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SHORT COMMUNICATION

Primary endoscopic repair of intermediate laryngeal clefts

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Abstract

Introduction: Traditionally, small laryngeal clefts may be closed endoscopically, while larger clefts necessitate an open anterior approach. We report the presentation, evaluation and outcome following endoscopic surgical repair of a series of laryngeal clefts.

Method: Retrospective study of children treated in a tertiary referral centre between 2003 and 2008. The presenting symptoms, patient demographics, cleft type, surgical outcome and complications were evaluated.

Results: Seven children underwent primary endoscopic repair of their laryngeal clefts (four Benjamin-Inglis type III clefts and three type II clefts). Presenting symptoms included stridor, cough and cyanosis with feeds, swallowing problems, weak cry, and recurrent lower respiratory tract infection. Treatment was ultimately successful in six of the seven children; treatment was ongoing for the remaining child, who underwent subsequent revision surgery via an open approach. Two children went on to require a second endoscopic repair, and two underwent an open repair of a residual defect. One child required a tracheostomy for failed extubation in the post-operative period.

Conclusion: Endoscopic repair is a safe, useful technique in the management of laryngeal clefts. Laryngeal clefts must be excluded in a child presenting with persistent aerodigestive tract symptoms, as described here.

Key words: Larynx; Congenital Abnormalities; Surgery; Endoscopy

Introduction

Laryngeal clefts, first described by Richter in 1792, are rare congenital anomalies that occur due to failure of development of the posterior lamina of the cricoid or the septum between the trachea and oesophagus.[1] The overall incidence is one in 10 000 to 20 000 live births, although mild forms are generally believed to be more common.[2,3]

Suspension laryngoscopy and palpation of the interarytenoid area remains the 'gold standard' for the diagnosis and assessment of the severity of a laryngeal cleft. The most widely used system for the classification of laryngeal clefts is that of Benjamin and Inglis, which describes four cleft grades of increasing severity.[4]

A type I cleft is a purely supraglottic defect extending up to but not involving the cricoid cartilage, and often responds to conservative treatment. The endoscopic approach for the repair of type I clefts that are persistently symptomatic has been widely reported.[5–10]

A type IV cleft is a severe defect extending into the thorax, requiring a multidisciplinary treatment approach. It has a high mortality rate and will not be discussed further here.

There is no consensus on the optimal management of type II clefts, which extend partially into the cricoid cartilage, or type III clefts, which extend completely through the cricoid cartilage and into the cervical tracheoesophageal wall but not into the thorax. The open approach, either anterior or lateral, is usually advocated for type II and III cleft repair.

Here, we describe an endoscopic approach for the repair of type II and type III laryngeal clefts.

Materials and methods

We retrospectively reviewed the case notes of seven children who had undergone primary endoscopic repair of type II ($n = 3$) or type III ($n = 4$) laryngeal clefts between 2003 and 2008. Two of our patients had clefts in association with Opitz–Frias syndrome, one type II and one type III. Only one child had no other congenital abnormalities. Table I shows the range of presenting symptoms in our patient group. It can clearly be seen that the children with type II clefts presented with milder symptoms than those with type III clefts, who naturally therefore came to surgery at a younger average age (Table I).

Surgical technique

Surgical repair was performed under general inhalational anaesthesia using a nasopharyngeal tube and spontaneous ventilation, providing an optimal view for suspension laryngoscopy.

Initially, the cleft was probed to determine its extent, and the apex of the cleft viewed using Lindholm vocal fold retractors (Karl Storz, Slough, UK) (Figure 1).

The edge and apex of the cleft were then denuded using a carbon dioxide laser set at 5 W in intermittent mode (Figure 2). The edge of the cleft was further freshened

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TABLE I
PATIENTS' PRESENTING SYMPTOMS AND AGES AT REPAIR, BY CLEFT TYPE

Presenting symptom(s)	Pt age at repair (mth)*†
Type II cleft	
Choke with feeds, weak cry	12
Recurrent chest infections	15
Cough with feeds	51
Type III cleft	
Stridor	1
Cyanotic episodes	2
Cyanotic episodes, aspiration pneumonia	4
Stridor	21‡

*Mean age at first repair = 15 months, median age = 12 months. †First surgical repair. ‡Patient (pt) referred from abroad.

using microscissors, which were also used to open the apex of the cleft. The two freshened layers produced in this way facilitated layered closure.

