CASE REPORT

Incidental diagnosis of an H-type tracheo-oesophageal fistula

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SUMMARY

A 6-day-old term neonate who was intubated on day 1 of life for apnoeic episodes, was transferred to the regional paediatric intensive care unit (PICU) for specialist opinion following 3 failed extubations in the neonatal unit. *Escherichia coli* congenital pneumonia was diagnosed and the child discharged to the local hospital. Chest radiographs and inflammatory markers were in keeping with infection. However, ongoing difficulties with secretions necessitated readmission to the PICU, following a significant cyanotic episode associated with coughing. On arrival at the PICU, a large leak around the endotracheal tube (ETT) was noted. On direct laryngoscopy, the ETT was found correctly positioned, through the cords, but air was noted to be coming back from the oesophagus. Advancing the ETT towards the carina terminated the leak and raised the suspicion of a tracheo-oesophageal fistula. An H-type tracheo-oesophageal fistula was confirmed on bronchoscopy. An uneventful fistula repair was performed and the baby discharged from the PICU on day 23 of life.

BACKGROUND

H-type tracheo-oesophageal fistulae account for only 2–3% of all tracheo-oesophageal fistulae.¹ Antenatal diagnostic sensitivity for all tracheo-oesophageal fistulae is only 9–42% and is more difficult with H-type fistulae, where the absence of a stomach bubble is not a diagnostic feature.² As a result of this, the diagnosis may be delayed without the clinician having a high index of suspicion. This case is particularly helpful as it shows how a diagnosis can be made at the bedside by an experienced intensivist through expert laryngoscopy.

CASE PRESENTATION

A female infant presented on day 1 of life, with apnoeas, on the postnatal ward of a district general hospital. She was born at 39+6 weeks, with birth weight 3.18 kg, by normal delivery after an uneventful pregnancy. There were no risk factors for sepsis and she was in good condition at birth. Initial chest radiograph was unremarkable. She was transferred to the local neonatal unit, where she was intubated and given intravenous antibiotics for a presumed congenital infection. Over the next 5 days, she failed extubation on three occasions (increased work of breathing and oxygen requirements together with copious oral secretions) and was subsequently transferred to the regional paediatric intensive care unit (PICU) for subspecialist review on day 6 of life.

On arrival in the PICU, the initial chest radiograph (figure 1) demonstrated evidence of consolidation, which led to a working diagnosis of congenital pneumonia. Inflammatory markers were also in keeping with infection. After performing a blind bronchoalveolar lavage, the antibiotic coverage was broadened. In view of the local team's concerns that there was an underlying cause for the failed extubations, the baby was reviewed by the respiratory, cardiac, neurology and ear, nose and throat (ENT) teams over the next few days. An echocardiogram demonstrated a structurally normal heart and the bronchoalveolar lavage cultured *Escherichia coli*. Neither the respiratory nor neurology teams noted concerns, and the ENT team's plan was to perform a bronchoscopy if there was a further failed extubation after treatment of the pneumonia. Regular chest physiotherapy allowed the infant's oxygen and ventilatory requirements to be weaned over the next few days. She was extubated on day 10 of life without event and transferred back to her base hospital the following day, self-ventilating in air, to complete a 7-day course of intravenous antibiotics.

In her local hospital, airway secretions remained problematic, causing frequent desaturations, especially while supine. Management of these episodes was symptomatic with facial oxygen and repositioning. These episodes culminated in a profound desaturation and bradycardia on day 14 of life, where milk was noted in the mouth and the baby was coughing profusely. For stabilisation, she

Figure 1 Chest radiograph showing endotracheal tube at T1 with patchy airspace disease in the upper lobes and in the left lower lobe in keeping with infection or aspiration.

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required reintubation, for which she was transferred back to the regional PICU.

On admission to the PICU, despite having an appropriately sized uncuffed endotracheal tube (ETT), seen in a good position on chest radiograph, a large leak was noted coming from around the ETT, and as this was felt to be affecting oxygenation and ventilation, a decision was made to change to a cuffed ETT. The baby was reintubated by the PICU Registrar, with the tube inserted to the same distance and the cuff inflated. End-tidal CO₂ was present, however, a large leak was still audible. Direct laryngoscopy was performed by the consultant paediatric intensivist to confirm tracheal placement in view of the unusual finding of a leak with the ETT cuff inflated. On insertion of the laryngoscope into the mouth, there was initial concern that the tube was not in the trachea due to the large amount of leak, however, on laryngoscopy, the ETT was clearly passing through the vocal cords, with the depth marker at the correct level. On further inspection, the air leakage was noted to be coming back up the oesophagus rather than from the trachea, raising concerns about the presence of an H-type tracheo-oesophageal fistula. With this in mind, the ETT was advanced under direct vision until the leak from the oesophagus disappeared (tip of ETT distal to the suspected fistula), at which stage the leak on the ventilator also disappeared, and ventilation and oxygenation significantly improved. Repeat chest radiograph showed the ETT to be at the level of the carina (T4) (figure 2).

INVESTIGATIONS

The diagnosis was confirmed on day 17 of life, when microlaryngoscopy and bronchoscopy demonstrated an H-type tracheo-oesophageal fistula halfway between the vocal cords and the carina (figure 3).

