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OTHER ARTICLES

Management of Symptomatic Grade I and II Laryngeal Cleft: Experience of a Tertiary Care Center and Review of Literature

Anindya Nayak[1] · Preetam Chappity[1] · Sidharth Pradhan[1] · Pradeep Pradhan[1] · Pradipta Parida[1] · Karakkandy Vinusree[1]

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Abstract Laryngeal cleft is a rare pathology and needs a high index of suspicion for diagnosis. Various classifications are used but the Inglis classification which describes 4 grades of cleft is most widely accepted. Grade 3 and 4 clefts are very rare and are usually associated with other congenital abnormalities. Grade 1 and 2 clefts are more common and can be easily corrected with good outcomes. We are presenting our experience with 9 cases of low grade (Grade 1 and 2) laryngeal cleft which were managed successfully in our department. Out of the 9 cases 4 failed conservative management and required a trans-oral repair. All patients improved with management, except one who intermittently developed another episode of pneumonia 2 months' post-surgery. However satisfactory cleft obliteration was confirmed on endoscopy and the patient has been under close follow up ever since. Majority of low grade laryngeal clefts can be managed with dietary modifications and feeding rehabilitation. Those who do not respond usually require surgical repair with near complete resolution of symptoms.

Keywords Laryngeal · Cleft · Conservative · Endoscopy

✉ Preetam Chappity
preetam82@gmail.com

[1] Department of ENT, All India Institute of Medical Sciences Bhubaneswar, Room No 175, Sijua, Patrapada, Bhubaneswar, Odisha, India

Introduction

Laryngeal clefts are rare congenital anomalies of the aerodigestive tract because of a deficient anatomical separation of the oesophagus and respiratory system at the level of the larynx and trachea. Incidence is reported as 1 in 10,000 to 1 in 20,000 live births, more common in boys than girls with a ratio of 5:3 [1, 2]. Depending on the extent of the cleft in the larynx and trachea, life-threatening disturbances of respiration can occur immediately after birth.

The condition was first described by Richter [3] in a new-born who presented with aspiration. It was Petterson [4] who attempted the first surgical reconstruction and also proposed a classification in 1955. Over the years, multiple classifications have been described with the one described by Benjamin and Inglis [5] being most widely used today.

Numerous theories [6] that have been proposed to explain the development of tracheoesophageal anomalies. They can be divided into due to intraembryonic pressure by heart and the curvature of esophagus, epithelial occlusion defect in esophagus, vascular occlusion due to persistent aberrant vessel causing low perfusion of the gut and differential cell growth.

Embryologically, a posterior laryngeal cleft is due to the incomplete formation of the interarytenoid membrane which may also include the absence of interarytenid muscle [7]. The classification proposed by Benjamin and Inglis [5] describes 4 types:

- Type 1—supraglottic interarytenoid defect, in which the cleft lies above the level of the posterior cricoid cartilage
- Type 2—cricoid lamina is partially involved with extension below the level of the true vocal cords;
- Type 3—total cricoid cleft, and

- Type 4—extending into the posterior wall of the thoracic trachea and may extend as far as the carina.

The signs and symptoms of patient with laryngeal cleft anomaly varies with the type of defect. It may be as subtle as mild aspiration to major life threating symptoms like stridor and respiratory distress.

In this manuscript we present a retrospective review of suspected laryngeal cleft cases that were managed at our centre. Institutional ethical clearance was waived off due to the retrospective nature of the study and non-deviation from accepted standard of care. As the study had no new intervention planned and was retrospective it was waived from CTRI registration.

In the present series we present a total of nine cases of suspected laryngeal cleft defect who were evaluated and were managed either conservatively or underwent surgical intervention at our tertiary care centre.

Case Series

Patients were referred from the department of Paediatrics for symptoms that included failure to thrive, difficulty in feeding, recurrent pneumonia and coughing while feeding which led to the suspicion of aspiration. In our series two symptoms were common to all the subjects, failure to thrive and recurrent pneumonia (Range 3–7 episodes over 6 months' period).

