

Laryngomalacia, laryngeal cleft and congenital unilateral vocal cord palsy: A unique case treated endoscopically without intubation or tracheostomy

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ABSTRACT

Laryngomalacia, congenital vocal cord palsy and laryngeal cleft are three separate pathologies which can independently cause stridor and failure to thrive in a neonate. We present a unique case all three entities. The diagnosis was confirmed on direct laryngoscopy and the cleft successfully repaired endoscopically. To our knowledge, this is the first case report where an infant presents with three contemporaneous pathologies; successfully treated endoscopically without intubation or tracheostomy.

Keywords: Laryngomalacia, laryngeal cleft, vocal cord palsy, paediatric, stridor, aspiration, failure to thrive

INTRODUCTION

Congenital laryngeal abnormalities are rare, occurring approximately 1 in 2,000 live births. Of these, less than 1% is due to laryngeal clefts. ^{1, 11} It was first described in 1792 by Richter ² and operated upon by Petersson in 1955. ³ Symptoms range from choking, aspiration, failure to thrive and stridor. It is an abnormal connection between the laryngo-trachea and oesophagus. There are many classifications on laryngeal clefts. ⁴⁻⁶

The most commonly used classification is by Benjamin & Inglis (Figure 1). ⁶

CASE REPORT

Baby A was born at term. There were no associated congenital cardiac, vertebral, fasciocervical and ear abnormalities. She had inspiratory stridor from birth and was diagnosed clinically with laryngomalacia. Worsening stridor and failure to thrive prompted a referral to our tertiary centre at age nine months. By this time, she also had cyanotic episodes/ cough during feeds and recurrent aspiration pneumonia. Clinical examination showed an infant with biphasic stridor and a

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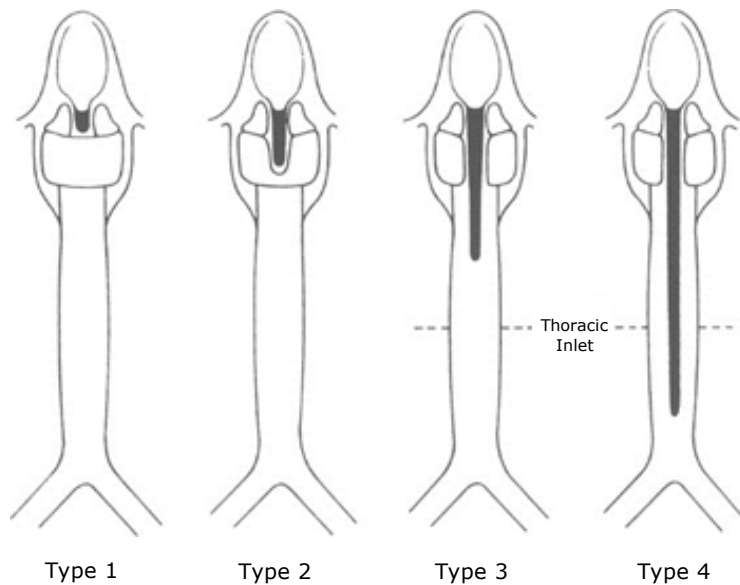


Fig. 1: The Benjamin–Inglis classification system of laryngeal clefts. Type 1 clefts are supraglottic interarytenoid clefts, where the cleft is above the level of true vocal cords. Type 2 clefts extend below the level of the vocal cords but do not involve the posterior cricoid lamina completely. Type 3 clefts extend completely through the cricoid cartilage, with or without further extension into the cervical tracheoesophageal wall. Type 4 clefts extend through the majority of the tracheoesophageal wall.⁶

predominantly expiratory wheeze. Fiberoptic laryngoscopy in clinic showed features of laryngomalacia with laryngeal cleft. She was started on a proton pump inhibitor for gastro-oesophageal reflux and fed in an upright position to prevent aspiration.

Fiberoptic laryngoscopy under spontaneous ventilation in the operating theatre confirmed the right vocal cord palsy and features consistent with laryngomalacia, i.e. a tubular epiglottis and redundant mucosa over both arytenoids (Figure 2a). We proceeded with direct laryngoscopy under suspension with a ventilating side port. A midline laryngeal cleft was seen posteriorly, extending to the upper border of the cricoid cartilage below (Figure 2b). The subglottis, lower airway and upper oesophagus was normal.

The patient underwent endoscopic repair of her laryngeal cleft without intubation the next day. She had LASER (*Light Amplifi-*

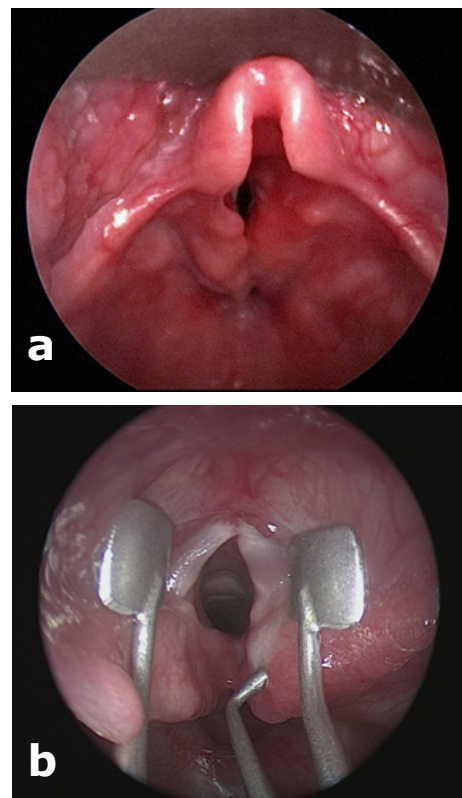


Fig.1: a) Direct laryngoscopy showing features of laryngomalacia, b) The laryngeal cleft was obscured by redundant mucosa prolapsing into the cleft, probe is shown demonstrating the site of laryngeal cleft and the arytenoids have been parted to show the cleft.

