CASE REPORT

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TYPE IV LARYNGEAL CLEFT: A THERAPEUTIC CHALLENGE

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ABSTRACT

Type IV laryngeal cleft (LC) is a rare congenital malformation often associated with poorer prognosis compared to lower grades LC. The difficulty in managing LC lies in the ventilation prior to cleft repair, intraoperative anesthesia, technical difficulties to repair a longer cleft and higher rates of post-operative complications. We report a case of a premature neonate with type IV LC who underwent early gastrostomy at birth and survived with optimal medical management despite suffering bouts of pneumonia and life-threatening event. After a late cleft repair at 4-month old, she survived for 3 months post-operatively without complication of relapse fistula. Despite our cleft repair, she was unable to be weaned off from ventilator attributed to her poor neurological recovery and subsequently succumbed to death at 7 months. We would like to discuss the management challenges with regards to ventilation, approach of surgery as well as post-operative complication to improve the management of such complicated disease.

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Introduction

Type IV laryngeal clefts (LC) are rare congenital anomalies that occur 1 in 2000 to 50000 live births. It represents less than 0.3% of congenital laryngeal anomaly [1]. The major issues in the treatment of these patients are ventilator leakage and carbon dioxide retention problems originating from an inherently longer cleft defect in type IV LC, non-standardized approaches of operation, intraoperative exposure insufficiency, and post-operative complications. To our understanding, there has been very limited cases reported regarding type IV LC surgery in our country and we reviewed the literature to compare our experience in managing this therapeutically challenging disease.

Case summary

A 33-weeker girl was delivered prematurely via emergency caesarean section for breech in labour with birth weight 1.85 kg. Antenatally, the mother

had iron deficiency anaemia and polyhydramnios. She was delivered with APGAR of 5 at 1 minute, and 10 at 5 minutes. The baby developed respiratory distress syndrome at birth and required continuous positive airway pressure (CPAP). She was nursed in neonatal intensive care unit for respiratory support and total parenteral nutrition. She started to deteriorate at day 3 of life due to severe aspiration pneumonia after starting on nasogastric tube feeding and subsequently required intubation with endotracheal tube (ETT) size 3.5 mm. Chest radiograph revealed right lung

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consolidation. In spite of intubation, there was high volume of leakage noted. An echocardiography revealed a small atrioventricular septal defect, with no other apparent craniofacial abnormality. A diagnostic

microlaryngobronchoscopy revealed a complete laryngeal cleft (LC) type IV measuring 3.5 cm in length extending from interarytenoid down to carina (Figure 1). Severe tracheobronchomalacia was also noted.

Figure 1. Bronchoscopic findings and schematic drawing of the type IV LC post gastric resection. [A] Findings at the level of supraglottic showing the beginning of the cleft at the interarytenoid (arrow). [B] Findings at the mid region of the cleft with feeding tube to drain out gastric content seen passing into the oesophagus. [C] Lower end of the LC at the level of carina (arrow) and bilateral main bronchus (dashed arrow).

Figure 2. (A) Bilateral endobronchial tubes ventilation was utilized throughout surgery before converting to a tracheostomy. (B) Bilateral ETT connected to double lumen tube connector (arrow) allowing a single port for ventilation.

Midline laparotomy, partial transection of gastrectomy and gastrostomy was performed at the same time to avoid spillage of stomach content into the airway as well as to allow for enteral feeding. Post-operatively the baby had to be on continuous suctioning to avoid saliva overflowing into the airway. After multidisciplinary discussion, it was decided that surgical repair would carry a high risk of morbidity and mortality for the patient and surgery was postpone for the purpose of optimizing the nutritional status till a body weight of 3 kg was achieved.

During this period of observation and optimization of nutritional status, she developed recurrent episodes of pneumonia leading to sepsis and an episode of acute life-threatening event. However the baby thrived with good weight gain and had improvement of the neurological deficit despite the hypoxic threats.

The baby underwent surgical repair at 4-month old with weight of 4 kg by a combined anterior cervical and lateral thoracotomy approach. The anaesthesia and oxygen delivery was made possible by endoscopic guided bilateral endobronchial intubation (Figure 2A) with ETT size 2.5 mm into the right and left main bronchus respectively and were connected to a circle circuit via double lumen tube connector (DLT) to allow simultaneous ventilation using one common connector (Figure 2B). An early episode of hypoxia induced bradycardia develop during the initial phase of changing the endotracheal tube into endobronchial intubation and this was promptly resuscitated. Apart from that ventilation

was smooth throughout the entire length of surgery following bilateral endobronchial intubation with no issues of hypoxemia nor carbon dioxide retention and minimal leakage issues.