The defect was then sutured with 6/0 PDS (polydioxanone) sutures (Ethicon, Livingston, UK) in two layers whenever possible, starting distally, with the knots tied on the oesophageal side (Figure 3). We preferred not to use an interposition graft, and we avoided tracheostomy whenever possible.

Post-operatively, all children were closely monitored on the paediatric intensive care unit, with tracheal intubation and insertion of a nasogastric tube avoided whenever possible. Parenteral steroids were given for 24 hours and nebulised adrenaline as required. Children were initially maintained on intravenous fluids, prior to commencement of oral fluids the day after surgery. All children received anti-reflux treatment. We did not routinely perform contrast or video swallow tests in the post-operative period; rather, we preferred to make use of these tests when required, depending on each individual child's progress.

Results

All the children with type II clefts were successfully treated with single-stage endoscopic repair, without any significant complications. Those with type III clefts had a much more variable course, summarised in table I.

Child one, who had undergone repair of a tracheoesophageal fistula on her second day of life, and subsequent Nissen fundoplication with gastrostomy, presented with coughing and cyanotic episodes with feeds plus an episode of aspiration pneumonia. At six months of age, an endoscopic repair of a cleft extending to the first tracheal ring was performed, and the majority of symptoms resolved. Subsequent endoscopy revealed a small, persistent, type I defect, which was again repaired endoscopically with good results. Child one continued to feed well and her gastrostomy was removed.

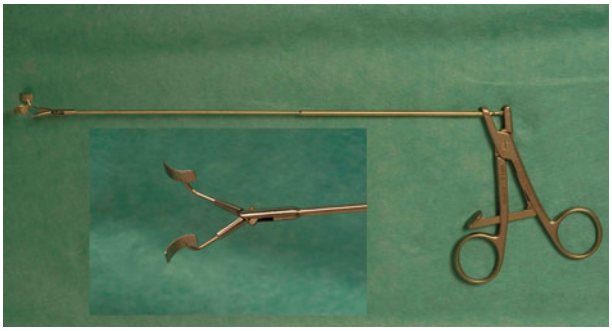

FIG. 1
Lindholm vocal fold retractors, used to visualise the apex of the cleft.

