

Case Report

Larygeotrache oesophageal cleft: a rare entity

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ABSTRACT

Larygeotrache oesophageal cleft is a rare anomaly in a newborn presenting with respiratory distress and choking with feeding symptoms identical with oesophageal atresia with or without tracheo-oesophageal fistula. It is an abnormal communication of the larynx and the trachea with the oesophagus occurring during fifth to seventh week of gestation as a result of cirroid cartilage failing to fuse dorsally. LTOC varies greatly in anatomical extent and clinical severity; more severe forms in which the some or all of the tracheal cartilaginous rings are incomplete are fatal unless corrected surgically. LTOC occurs in less than 1/10,000 to 1/20,000 live births with slightly male predilection and has a autosomal dominant mode of inheritance. Routine chest X-rays and barium oesophagogram are usually not conclusive but bronchoscopy will delineate anatomy of cleft clearly. We present detailed case report of neonate with respiratory distress and choking on feeding and illustrating maneuvers at endoscopy and subsequent individualized surgical management of child.

Keywords: LTOC, Respiratory distress, Choking, Bronchoscopy, Larynx, Cirroid cartilage

INTRODUCTION

Laryngotracheo oesophageal cleft (LTOC) though rare should be considered in the differential diagnosis of any newborn presenting with respiratory distress and choking with feeding. The estimated annual incidence of LTOC is 1/10,000 to 1/20,000 live births.¹ Once the cleft is diagnosed, it is essential to determine its length to orient the management and treatment approach. The prognosis is variable depending on the severity of the LC and associated malformations. Early diagnosis and appropriate treatment and management help to reduce mortality and morbidity.

CASE REPORT

A three day old full term male neonate 2.5 kg came to us with history of choking, dusky facial discoloration on feeds with respiratory distress. On examination, neonate had audible crepts on right side of lung. Haemogram was within normal range; 2D echocardiography showed 4 mm

ASD with patent ductus arteriosus. Ultrasound KUB and abdomen was within normal limits. X-ray showed right side pneumonitis (Figure 1). Cine-oesophagogram, using barium with patient in prone position and gradually withdrawing the end hole catheter, revealed spillage of dye into the trachea. Bronchoscopy using cystoscope with camera was done which showed a common channel extending beyond the cricoid lamina to the cervical trachea i.e. type 3 variety of tracheo-oesophageal cleft (Figure 2). Under general anesthesia, a large bore nasogastric tube (10F) was passed into the esophagus and keeping the endotracheal tube near the carina patient was approached by right cervical incision at the level of cricoid cartilage with a muscle cutting incision (Figure 3). Normal esophagus and trachea are identified (Figure 4). Stabilization of an endotracheal tube was done with a loop passed around trachea which also draws the endotracheal tube forward. The cleft was divided in a manner so that more tissue is left, with trachea to create neomembranous portion of neotrachea (Figure 5). While dissecting, vagus and the recurrent laryngeal nerves were

taken care. Closure of esophagus and trachea was done from caudal to cranial direction using absorbable vicryl sutures followed by placement of interposition sternocleidomastoid flap (Figure 6). Post operatively patient was kept on elective ventilation. But develop bilateral pulmonary infiltrates and died on postoperative day 8 due to septicemia and acute respiratory distress syndrome.

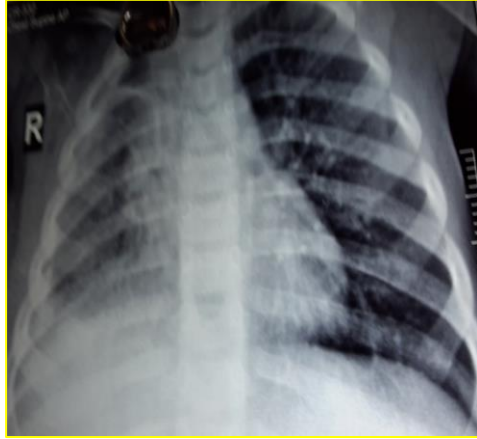


Figure 1: X-ray showing right side pneumonitis.



Figure 2: Bronchoscopy showing type 3 cleft.



Figure 3: Intraoperative photograph showing right cervical incision.



Figure 4: Intraoperative photograph showing normal esophagus and trachea.



Figure 5: Intraoperative photograph showing division of cleft.



Figure 6: Intraoperative photograph showing placement of interposition sternocleidomastoid flap.