Patients were reviewed by our Paediatric department for any syndromic association. We performed fibre optic laryngoscopy with swallowing evaluation in all the patients, either trans nasally or trans orally in our minor operative room with Paediatric back up. We avoided radiology to avoid radiation exposure. In 1 case where child had a cardiac anomaly associated (Atrial septal defect) a non-ionic contrast based study was done to rule out a tracheoesophageal fistula.

A routine diagnostic direct laryngoscopy was not done unless the conservative measures failed to improve the symptoms. Patients who improved with minor dietary and positional modifications in likely grade 1 cleft were excluded from the study. These conservative measures included changes in feeding habits such as the thickening the consistency and viscosity of feed, feeding strategies and positioning. Treatment of coexisting gastroesophageal reflux disease in the forms of PPIs was also undertaken. Other underlying causes such as reactive airway disease, food allergies and TEF (Tracheoesophageal fistula) were also ruled out.

Our protocol was to initiate conservative measures immediately. Those who improved by 1–2 months were not considered symptomatic for the cleft. Those whose symptoms persisted beyond 2 months of conservative management were considered to be highly suspicious of a symptomatic cleft and were admitted for swallowing therapy and parent education and included in our study. The patients were closely monitored and either significant worsening or non-improvement of symptoms were considered for surgical intervention.

Of the nine cases only four required surgical intervention. Diagnostic laryngoscopy was done and the diagnosis was confirmed with palpation of the cleft (Figs. 1, 2). A diagnostic bronchoscopy was also performed in the same setting to rule out any other pathology in the airway.

A trans oral approach was utilized. The margins of the cleft were made raw with coblation MLW wand (Fig. 3) in 3 cases and using Co2 laser in 1 case. Tubeless anaesthesia with spontaneous breathing was continued all through the surgical procedure. The repair was done under an operating microscope with an endoscopic needle holder and knot pusher (Fig. 4). A blunt right angled probe was used to palpate the depth of the interarytenoid groove with care taken not to manipulate the interarytenoid mucosa. The cricoid was palpated for evidence of dehiscence. A 6-0 resorbable vicryl suture was used for closure. Two independent sutures were applied for stability and strength from distal to proximal direction. The endoscopic picture after repair is shown in Fig. 5.

The patients were managed in Paediatric ICU for 24 h and a repeat fibre optic endoscopy was performed on post-operative day 1 to rule out overclosure which would lead to restriction of vocal cord mobility resulting in stridor.

Results

The patients were in the age group of 3–38 months. Only one child presented late at the age of 3 years and 2 months as they had no tertiary care services in their rural set up. The rest of the patients were all under 1 year of age (3 months, 7 months, 8 months and 11 months). The weight of the child at presentation was 10 kg and required