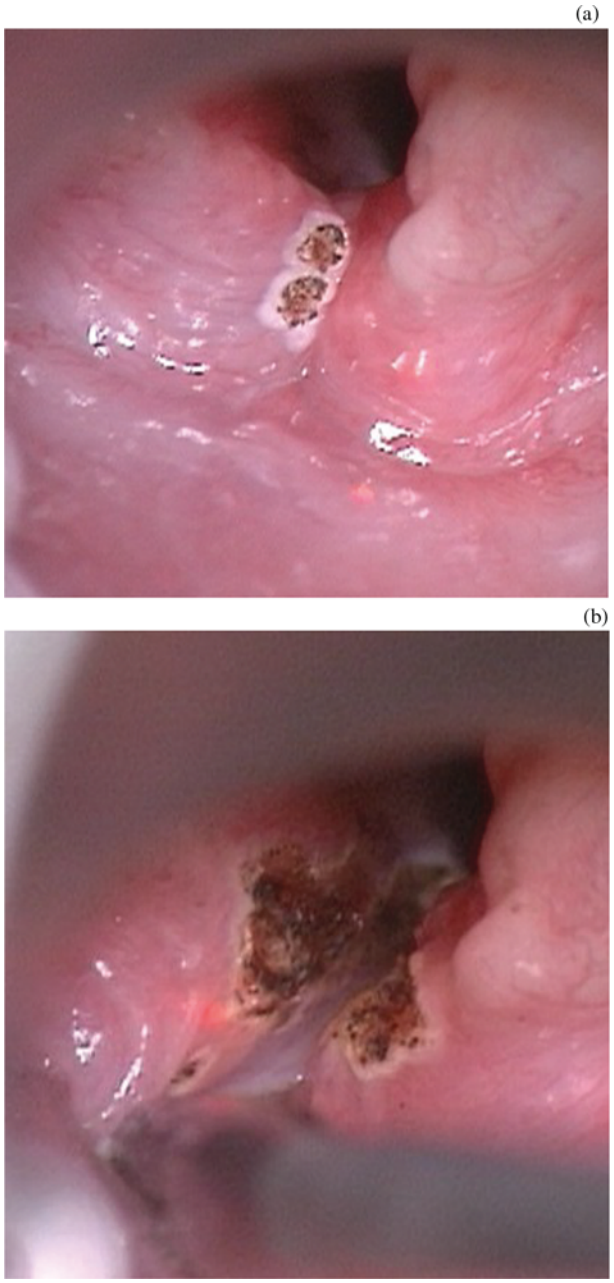

FIG. 2
(a) Edge and (b) apex of the cleft, denuded using a carbon dioxide laser.

Child two presented at nine months of age with stridor from birth, and in the absence of other symptoms was clinically diagnosed with laryngomalacia. Continued stridor and developmental delay prompted a further referral to our service at the age of 21 months. At this time, direct laryngoscopy confirmed a type III cleft, a prominent cricoid anteriorly, and evidence of left bronchial narrowing. Further investigation confirmed a diagnosis of Opitz–Frias

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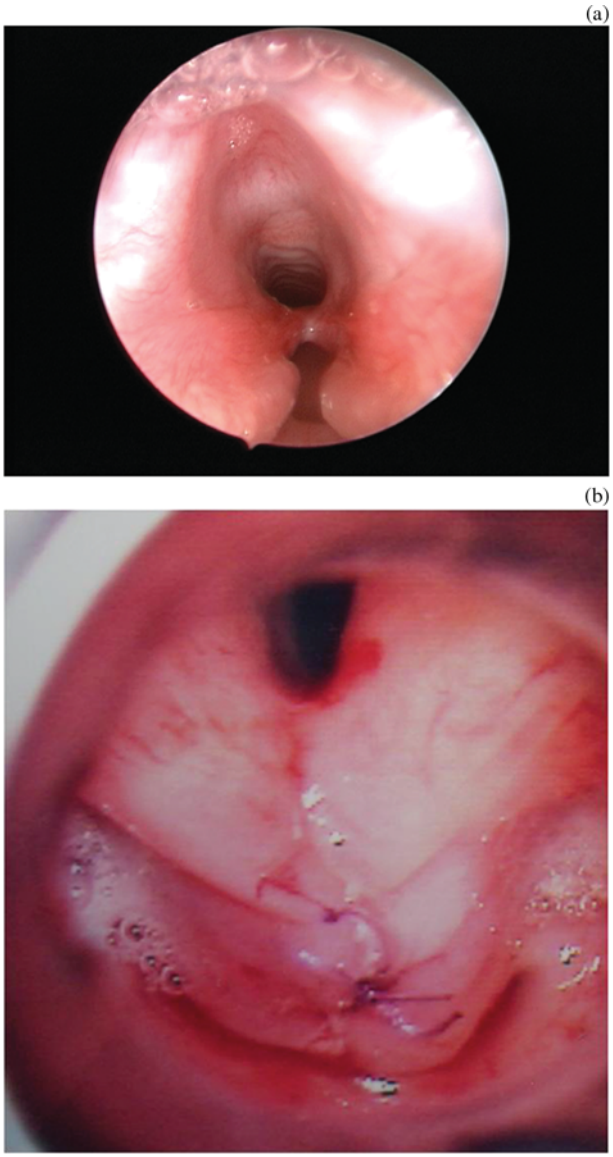

FIG. 3
Type III laryngeal cleft (a) before and (b) after suture repair.

syndrome, and demonstrated a left-sided superior vena cava that did not require surgical intervention. Endoscopic repair of the laryngeal cleft was performed at 25 months of age. Considering the child's other laryngeal and chest abnormalities, it was decided to place a covering tracheostomy, which remained in place for two weeks. The child progressed well, but still required some thickener in his feeds. Nine months after the initial repair, videofluoroscopy showed some minor aspiration of liquids. Subsequently, direct laryngoscopy showed a small, persistent, type I defect. This was again repaired endoscopically, following which there were no persistent symptoms.