DISCUSSION

A laryngotracheo oesophageal cleft is a congenital malformation characterized by an abnormal, posterior, sagittal communication between the larynx and the pharynx, possibly extending downward between the trachea and the esophagus. LTOC accounts for 0.2% to 1.5% of congenital malformations of the larynx.¹ A slightly higher incidence has been reported in boys than in girls with a ratio of 1.2 to 1.8.^{2,3} Laryngeal clefts result from failure of fusion of the posterior cricoid lamina and abnormal development of the tracheo-esophageal septum

which develops from the fourth and sixth branchial arches.⁴ LTOC appears to be mostly sporadic although some familial cases with suspected autosomal dominant transmission have been reported.⁵ The first patient with laryngeal cleft was described by Richter in 1792.⁶ In 1955, Pettersson reported the first surgical repair of a type 1 laryngeal cleft.⁷ Over the past 50 years, many classifications of LTOC have been proposed, all based on the downward extension of the cleft, Petterson (1955), Armitage (1984), Evans (1985), Benjamin (1989), Meyer (1990), DuBois (1990), and Sandu (2006).⁸ In 1989, Benjamin and Inglis presented a classification system in which 4 types of cleft were described: type 1 is a supraglottic interary-tenoid defect that extends inferiorly no further than the level of the true vocal folds; in type 2, the cricoid lamina is partially involved, with extension of the cleft below the level of the true vocal folds; type 3 is a total cricoid cleft that extends completely through the cricoid cartilage with or without further extension into the cervical trachea; and type 4 extends into the posterior wall of the thoracic trachea and may extend as far as the carina.⁹ Depending on the severity of the malformation, patients may present with stridor, hoarse cry, swallowing difficulties, aspirations, cough, dyspnea and cyanosis through to early respiratory distress. LTOC are frequently associated with various syndromes (16% to 68%) like opitz/BBB syndrome, Pallister Hall syndrome, vacterl/vater association, and charge syndrome.¹⁰ Routine chest X-rays are usually not conclusive and may only provide signs of parenchyma anomalies secondary to aspiration. Barium oesophagogram will show, contrast in oesophagus and trachea, however, it is difficult to differentiate whether there is spillage of dye due to cleft or spillover, at level of larynx.¹¹ For this purpose, it is better to do, oesophagogram in prone position, with the end hole catheter, which was slowly withdrawn, hence we will see the caudal most part of the cleft. Bronchoscopy will, delineate anatomy of cleft clearly. Under general anaesthesia during intubation preferential slippage of endotracheal tube into oesophagus raises suspicion of cleft. The posterior glottis must be carefully inspected and palpated, looking for a sagittal cleft between the digestive and the respiratory tracts, a LTOC may be inadvertently overlooked because of the redundant mucosa between the oesophagus and trachea, prolapsing into the cleft. Several probes have been designed and proposed for cleft palpation.¹² Once the diagnosis is made, it is fundamental to assess the length of the cleft. For endoscopy in our patients we have used cystoscope with camera. Once a LTOC has been diagnosed, a systematic course of action is critical first by ruling out associated anomalies by doing cardiac and renal ultrasonographies, spinal (cervical) X-rays and hearing screening. Medical management aims to: maintain satisfactory ventilation in children presenting an obstructive form of LTOC (mostly by prolapsing mucosa), prevent secondary pulmonary complications as a result of repeated aspiration and ensure adequate feeding of the child. In cases of respiratory distress

(possibly neonatal), an endotracheal intubation may be required. For grade 4 LCs, the stability of the tube in the airway and the quality of mechanical ventilation may be difficult to maintain. Placing the tip of the tube very close to the carina is helpful. If a tracheotomy is decided upon, the placement of the cannula also requires an endoscopic control. In extreme type IV LTOC, trachea and oesophagus are merged in one single tract and ventilation is extremely hard to maintain; the prognosis is therefore very guarded. In some cases of significant type III or type IV clefts, the risk of aspiration is so high that parenteral nutrition may temporarily be necessary. High grade LTOC often require a mid to long term gastrostomy (often with fundoplication).¹³ Two surgical standards exist for LCs: the external and endoscopic approaches. External approach; different cervical approaches are possible: lateral with lateral or posterior pharyngotomy, and anterior laryngotracheal depending on the type of LTOC and the experience of the surgical team. The lateral approach with lateral pharyngotomy has been recommended for low grade LCs with less than 2 cm of cervical trachea involved.⁸ Its drawback is the risk of recurrent and pharyngeal injuries to the nerves. The anterior laryngotracheal approach is the most widely used open technique. It provides an excellent exposure of the cleft with minimal neck dissection, and presents no risk of nerve damage.¹⁴ Several closure techniques has been described; a multi-layer closure after excision of excess mucosa and the use of asymmetric flaps with non-overlapping suture lines. In order to lower the risk of secondary opening, interposition grafts can be used, tibial periosteum or auricular cartilage, sternocleidomastoid muscle flap, fascia temporalis, costal cartilage, pleural flap, or even pericardium.¹³⁻¹⁵ Endoscopic approach has also been described in numerous publications for the closure of type I and type II LC.¹⁶ The risk of relapse of the LC (secondary re-opening of the cleft) requiring a revision is high: currently documented at 11% to 50% requiring multiple procedures are common.¹⁶ Tracheomalacia is a frequent issue in the post-operative management of type III and type IV clefts, and may require the use of a stent by tracheotomy excision of the malacic segment, aortopexy, or positive pressure ventilation.¹⁷ The overall mortality rate in a series of patients in 1983 was 46% and has dropped to 6%-25% in most recent reports.¹⁶