cation by Stimulated Emission of Radiation) treatment to her inter-arytenoid area and redundant mucosa overlying the arytenoids. The mucosa was denuded prior to suturing. We used Safil polysorb 5/0 anteriorly and 6/0 posteriorly. She had pre- and post-operative intravenous steroid and antibiotics. She was transferred to the Paediatric Intensive Care Unit (PICU) post-operatively and nursed with oxygen mask and without a feeding tube. She was extubated two days after surgery. Unfortunately she developed respiratory distress two days later.

Intravenous antibiotics and steroid were commenced. Despite Continuous Pressure Airway Pressure (CPAP) followed by Bi-level Positive Airway Pressure (BIPAP) therapy, her oxygen saturation deteriorated. She was taken back to the operating theatre for direct laryngoscopy. Under direct vision, the anastomosis was seen to be intact, with a thin layer of overlying fibrin overlying. The arytenoids and epiglottis appeared swollen, and there were a lot of secretions. She was taken back to PICU where she remained intubated and sedated for five days with regular tracheal suctioning. She was successfully extubated and bottle-fed. Her condition continued to improve on twice daily chest physiotherapy and she was discharged home.

A repeat direct laryngoscopy performed three months (Figure 3) later showed a well healed anastomotic site. A leak test was performed, whereby a 6F feeding tube was inserted, and methylene blue dye injected into the feeding tube in the oesophagus. No dye was seen to leak into the larynx or lower airway.

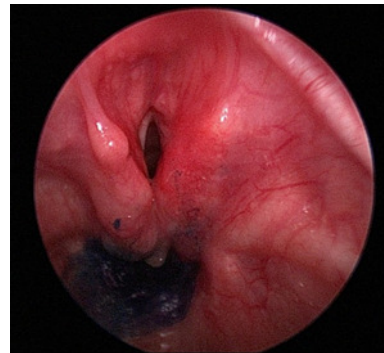


Fig. 3: Methylene blue (blue-black area below the vocal cord complex) in hypopharynx with no leakage into glottis and no persistent fistula. Anastomosis was seen to be intact.

DISCUSSION

Laryngeal cleft is a rare congenital abnormality. It affects more boys than girls with a ratio of 5:3.^{9, 10, 14} It can present with a myriad of symptoms; from choking during feeds and inspiratory stridor to aspiration pneumonia and failure to thrive. In this patient, these symptoms were initially attributed to laryngomalacia. However, as flexible laryngoscopy showed presence of the laryngeal cleft, endoscopic evaluation of the laryngotrachea was undertaken. The correct diagnosis was then made and the appropriate surgical measures performed.

The symptoms often depend on the length of the cleft. Some authors⁷ advocate endoscopic repair for Type 1 and 2 clefts, whilst others¹⁵ have reported a series of successful endoscopic repair on all types of laryngeal cleft. Repair of the cleft endoscopically without intubation provides many advantages¹⁵ over the open approach. It avoids an anterior neck incision over the larynx/ trachea, thus the risk of neck infection/ pharyngeal fistula and injury to the recurrent laryngeal nerve is zero. Secondly, the surgeon has an

excellent axial view over the airway. The endoscopic approach also affords easier exclusion of excess trimmed mucosa into the newly reconstructed airway which can then lead to laryngotracheal stenosis. Finally, this approach preserves the vascular integrity of the mucosa which is used for repair of the cleft itself.

Up to 50% of clefts may have associated congenital abnormalities.⁸ Three syndromes must be ruled out prior to any intervention. These are Opitz-Frias, Pallister-Hall and VACTERL (abnormalities of the vertebrae, anus, cardiac, tracheoesophageal fistula, renal and limb defects) associations. Opitz-Frias syndrome is characterised by cleft lip and palate, laryngeal cleft, cardiac anomalies, hypospadias or defects of the corpus callosum. Pallister Hall syndrome is characterised by hypothalamic hamarblastoma, hypopituitarism, imperforate anus, polydactyly and laryngeal abnormalities which can include laryngeal cleft. The only similarity between our patient and these other syndromes is laryngeal cleft. However, a thorough neonatal/ birth history must be taken, as well as a full systemic examination to rule out possible associated congenital malformations.¹²

Our patient had no persistent or recurrent cleft as proven by diagnostic direct laryngoscopy at three months post-op. She continues to be seen under the Speech and Language Therapist's follow-up as well as our Paediatric Airway clinic at our centre. She has improved clinically and has no further aspiration, pneumonia, chronic cough, stridor or cyanotic episodes. We plan to discharge her from hospital follow-up after 1 year.

In conclusion, although laryngomala-

cia remains a common finding in an infant with stridor, coexisting abnormalities must be borne in mind should the patient's condition continue to deteriorate. Intra-operative endoscopic examination remains the gold standard diagnostic modality for assessing laryngeal clefts.¹³ To our knowledge, this is the first case report where an infant presents with three contemporaneous pathologies; laryngomalacia, unilateral vocal cord palsy and laryngeal cleft, successfully treated using the endoscopic approach without intubation. We hope this case illustrates the need for a high level index of suspicion of concurrent in a deteriorating infant initially diagnosed with laryngomalacia.

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