Child three had no congenital abnormalities and presented with cyanotic episodes during feeds. Her cleft extended to the lower border of the vocal folds. The initial repair was performed endoscopically at one month of age. Initially, feeding improved dramatically, but subsequently deteriorated, and repeated endoscopy five weeks after the initial surgery revealed a breakdown of the repair at its apex. This was repaired via an anterior laryngofissure, with a covering tracheostomy remaining in place for three weeks. Following decannulation, this child made an excellent recovery.

Child four presented with microcephaly, developmental delay, and stridor following birth at 33 weeks' gestation. This child's laryngeal cleft extended to just below the vocal folds, and was repaired endoscopically at two months of age. Attempts at extubation on post-operative days 6 and 10 were unsuccessful, so a tracheostomy was created to enable weaning from ventilatory support. Shortly after this, a percutaneous gastrostomy was inserted. Unfortunately, the initial endoscopic repair failed to heal, and so at five months of age the cleft was repaired via an external approach (anterior laryngofissure). At seven months of age, direct laryngoscopy revealed a small, persistent, type I defect, which was repaired endoscopically. Despite making a good recovery, with successful cleft repair, this child died four months later as a result of his significant comorbidity.

Discussion

We describe our experience of the endoscopic repair of laryngeal clefts, highlighting the difficulties that can be encountered when treating type III defects. Two of our four children with type III clefts were successfully managed endoscopically, although one required a temporary covering tracheostomy and both required a second endoscopic procedure to correct small, residual, type I defects. The other two children failed endoscopic repair and required an open approach with tracheostomy.

The endoscopic approach for the repair of laryngeal clefts has several potential advantages over open approaches. Endoscopic repair avoids the risks of the anterior and lateral open approaches, including wound complications, laryngeal destabilisation and subsequent dysphonia, and injury to the recurrent laryngeal nerve. In addition, endoscopic repair may enable avoidance of tracheal intubation and tracheostomy, procedures which can be detrimental to healing of the repair due to a direct pressure effect or introduction of infection.

Since the endoscopic approach was first described by Koltai *et al.* in 1991, experience with endoscopic techniques has increased, and this method of repair is now commonly used for type I clefts that fail conservative treatment.[5–10]

However, use of the endoscopic approach for type II and type III clefts remains controversial, and there are no large studies demonstrating its efficacy in the long term.

Kubba *et al.* have recommended the endoscopic approach for type I and shorter type II clefts.[6] The anterior open approach, with covering gastrostomy and tracheostomy, is preferred for longer clefts; follow up of 10 patients with type III clefts treated in this way identified one child who died (cause unknown), five who experienced complications and three who required revision procedures. This highlights the difficulties encountered in treating intermediate laryngeal clefts in a group of children with a range of comorbidities, whatever the approach.

Rahbar *et al.* have advocated the endoscopic approach for type I and all type II clefts, and describe its successful use in a single case of type III cleft.[9]

Sandu and Monnier have reported the successful use of endoscopic repair in four children with type III clefts, without complication.[11] Their protocol for an endoscopic approach in type III clefts excludes children with associated

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laryngeal abnormalities, as well as those with Opitz–Frias or Pallister–Hall syndromes. They describe a mucosal incision with a laser, and subsequent two-layer closure, in three cases with complete division of the cricoid plate and one with extension of the cleft to the level of the sternal notch. All four children in their series achieved normal feeding with no clinical signs of aspiration, and all had a good voice. All had slight exertional dyspnoea; the authors hypothesise that this was due to a laterally situated posterior cricoarytenoid muscle, associated with the cleft condition, which prevented complete abduction of the vocal folds.

It is interesting to note that three of our four patients with type III cleft required a second endoscopic repair. We do not consider this need for further surgery, in itself, to represent failure of the endoscopic technique, as we tended to err on the side of under-treatment during the initial repair. This was because overclosure of a laryngeal cleft can lead to supraglottic stenosis, which can be very difficult to treat.

The current paper represents the second reported series of endoscopic repair of type III laryngeal clefts; two of our four patients with type III clefts were successfully treated in this manner.

Conclusion

This small series demonstrates the successful use of endoscopic repair for type II laryngeal clefts. Endoscopic repair of type III laryngeal clefts had a more variable outcome. We believe that endoscopic repair is a promising technique, but it is clear that each case must be treated individually by an experienced team who are able to offer the full range of surgical approaches.

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