CONCLUSION

LCs are rare malformations of the larynx, whose prognosis is highly dependent on the extension of the cleft and other associated malformations. The improvement in survival in recent years can be explained not only by the advances in treatment and management, but also by an earlier diagnosis.

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REFERENCES

1. Mounghthong G, Holinger LD. Laryngotracheo-oesophageal clefts. *Ann Otol Rhinol Laryngol.* 1997;106:1002-11.
2. Andrieu-Guitrancourt J, Narcy P, Desnos J, Bobin S, Dehesdin D, Dubin J. Diastema or laryngeal or posterior laryngotracheal cleft, Analysis of 16 cases. *Chir Pediatr.* 1984;25:219-27.
3. Myer CM, Cotton RT, Holmes DK, Jackson RK. Laryngeal and laryngotracheoesophageal clefts: role of early surgical repair. *Ann Otol Rhinol Laryngol.* 1990;99:98-104.
4. Kluth D, Steding G, Seidl W. The embryology of foregut malformations. *J Pediatr Surg.* 1987;22:389-93.
5. Phelan PD, Williams H, Danks D. Familial occurrence of congenital laryngeal clefts. *Arch Dis Child.* 1995;72:98.
6. Pezzettigotta SM, Leboulanger N, Roger G, Denoyelle F, Garabedian EN. Laryngeal cleft. *Otolaryngol Clin North Am.* 2008;41:913-33.
7. Pettersson G. Inhibited separation of larynx and the upper part of trachea from oesophagus in a newborn; report of a case successfully operated upon. *Acta Chir Scand.* 1955;110:250-4.
8. Evans JN. Management of the cleft larynx and tracheoesophageal clefts. *Ann Otol Rhinol Laryngol.* 1985;94:627-30.
9. Benjamin B, Inglis A. Minor congenital laryngeal clefts: diagnosis and classification. *Ann Otol Rhinol Laryngol.* 1989;98:417-20.
10. Rose KG, Benz-Bohm G, Gunther H. Laryngo-tracheo-oesophageal cleft, clinical features, diagnosis and therapy. *Eur J Pediatr.* 1983;140:41.
11. Eriksen C, Zwillenberg D, Robinson N. Diagnosis and management of cleft larynx, literature review and case report. *Ann Otol Rhinol Laryngol.* 1990;99:703-8.
12. Parsons DS, Stivers FE, Giovanetto DR, Phillips SE. Type I posterior laryngeal clefts *Laryngoscope.* 1998;108:403-10.
13. Nicolas L, Eréa-Noël G. Laryngo-tracheo-oesophageal clefts. *Orphanet J Rare Dis.* 2011;6:81.
14. Albanese CT, Harrison MR, Jennings RW. Anterior cervical approach for repair of laryngotracheo-oesophageal cleft. *J Pediatr Surg.* 1998;33:400-2.
15. EN, Ducroz V, Roger G, Denoyelle F. Posterior laryngeal clefts: preliminary report of a new surgical procedure using tibial periosteum as an interposition graft. *Laryngoscope.* 1998;108:899-902.
16. Rahbar R, Chen JL, Rosen RL, Lowry KC, Simon DM, Perez JA, et al. Endoscopic repair of laryngeal cleft type I and type II: when and why? *Laryngoscope.* 2009;119:1797-802.
17. Mitchell DB, Koltai P, Matthew D, Bailey CM, Evans JN. Severe tracheobronchomalacia associated with laryngeal cleft. *Int J Pediatr Otorhinolaryngol.* 1989;18:181-5.

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