and lungs.⁴ There are three morphologically distinct types of TA. Type I is characterised by agenesis of the proximal trachea and the presence of a distal tracheoesophageal fistula (TEF); type II is defined by a complete absence of the trachea and the presence of normal bifurcating bronchi as seen in case 2. In type III, the two main bronchi arise independently from the oesophagus.⁵

Bilaterally echogenic and enlarged lungs seem to be the typical prenatal sonographic signs of laryngotracheal obstruction in the absence of TEF, due to over distension of the respiratory alveoli by fluid.⁶ Prenatal diagnosis of TA is rare and only possible in the absence of TEF, as the patient would present with classical congenital high airway obstruction syndrome (CHAOS), characterised by enlarged hyperechogenic lungs, fluid-filled dilated trachea and bronchi, and an absent flow in the trachea during fetal breathing. As phospholipids are secreted into the amniotic fluid by lungs, the absence of it in cases of CHAOS may support the diagnosis of TA.⁷ CHAOS should be suspected in pregnancies with a history of oligohydramnios in early pregnancy and/or polyhydramnios later on in pregnancy due to compression of oesophagus resulting in decreased fetal swallowing.⁸ When TA without CHAOS is suspected, fetal MRI has been reported to be a promising diagnostic tool.⁹ Associated anomalies were detected in cases of TA and CTS. Antenatal ultrasound scan is an excellent tool for diagnosing associated limb defects. By 4–8 weeks, limb formation occurs and primary ossification centres develop by the 12th week in all the long bones.¹⁰ In case 2, the vertebral defect, absence of right radius, thumb and index finger were not detected in the fetal ultrasound scan, but the detection of other abnormal findings prompted the sonologist to refer the patient for further imaging at a tertiary centre, which she deferred.

A strong suspicion of tracheal malformation in a fetus during antenatal scan warrants referral to a tertiary centre for further evaluation and management. At birth, stridor and cyanotic spells can be the presenting symptoms in infants with CTS. Older infants can present with respiratory insufficiency. It is often found incidentally, or during a workup for wheezing in older children. At birth, TA and severe CTS can present with asphyxia, absent crying, respiratory distress and failure to intubate beyond the vocal cords.^{7,11} In both the reported cases, the presenting symptoms were severe respiratory insufficiency, failure to advance the ET tube beyond the vocal cords in case 1 and inability to intubate the trachea in case 2. A laryngeal mask

airway (LMA) may be considered as an alternative to tracheal intubation if face mask ventilation is unsuccessful in achieving effective ventilation in infants more than 34 weeks of gestation and weighing more than 2000 g. LMA is a supraglottic airway device designed to be inserted into the patient's hypopharynx. If tracheal intubation fails, LMA can be used as a rescue device, while preparing for tracheotomy.¹² Although tracheotomy is an option, its success depends on the length of trachea affected in TA. It can be successful if done distal to a short-segment severe CTS.¹³ In TA with TEF, alternative management lines include intubating the oesophagus and ventilating the infant using the fistula as a conduit, even though there will be cycles of stabilisation and deterioration as observed in case 2. Chest movement should be observed if this manoeuvre is useful.⁷ If oesophageal intubation can stabilise the infant, oesophageal ligation distal to the site of TEF can be considered to ventilate through the fistula and a gastrostomy is needed to feed the infant.¹⁴ In cases of complete or near-complete obstruction of the fetal airway with an antenatal diagnosis of CHAOS, an ex utero intrapartum treatment (EXIT) procedure can be planned in tertiary centres having the surgical expertise. Vaikunth *et al* reported the survival of the first case of TA causing CHAOS following EXIT.¹⁵

Rigid bronchoscopy remains the gold standard for diagnosis of tracheal malformations. Virtual endoscopy using high-resolution, multirow detector CT is used to evaluate the distal airway in CTS.^{7,11} MRI is useful in delineating associated cardiovascular anomalies.¹¹ In TA, using the oesophagus as an air conduit and inflating the chest strongly suggests TA in the presence of a fistulous communication between oesophagus and airway. Contrast oesophagogram under fluoroscopy is useful for diagnosis of TA, as shown in case 2.

Isolated CTS occurs in 10%–30% of cases. Cardiovascular anomalies occur in up to 70% cases of CTS, including pulmonary artery sling, Fallot's tetralogy, double aortic arch and transposition of the great arteries.¹¹ In case 1, CTS was an isolated anomaly. In both TA and CTS, associated anomalies that are gastrointestinal, renal and musculoskeletal have been reported, especially radial and vertebral defects. The infant in case 2 had three major congenital malformations including a vertebral anomaly, TA with carinoesophageal fistula and limb defects, satisfying the criteria of VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) association.^{16,17}

In CTS, the severity and extent of airway stenosis and the presence of associated anomalies determine the outcome. A subset of patients with CTS outgrows the stenosis by 7–9 years of age.¹¹ ET stents have been successfully used in few patients recently. Even though surgical repair of CTS has been performed as early as 1964, the outcome has been poor. Current surgical options include resection and primary anastomosis, pericardial patch or rib cartilage tracheoplasty, tracheal autograft, tracheal homograft and slide tracheoplasty, and tracheal transplant.^{11,18–20} Slide tracheoplasty is currently the operation of choice for CTS, first described by Tsang *et al* in 1989.^{19,20} The reported case of CTS was successfully subjected to slide tracheoplasty. Low morbidity and mortality in patients who underwent slide tracheoplasty with cardiopulmonary bypass for tracheal stenosis were reported in two large series.^{20,21} In older children, the indications of surgery are based on functional status: any patient presenting with severe respiratory symptoms meets the criteria for surgical intervention. A paediatric patient can tolerate up to 50% of the narrowing of the tracheal diameter before developing significant symptoms.¹¹

TA is considered a lethal anomaly. Resuscitation manuals do not mention what to do when challenged with a neonate who cannot be intubated.²² In infants with suspected TA, securing the airway using LMA or wilful oesophageal intubation can be lifesaving. Initial stabilisation with oesophageal banding and gastrostomy can provide effective ventilation and feeding. Recent reports of successful airway and oesophageal/alimentary reconstruction by Tazuke *et al*²³ seem propitious. Neotrachealisation using oesophageal stenting and establishing a gastrointestinal continuity using gastric or jejunal interposition have shown promising results.^{23,24} Reconstruction of trachea in future using regenerative medicine and tissue engineering of amniotic fluid cells to develop a tracheal graft following prenatal diagnosis may improve outcome in patients with TA.²⁵

Learning points

- ▶ When resuscitating newborn infants with difficult endotracheal intubation at birth, experienced medical staff should have a high index of suspicion of tracheal malformations.
- ▶ Prompt recognition of airway malformations enables swift execution of directed resuscitation and appropriate interventions.
- ▶ Many tracheal anomalies such as tracheal stenosis can be managed with favourable surgical outcomes.

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