DIFFERENTIAL DIAGNOSIS

- ▶ Laryngeal cleft
- ▶ Congenital infection (pneumonia)
- ▶ Other structural upper airway abnormalities

TREATMENT

An uneventful fistula repair was carried out on day 19 of life and the baby was successfully extubated 2 days later. The closure was completed through a right cervical incision. During the surgical procedure, the oesophagus and trachea were noted to have a common wall lower down to the fistula. This was confirmed not to be an extension of the fistula, via a negative water leak test and a recheck bronchoscopy.

OUTCOME AND FOLLOW-UP

The baby made a full recovery and was discharged home from hospital on day 32 of life. She has been under follow-up with paediatric surgery, ENT team and general paediatrics since discharge. The early postoperative period was complicated by vocal cord palsy, which resolved spontaneously. On review at 4 months of age, she was feeding well and gaining weight adequately.

DISCUSSION

Tracheo-oesophageal fistula is a congenital communication between the trachea and oesophagus, affecting between 1 in 2000 and 1 in 4000 live births.² It occurs when there is defective separation of the oesophagus and trachea during embryogenesis.¹ Five anatomical subtypes are encountered in clinical practice, as described by Gross.³ Of these, the second least

Figure 2 Chest radiograph showing endotracheal tube at T4, at the level of the carina.

Figure 3 Bronchoscopic images of the fistula with operator annotations. Arrow indicating fistula.

common is the H-type fistula, which accounts for 2–3% of cases.¹

The underlying aetiology remains unknown but is likely multifactorial.² The majority of cases are sporadic with only ~1% being familial.² While various environmental factors have been implicated, no consistently strong evidence supports these claims.² Oesophageal atresia occurs in combination with other congenital anomalies in up to 50% of cases, commonly occurring as part of the CHARGE (coloboma, heart defects, atresia of the chonae, retardation of growth and/or development, genital and/or urinary abnormalities, ear abnormalities and deafness) and VACTERL (vertebral defects, anal atresia, cardiac defects, trachea-oesophageal fistula, renal abnormalities, limb abnormalities) associations.²

The diagnosis can be suspected antenatally on ultrasound in the presence of polyhydramnios with a small or absent gastric bubble.¹ However, overall antenatal sensitivity is only 9–42%.²

Most children with oesophageal atresia and tracheo-oesophageal fistula are diagnosed shortly after birth. They typically suffer recurrent cyanotic choking episodes on feeding attempts, with excessive upper airway secretions.^{1 2} This relates to the infants' inability to swallow due to significant oesophageal obstruction. Respiratory distress may result from aspiration events. The classical method of diagnosis involves the passage of a large gauge

Figure 4 H-type tracheo-oesophageal fistula with nasogastric tube sited within the stomach.

nasogastric (NG) tube, which arrests prior to reaching the gastric bubble on chest X-ray.^{1 2}

However, in an H-type fistula, the NG tube will pass freely into the stomach, so a high index of suspicion must be maintained (figure 4). As such, significant diagnostic delays can occur. The average duration of time from symptom onset until diagnosis is 14 days, although delays as long as 4 years have been described.^{4 5} These children may then present in later life with chronic cough, recurrent chest infection or feeding difficulties.^{1 4 6}

In our case, while the possibility of an upper airway anatomical anomaly was postulated, confirmation came with the disappearance of a significant ventilation leak on ETT repositioning. This was an incidental finding that prompted definitive investigation. As such, a tracheo-oesophageal fistula should be considered in a patient with a significant upper airway leak despite suitable ETT size and positioning (especially in the setting of recurrent aspirations or choking episodes).

Suspicion of an H-type tracheo-oesophageal fistula warrants bronchoscopy, oesophagoscopy and consideration for upper gastrointestinal contrast studies.¹ Contrast studies are valuable in confirming a diagnosis, however, worth noting is that, in a recent multicentre study, <50% of cases were confirmed in a single contrast study.⁷ Contrast studies may give false-negative results and one review article reports statistically significant improvement in diagnostic accuracy with direct visualisation (tracheo/bronchoscopy) compared with an oesophagogram.⁸ There are reports that all infants with an oesophageal atresia should undergo tracheostomy prior to surgical repair to visualise

the tracheobronchial tree.^{8–10} It is important to note, however, that there is little in terms of an evidence base to guide the definitive approach to surgical repair.⁹ Interestingly, one review article specifically highlights the importance of positioning an ETT between the fistula and the carina for these patients—in agreement with the approach for this case.¹⁰

Following diagnosis, the child should be kept nil by mouth until the fistula can be surgically divided.¹ Those with isolated tracheo-oesophageal fistulae without associated congenital abnormalities have a good outcome and live healthy lives.¹

Learning points

- ▶ A high index of suspicion is required to make a diagnosis of an H-type tracheo-oesophageal fistula (TOF).
- ▶ An H-type TOF is best diagnosed by conventional gold standard investigations.
- ▶ If a TOF is suspected, attempt to position the endotracheal tube (ETT) distal to the fistula (as close to the carina as possible) to facilitate adequate oxygenation and ventilation.
- ▶ The authors do not suggest that intubation is a gold standard or suitable test for the diagnosis of an H-type TOF, however, if a large air leak is detected, it is important to consider the diagnosis of a fistula and direct laryngoscopy will be of benefit to confirm appropriate ETT placement.

Competing interests None declared.

Patient consent Obtained.

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