Surgical repair was done via an open anterior cervical approach using an extended laryngofissure made from thyroid notch till the lower end of cervical trachea for exposure of the cleft (Figure 3A). Incision was made along the mucosal borders of the cleft starting from cuneiform cartilage level down to caudal end of the cervical cleft separating the laryngotracheal and pharyngoesophageal layers. The two layers were then closed up with absorbable suture vicryl 5/0 interposed with fascial lata graft (Figure 3D – F). The defect in the anterior wall created after laryngofissure was left open for the next part of the surgery and also to provide a place for anchoring the bilateral endobronchial tubing without manipulation of the airway. The thoracic cleft repair was performed via a thoracotomy made from the right axillary approach. The right lung was retracted anteriorly and the visualised azygos vein was identified and ligated. The fistula was sealed by approximation of the dilated left and right anterior oesophageal wall using approximation with a fine Polydioxanone (PDS) 6/0 suture. Suture line between the lower thoracic trachea and oesophagus was reinforced with interposition of parietal pleural tissue. Lastly, the laryngofissure incision was closed approximating the anterior commissure. The bilateral endobronchial tube was then replaced with a tracheostomy tube size 3.5 mm uncuffed

and the tip of the tube was confirmed to be just above the carina with flexible endoscope.

Post cleft repair the child again developed high carbon dioxide retention secondary to severe malacic airway and excessive tracheobronchial secretion requiring regular bronchial toilet and readjustment of trachea tube. A barium contrast study was performed 2 weeks post-surgery (Figure 4) and confirmed to have no anastomotic

leak. However the baby had poor neurological outcome with spastic quadriplegia. The effort to wean off ventilation was futile as the child had later developed gastrointestinal tract infection, sepsis and soon after passed away at 3 months post-surgery.

Figure 3. (A) Anterior laryngofissure from the thyroid notch (dotted lines) till the lower end of the cervical cleft (solid arrow). (B) Exposure of the cleft after retracting the anterior tracheal wall laterally. (C) ETT tube seen passing through the oesophagus posterior to the cleft. (D) Approximation of the cleft using double layer suture. (E) The closure of cleft after suturing. (F) Dashed arrow showing the approximated trachea retracted to the left, while solid arrow showing insertion of fascia lata graft.

Figure 4. Barium study post contrast images showing smooth flow of contrast into the distal pouch of the transected stomach without evidence of anastomotic leak.

DISCUSSION

The most widely use classification of laryngeal cleft used to date is known as the Benjamin-Inglis classification [2]. This classification divides the cleft into 4 types, which are: type I involving the supraglottic, interarytenoid and above the vocal fold, type II where cleft extending into the posterior cricoid cartilage, type III extension of the cleft beyond the cricoid cartilage into the cervical trachea, and type IV also known as laryngotracheoesophageal cleft (LTEC) when the cleft extend into the thoracic trachea down to the carina as in our case. Other classifications has been described but are used less often [3].

Associations with genetic syndrome occurring in LC is reported in up to 16 to 68% of cases namely Opitz G, CATCH 22, Pallister Hall syndrome, VACTERL association and CHARGE syndrome [4]. In most recent study, type III and IV LC has been consistently associated with other comorbidities [5]. One such comorbidity includes cardiovascular malformation who was found present in all the patient who had type III and IV LC mostly diagnosed with atrial septal defect (ASD) followed by ventricular septal defect (VSD) which was present in our patient. Prematurity was found in up to 56% of children reported [5].

The hallmark of LC diagnosis is by direct microlaryngobronchoscopy. In low grade cleft as in type I and II, laryngoscopy with palpation of the interarytenoid area will confirm the diagnosis,

while bronchoscopy is required to assess the extent of the cleft in type III and IV LC.

Optimal ventilator support can be a challenging aspect for type IV LC especially during surgical intervention. Ideally, a tubeless airway is desired for adequate assessment of the airway and surgical intervention. In short length LC (type I and II), repair is usually performed endoscopically, and anesthesia can be achieved with induction using gas inhalation via a nasal prong or oxygen mask, followed by total intravenous anesthesia (TIVA) with advantage of maintaining spontaneous respiration [6]. Ventilation with a T-piece attachment to the side arm of bronchoscopy is a well-practiced technique. However, in type III and IV LC, the above technique does not address the possible large amount of gas leak owing to the longer length of cleft and hence we proceeded with bilateral endobronchial intubation made possible with endoscopic guidance prior to exposure of the cleft. We were able to maintain adequate oxygenation throughout the surgical repair with this technique despite an initial hypoxic event during the early stage of intubation. This bilateral endobronchial technique also offers the flexibility of manual ventilation as well as one lung ventilation during the stage of the alternate bronchial repair. To our knowledge, this technique has not been emphasized in other studies.