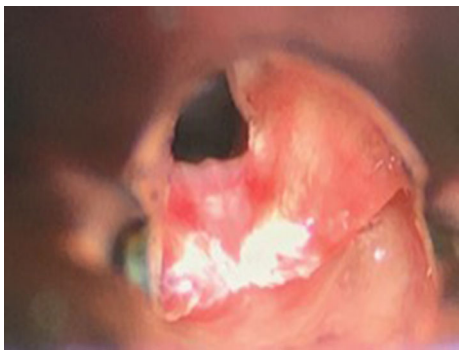

Fig. 1 Microscopic view of a type 2 laryngeal cleft

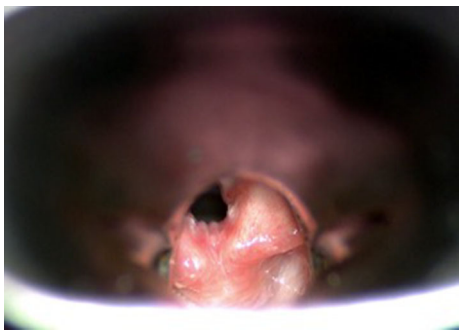

Fig. 2 Microscopic view of a type 1 laryngeal cleft

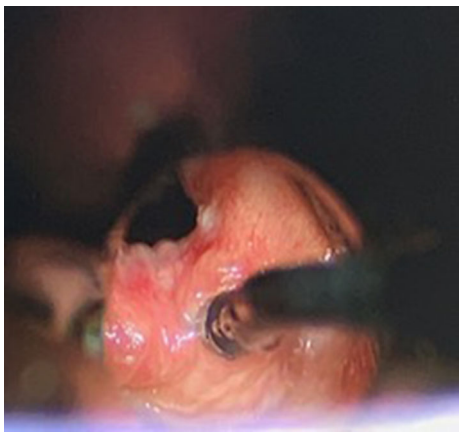

Fig. 3 Microscopic view of margins being freshened with MLW coblation wand

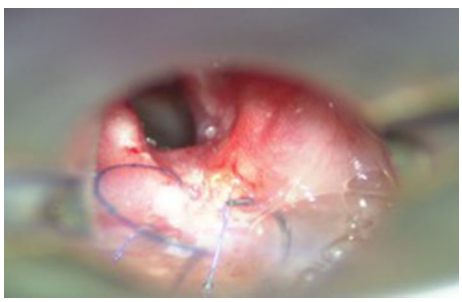

Fig. 4 Suturing of freshened margins with 6-0 vicryl

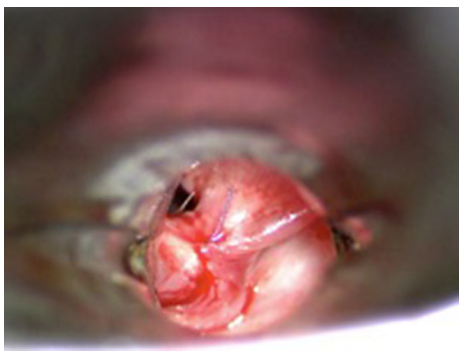

Fig. 5 Microscopic view of laryngeal cleft post repair

prompt surgical intervention in 2 months' time, due to non-improvement of symptoms. The child gained weight to 15 kg at 8 months follow up post-surgery with no further episodes of pneumonia.

Amongst all the symptoms, all the 9 cases had history of failure to thrive and recurrent pneumonia without any definitive pulmonary signs. The frequency of pneumonia varied from 1 to 3 episodes per month. Seven cases had coughing while suckling or feeding.

Five of the total number of cases were found to have a deep interarytenoid groove that was suggestive of a type 1 laryngeal cleft. All such cases underwent conservative management and dietary modifications. The minimum follow-up period has been 5 months. The weight gain has been adequate with no further pulmonary complaints.

Four of the remaining cases had type 2 laryngeal clefts. They required surgical intervention as there was no improvement with conservative management.

Of the 4 cases 3 had weight gain and no further episodes of pneumonia post-surgery. One case developed a repeat bout of pneumonia after 2 months of surgery even though the repeat endoscopy showed closure of the cleft, with no evidence of micro-aspiration. He is otherwise asymptomatic with regards to aspiration. He is being kept on regular follow up for evaluation of any immunodeficiency.

The minimum follow-up for all cases in our study was 5 months (range 5–48 months'). The intervention was considered successful if there was an upward trend noticed in the age appropriate weight gain of the baby over 3 months' period of time, with no further episodes of pneumonia. Repeat endoscopy was performed at monthly intervals to rule out any evidence of micro aspiration.

Discussion

Laryngeal cleft is a rare anomaly with potentially grave outcomes. Due to its wide spectrum of presentation, diagnosis is often difficult and requires a multi-modality approach. It is important to have a degree of suspicion in any child who presents with recurrent chest infections, aspiration during feeding and generalised failure to thrive.

