Simultaneous anterior cervical repair of type IV laryngeal cleft and tracheo-oesophageal fistula

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DESCRIPTION

A boy with antenatal polyhydramnios, born at 38 weeks via normal vaginal delivery, required prompt intubation due to cyanosis. Following successful extubation onto high-flow nasal cannula (HFNC), he was transferred to a tertiary paediatric ICU for management of lung collapse and pneumonia. Microlaryngoscopy and bronchoscopy (MLB) identified a 3 cm posterior laryngeal cleft (LC) with a pit connecting the trachea to oesophagus at the midpoint between the distal cleft and carina (tracheo-oesophageal fistula, TOF). He was noted to have dysmorphic features; hypertelorism and hypospadias prompting genetic testing, confirming Opitz G/BBB syndrome.

At 9 days, he was transferred to a quaternary centre. Repeat MLB confirmed a type IV LC (figure 1), H-type TOF 1 cm superior to the carina and 0.9 cm from the cleft (figure 2) with distal tracheobronchomalacia. The repair of LC and TOF was performed simultaneously via an anterior cervical approach. The airway was entered anterior to the length of the cleft via a midline incision. The TOF was repaired with a double-layer closure, with simultaneous closure of the oesophageal mucosa superior and inferiorly, trachealis, tracheal mucosa and anterior tracheal wall. Admission continued on paediatric intensive care unit (PICU) (73 days) with transfer back to the local hospital once extubated onto HFNC.

Type IV LC is believed to be a result of incomplete fusion of the tracheo-oesophageal septum distal to the cricoid cartilage.¹ The Benjamin and Inglis classification is most common. Type I cleft is above the vocal folds (most common), type II extends below, type III extends through the cricoid cartilage with/without extension into trachea

Figure 2 Type 4 laryngeal cleft with type H TOF visible distal to the cleft.

and type IV extends into thoracic trachea (least common).² Symptoms range from asymptomatic to varying degrees of dysphagia, aspiration and stridor in type I, to respiratory failure in type IV, depending on severity of the cleft.¹ LC accounts for 0.2–1.5% of congenital laryngeal abnormalities. The gold standard for diagnosis is MLB.³ They are associated with other congenital abnormalities (laryngomalacia and TOF) and syndromes (Opitz G syndrome and VACTERL).⁴ TOF has an incidence of approximately 1 in 3500 live births; it is confirmed with contrast studies or bronchoscopy.⁵ Type-H TOF (without oesophageal atresia) accounts for only 3% of these. Presentation includes cyanotic episodes and respiratory distress due to aspiration.⁶ The incidence of TOF in children with LC is unknown. However, there are reports of an association in 12–45% cases.^{7–9}

Opitz G/BBB is a congenital disorder affecting midline structures; it is characterised by craniofacial, genitourinary and laryngotracheo-oesophageal defects.¹⁰ VACTERL is defined by the presence of three of the following: vertebral defects, anal atresia, cardiac defects, TOF and renal and limb abnormalities.¹¹

Type IV LC is most commonly repaired via anterior laryngotomy, avoiding damage to neurovascular structures.¹² Standard repair of proximal TOF is via lateral cervicotomy.^{13 14} For distal TOF, thoracotomy is preferred.^{15 16} Cohen presents a case of combined repair via lateral pharyngotomy.¹⁷ We present a case of a combined repair of a type IV LC and type-H TOF via the anterior approach, not previously reported, in a child with Opitz G syndrome.

Figure 1 Type IV laryngeal cleft with correctly sited endotracheal tube protruding posteriorly into the oesophagus.

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Patient's perspective

As a new mother, I found the experience very difficult. Due to the COVID-19 pandemic, only one parent was permitted to be with patients. I found it very difficult to deal with it all alone especially as I was far from home. However, the staff were very helpful. Since being discharged, I feel he has been very well and one wouldn't think there had been any problems other than having two scars.