Timing of surgery should be as early as possible to negate the prolong effect of aspiration and hypoxia. In recent study [5], most type III LC underwent surgery within first 2 months of life while type IV LC within the first 4 weeks. However, as in our case where early surgery is difficult to achieve due to cardiorespiratory instability, jejunostomy and gastric ligation was performed first. A second stage surgical repair is usually done in 6-20 weeks once patient has achieved cardiorespiratory stability and adequate weight gain.

Surgical management of LC can be divided into external or endoscopic approaches. Endoscopic surgical repair is used for short length cleft such as type I and II LC with some type III. Endoscopic cleft closure is commonly performed using suspension laryngoscopy and with cold instrument, and endoscopic needle driver and knot pusher.

Open surgery is advocated for most type III and IV clefts. Open surgery can be further divided into cervical or thoracic approach. Cervical approach can be either from the anterior or lateral approach with the latter either as lateral pharyngotomy or posterior pharyngotomy. Anterior approach is recommended because it provides an excellent exposure of the cleft and possesses less risk of recurrent laryngeal nerve injury [1]. Lateral approach with lateral pharyngotomy is recommended with low grade LC with less than 2 cm cervical trachea involved [7] while posterior pharyngotomy can be addressed for deeper cervical trachea cleft. Two types of thoracic approaches have been described: anterior (with sternotomy) and lateral thoracotomy.

We used a one-stage, two-approach repair technique. The cervical tracheal repair is approach from a wide horizontal cervical incision followed by an extended anterior laryngofissure. Our combine approaches differ from the anterior cervicothoracic approach alone or in combination with limited median sternotomy used in other case reported [8,9]. Extended anterior laryngofissure [10] alone is limited inferiorly by the presence of the innominate artery. This technique has been used to approach the cleft up to 1 cm below the vessels [11]. However in our case the cleft extended well below the thoracic inlet this technique alone, do not provide sufficient access. Lateral thoracotomy provided

better access of thoracic trachea without the need for sternotomy and has also been recently described [12].

The gist of surgical therapy for type III and IV clefts is to recreate a separate functioning trachea and oesophageal lumen by means of various graft interposition to prevent relapse fistula [1]. Various tissues have been used as interposition grafts such as the pericardium, sternocleidomastoid muscle flaps, pleura, strap muscle, jejunum, and tibial periosteum [13]. In our case, a combination of fascia lata and pleural flap was used. Cleft closure usually includes two-layer closure with either asymmetric flaps with non-overlapping suture lines [14, 15]. More recent techniques have been described such as the usage of tracheal flap using strep muscles [15] or costal cartilage graft [9] with good functional separation result.

Common complications post-surgery include tracheoesophageal fistula, tracheomalacia, and persistent LC [5]. Tracheomalacia is a frequent issue post-operatively and this can be managed with keeping a tracheostomy tube for longer period of stenting with positive pressure ventilation [6]. In our case, child had few episodes of desaturation post-operatively requiring an endoscopic transtracheal evaluation and prolonged usage of tracheostomy due to tracheomalacia. However, we manage to maintain a functional separation between the airway and the digestive tract. This was proven by a repeated a barium swallow 2 weeks post-operatively.

In our case, withdrawal of ventilator was attributed to the poor neurological recovery of child. A substantial number of patients with LC have been found to have coexisting neuromuscular dysfunction [16] in the form of dysphagia or aspiration. LC patients to have high incidence of associated abnormal magnetic resonance imaging (MRI) of brain, such as white matter changes, ventricular irregularities and even Arnold-Chiari malformation. In retrospect, a formal referral to a neurologist and an MRI brain could have benefitted our child to look out for any concurrent brain abnormalities in this otherwise non-dysmorphic child.

The overall mortality of LC in recent decades of technology advancement appears to have dropped to 6-25%. The mortality rate for type IV

LC has also been reduced from 93% in 1985 to as low as 11% in 2020 [5].

CONCLUSION

Type IV LC remains an extremely rare entity and is a therapeutically challenging disease for any attending physicians. Cleft repair is recommended as early as possible. Despite the difficulties of ventilation, aspiration, surgery and its post-operative complication, long-term prognosis for type IV LC has improved over the years with better understanding of this disease.

KEY POINTS

- 1) In Type IV LC, surgery should be performed as soon as possible. Gastric ligation and jejunostomy should be performed if cleft repair needs to be delayed.
- 2) Bilateral endobronchial intubation is able to provide stable oxygenation and ventilation during surgery for type IV LC.
- 3) Combine approach of extended anterior laryngofissure with lateral thoracotomy provides adequate access for long segment type IV LC.
- 4) Tracheobronchomalacia remains the most common problem and parents should be informed of the longer duration of tracheostomy tube in their child.
- 5) Poor neurological recovery should arouse clinical suspicion for associated brain abnormalities and formal brain imaging should be undertaken prior to surgery.

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