The differential diagnosis includes swallowing disorders due to neuromuscular causes, CNS causes like hydrocephalus, Arnold Chiari malformation. Other causes like a high vagal palsy leading to vocal cord palsy should also be kept in mind. As many as 50% of patients with laryngeal cleft have associated congenital abnormalities like tracheo-esophageal fistula (TEF), tracheomalacia, cleft lip and palate, anomalous right subclavian artery, pyloric atresia, imperforate anus, pancreatic ectopia, congenital heart defect, and congenital subglottic stenosis [8, 9]. In the present study, excepting 1 case who had an atrial septal

defect, rest did not have an associated congenial abnormality. This might be attributed to the low grade of clefts that we encountered (Grade 1 and 2).

All cases were evaluated in our series by routine radiological investigations (chest x-ray), cardiac asessment and a flexible laryngoscopic evaluation with swallowing assessment. Most authors prefer a preliminary modified barium swallow or a FEES prior to endoscopy. However, it is widely accepted that intraoperative palpation remains the gold standard of diagnosis [10].

All cases of type 1 laryngeal cleft were managed by lifestyle changes with 100% success in our series. This is in agreement with other studies reported [7, 8, 11, 12]. Surgical intervention was done in cases of failure of conservative management. We did not encounter higher grade of cleft as usually these cases are also associated with other congenital abnormalities and have grave prognosis.

A number of techniques have been described for repair. Waltzman and Bent [13] described anteriorly and posteriorly based mucosal flaps over opposite arytenoid with an S-shaped incision across the interarytenoid cleft. The flaps were rotated so that each covered the demucosalized portion of the contralateral arytenoid, creating a double mucosal layer in the interarytenoid region. Chien and colleagues inserted an age-appropriate endotracheal tube to be used as a laryngeal stent for up to 10 days [11]. Garabealdian reconstructed several type III clefts that extended to the first two tracheal rings in infants aged 10–15 days using a bi positive airway pressure during wake up phase [14]. Nakahara et al. [15] described the use of collagen injections in a type I cleft, with good results at 9 months follow-up. Kennedy et al. [16] reported Gelfoam injection in cleft type I with favourable results. Ahluwalia et al. [17] suggested the use of Bioplastic injections as an alternative to gelfoam.

The overall low mortality and morbidity associated with endoscopic repair of lower grade laryngeal clefts have made it the procedure of choice. Higher grade clefts may require open surgery. A cervical or cervicothoracic approach is used. Three types of cervical approaches have been described [14].

(1) The lateral approach with lateral pharyngotomy,
(2) The lateral approach with posterior pharyngotomy, and
(3) The anterior translaryngotracheal approach.

In spite of major advances in medical and surgical management, the prognosis of laryngeal cleft ultimately depends on the type of cleft. Roth et al. [1] found an overall mortality rate of 46 with all types combined (43%for types I and II, 42% for type III, and 93% for type IV) [1]. The presence of other coexisting anomalies or syndromes worsen the prognosis. Myer et al. [18] in their study found that more than 60% of deaths were in patients with other anomalies.

A multimodality approach and early diagnosis and treatment are therefore important in managing this condition.

The high success rate reported in our series can be attributed to the fact that most cases were of low grade clefts. The surgical outcome reported by us is comparable to literature.

Conclusion

A high index of suspicion is required to diagnose cases of laryngeal cleft. High grade of cleft is usually associated with other congenital abnormalities and carry a grave prognosis requiring open and more morbid procedures. The commonest mode of presentation is failure to thrive and recurrent pneumonia. A fibre optic laryngoscopy with swallowing evaluation is adequate to suspect a cleft after ruling out other possible differential causes. A routine radiology or diagnostic endoscopy under general anaesthesia is not necessary and not feasible in high volume centres. A trial of 3 months of conservative management can be tried and if no improvement is noted surgical intervention is required. Trans oral microscopic or endoscopic single layer repair with at least 2 sutures is required for successful repair. If the patient has severe symptoms, then an early intervention should be planned. A single layer closure with tubeless anaesthesia is preferable to produce the best outcomes. The conservative management should continue into the post-operative period to prevent wound breakdown before complete healing.