Learning points

- ▶ Type I clefts can often go undiagnosed due to symptoms suggestive of more common pathologies. Whereas type III or IV clefts usually present in the first few days of life.
- ▶ A high index of suspicion is required for a tracheo-oesophageal fistula (TOF) in children with laryngeal clefts (LC) as they can co-exist resulting in worsening symptoms. We may consider future classifications of TOF to include LC.
- ▶ We advocate formal assessment with microlaryngoscopy and bronchoscopy at a tertiary paediatric centre for children with suspected LC.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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REFERENCES

- 1 Johnston DR, Watters K, Ferrari LR, *et al.* Laryngeal cleft: evaluation and management. *Int J Pediatr Otorhinolaryngol* 2014;78:905–11.
- 2 Leboulanger N, Garabédian E-N. Laryngo-tracheo-oesophageal clefts. *Orphanet J Rare Dis* 2011;6:81.
- 3 Griffith CL, Liversedge TFG. Laryngeal clefts. *BJA Educ* 2015;15:237–42.
- 4 Jesse C, Jonathan S, Jeremy N, Chen J, Stern J, Neuman J, *et al.* A rare case of laryngeal cleft in association with VACTERL and malrotation. *Radiol Case Rep* 2019;14:315–9.
- 5 Depaepe A, Dolk H, Lechat MF. The epidemiology of tracheo-oesophageal fistula and oesophageal atresia in Europe. EUROCAT Working group. *Arch Dis Child* 1993;68:743–8.
- 6 Donnelly P, McVea S, Flannigan C, *et al.* Incidental diagnosis of an H-type tracheo-oesophageal fistula. *BMJ Case Rep* 2016;2016. doi:10.1136/bcr-2016-215419. [Epub ahead of print: 29 Jun 2016].
- 7 Rahbar R, Rouillon I, Roger G, *et al.* The presentation and management of laryngeal cleft: a 10-year experience. *Arch Otolaryngol Head Neck Surg* 2006;132:1335–41.
- 8 Evans KL, Courteney-Harris R, Bailey CM, *et al.* Management of posterior laryngeal and laryngotracheoesophageal clefts. *Arch Otolaryngol Head Neck Surg* 1995;121:1380–5.
- 9 Fraga JC, Adil EA, Kacprowicz A, *et al.* The association between laryngeal cleft and tracheoesophageal fistula: myth or reality? *Laryngoscope* 2015;125:469–74.
- 10 Tajima H, Itoh H, Mochizuki A, *et al.* Case of polyhydramnios complicated by Opitz G/BBB syndrome. *J Obstet Gynaecol Res* 2010;36:876–81.
- 11 Solomon BD. VACTERL/VATER association. *Orphanet J Rare Dis* 2011;6:56.
- 12 Walner DL, Stern Y, Collins M, *et al.* Does the presence of a tracheoesophageal fistula predict the outcome of laryngeal cleft repair? *Arch Otolaryngol Head Neck Surg* 1999;125:782–4.
- 13 Genty E, Attal P, Nicollas R, *et al.* Congenital tracheoesophageal fistula without esophageal atresia. *Int J Pediatr Otorhinolaryngol* 1999;48(1):231–8. 25.
- 14 Riazulhaq M, Elhassan E. Early recognition of H-type tracheoesophageal fistula. *APSP J Case Rep* 2012;3:4.
- 15 Ou P, Seror E, Layouss W, *et al.* Definitive diagnosis and surgical planning of H-type tracheoesophageal fistula in a critically ill neonate: first experience using air distension of the esophagus during high-resolution computed tomography acquisition. *J Thorac Cardiovasc Surg* 2007;133:1116–7.
- 16 Crabbe DCG. Isolated tracheo-oesophageal fistula. *Paediatr Respir Rev* 2003;4:74–8.
- 17 Cohen SR. Cleft larynx. a report of seven cases. *Ann Otol Rhinol Laryngol* 1975;84:747–56.

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