Summary

- Laryngeal cleft is a rare pathology and needs a high index of suspicion for diagnosis.
- Grade 1 and 2 clefts are more common and can be easily corrected with good outcomes with conservative measures and endoscopic surgery.
- In the present series we present a total of 9 cases of suspected laryngeal cleft defect.
- Of the 9 cases only 4 required surgical intervention. This was done transorally.
- The remaining cases were successfully managed conservatively.
- All cases who underwent surgical intervention had a successful cleft repair.
- We recommend a trans oral microscopic or endoscopic single layer repair with at least 2 sutures for successful

repair. A single layer closure with tubeless anaesthesia is preferable to produce the best outcomes.
- The conservative management should continue into the post-operative period to prevent wound breakdown before complete healing.

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Compliance with Ethical Standards

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References

1. Roth B, Rose KG, Benz-Bohm G, Gunther H (1983) Laryngotracheoesophageal cleft: clinical features, diagnosis and therapy. Eur J Pediatr 140:41–46
2. Eriksen C, Zwillenberg D, Robinson N (1990) Diagnosis and management of cleft larynx. Literature review and case report. Ann Otol Rhinol Laryngol 99:703–708
3. Richter CF (1792) Dissertatio Medico de infanticide in artis obstetriciae. Thesis for Doctor of Medicine, Leipzig
4. Pettersson G (1955) 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 110:250–254
5. Benjamin B, Inglis A (1989) Minor congenital laryngeal clefts: diagnosis and classification. Ann Otol Rhinol Laryngol 98:417–420
6. Merei JM, Hutson JM (2002) Embryogenesis of tracheoesophageal anomalies: a review. Pediatr Surg Int 18(5–6):319–326
7. Johnston RD, Watters K, Ferrari LR, Rahbar R (2014) Laryngeal cleft: evaluation and management. Int J Pediatr Otorhinolaryngol 78:905–911
8. Rahbar R, Rouillon I, Roger G et al (2006) The presentation and management of laryngeal cleft: a 10 year experience. Arch Otolaryngol Head Neck Surg 132:1335–1341
9. Evans KL, Courteney-Harris R, Bailey CM et al (1995) Management of posterior laryngeal and laryngotracheoesophageal clefts. Arch Otolaryngol Head Neck Surg 121:1380–1385
10. Cohen SR (1975) Cleft larynx. A report of seven cases. Ann Otol Rhinol Laryngol 84:747–756
11. Bowe SN, Hartnick CJ (2017) Management of type 1 and type 2 laryngeal clefts: controversies and evidence. Curr Opin Otolaryngol Head Neck Surg 25:000–000
12. Chien W, Ashland J, Haver K, Hardy SC, Curren P, Hartnick CJ (2006) Type I laryngeal cleft: establishing a functional diagnostic and management algorithm. Int J Pediatr Otorhinolaryngol 70(12):2073–2079
13. Waltzman M, Bent J III (2001) Repair of type 1-A and 1-B posterior laryngeal clefts. Oper Tech Otolaryngol Head Neck Surg 12(4):252–255
14. Pezzettigotta SM, Leboulanger N, Roger G, Denoyelle F, Garabealdian E (2008) Laryngeal cleft. Otolaryngol Clin N Am 41:913–933
15. Nakahara S, Tayama N, Tsuchida Y (1995) Aminor laryngeal cleft (type 1-a) diagnosed in infancy. Int J Pediatr Otorhinolaryngol 32(2):187–191
16. Kennedy CA, Heimbach M, Rimell FL (2000) Diagnosis and determination of the clinical significance of type 1A laryngeal clefts by gelfoam injection. Ann Otol Rhinol Laryngol 109(11):991–995
17. Ahluwalia S, Pothier D, Philpott J et al (2004) Laryngeal cleft type I: a novel method of repair using bioplastique. J Laryngol Otol 118(8):648–650
18. Myer CM 3rd, Cotton RT, Holmes DK et al (1990) Laryngeal and laryngotracheoesophageal clefts: role of early surgical repair. Ann Otol Rhinol Laryngol 99(2 Pt 1):98–104