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Laryngeal Cleft: Evaluation and Management

Heather Nardone and Reza Rahbar

Laryngeal cleft is an uncommon congenital anomaly, with an incidence of approximately 1 in 10, 000 to 20, 000 live births. The developmental defect is the result of an abnormal communication between the posterior laryngotrachea and the esophagus. During the 5th-7th weeks of development, the cricoid cartilage closes from two lateral centers and the tracheoesophageal folds fuse, forming a complete cricoid ring and separating the airway from the esophagus. Arrest at various stages in this process results in laryngeal clefts of varying degree. Histolopathologic evaluation of the cleft larynx by Lim (1979) revealed cartilaginous defects in the posterior lamina of the cricoid as well as arrested foci of developing cricoid. Partial agenesis and/or atrophy of the interarytenoid muscle was also observed.

While the majority of cases appear to be due to a sporadic event, laryngeal cleft is also a part of two genetic syndromes, Opitz-Frias and Pallister-Hall syndromes (Phelan 1973). Both inherited in an autosomal dominant fashion, Opitz-Frias consists of cleft lip and palate, hypertelorism, hypospadias, and cleft larynx, while Pallister-Hall is characterized by hypopituitarism, hypothalamic hamarblastoma, imperforate anus, postaxial polydactyly, and laryngeal cleft.

Though several different classification systems have been described, the classification system proposed by Benjamin and Inglis (1989) remains the most widely used. In this system, four types of clefts are described. Type 1 cleft is an interarytenoid defect, above the level of the vocal cord. Type 2 cleft extends to the level of the vocal cords and partially involves the cricoid lamina. In type 3 clefts, the entire cricoid is involved, with possible extension to the cervical trachea. Type 4 clefts extend into the thoracic trachea and may extend to the carina. The severity of symptoms is typically proportional to the extent of the cleft and ranges from hoarseness and recurrent respiratory tract infections to severe aspiration pneumonia leading to cardiopulmonary collapse.

Infants and children with laryngeal cleft present with somewhat non-specific symptoms, including chronic cough, recurrent aspiration, stridor, and/or cyanosis with feeds. The differential diagnosis of such symptoms is broad and includes esophageal stricture, tracheoesophageal fistula, cricopharyngeal spasm, neuromuscular abnormalities, laryngomalacia, GER, vocal cord paralysis, and bilateral choanal atresia. Pediatricians and pediatric specialists caring for children

with these symptoms must have a high index of suspicion for laryngeal cleft, as early detection and management help to lessen morbidity and mortality.

Work-up for children with these respiratory and/or feeding difficulties may include chest x-ray, modified barium swallow (MBS), and fiberoptic endoscopic evaluation of swallowing (FEES). Chest x-ray is useful as it may show pulmonary infiltrates from repeated aspiration. MBS and FEES are also valuable tools for evaluating aspiration. MBS allows examination of the dynamic swallowing process, showing the entire swallowing phase and thus allowing aspiration due to neuromuscular discoordination to be ruled out. While FEES also plays an important role in evaluating the swallow, it is an invasive procedure and is challenging to perform in infants and young children. In addition, FEES fails to evaluate the entire swallowing cycle and so does not allow one to rule out aspiration due to a lack of neuromuscular coordination. Finally, while the finding of aspiration on either study is a significant one, neither a negative MBS nor FEES rules out intermittent aspiration. These tests offer only a snapshot of a child's swallowing cycle and thus may miss aspiration if it occurs only intermittently.

Microdirectlaryngoscopy under general anesthesia is the gold standard for diagnosis of a laryngeal cleft. Because of a redundancy of tissue in the posterior glottic region and inward collapse of the posterior supraglottic structures with inspiration, a laryngeal cleft is sometimes difficult to appreciate on visual inspection, even during microdirectlaryngoscopy. Therefore separation and palpation of the arytenoids with a probe is essential in order to thoroughly evaluate the posterior glottis. If a cleft is diagnosed, the depth of the cleft in relation to the true vocal cords and cricoid should be noted.

Complete evaluation of both the tracheobronchial tree and esophagus should be performed in order to evaluate for associated anomalies, which may impact the timing and approach taken for repair. Concomitant abnormalities of the airway and esophagus are not infrequently described. Tracheobronchomalacia is one of the most common findings at the time of endoscopy, occurring in approximately one third of cleft children (Rahbar 2006). Its presence can cause continued airway difficulty despite successful cleft repair and may necessitate the use of CPAP or even tracheotomy until the child grows and the condition improves.

Tracheoesophageal fistula occurs in 19%-56% of children with laryngeal cleft (Lim 1979, Tyler 1985, Walner 1999). Given that the pathogenesis of both conditions involves an error in the separation of the embryonic respiratory and alimentary tracts, a common mechanism may be responsible for both disorders. It is important for physicians evaluating children with TEF to be aware of this association, so that they may provide their patients with the greatest likelihood of a functional swallow and pulmonary health, and so they may counsel families appropriately in relation to the expected success of the cleft repair. In a series by Walner (1999), patients with cleft larynx with a history of TEF repair had a much higher incidence of breakdown of their cleft repair (86%) than those without history of TEF repair (9%).

Other associated abnormalities, including cleft lip and palate, gastrointestinal, genitourinary tract, and cardiovascular anomalies are also described in children

with cleft larynx. The presence and severity of such anomalies often dictate when a patient is medically stable for repair.

Early diagnosis is crucial to avoid the long-term pulmonary sequela of repeated aspiration. Many advocate a trial of conservative management in those with a type 1 and type 2 clefts, with the use of antireflux medications, positioning maneuvers during feeds, and thickened food. Rahbar et al (2008) found that 48% of children with both type 1 and 2 clefts responded favorably to conservative therapy, avoiding surgical repair.

Endoscopic repair is typically reserved for children with type 1 and type 2 clefts who fail conservative management. Endoscopic repair is best approached by avoiding endotracheal intubation; an endotracheal tube hinders exposure and one's ability to suture. Rather, general anesthesia with spontaneous breathing should be used. Using this method, the larynx is exposed using a Lindholm laryngoscope, the mucosal margin of the cleft is denuded using a carbon dioxide laser, and interrupted absorbable sutures are placed to close the cleft.

For type 1 and type 2 clefts, endoscopic repair shows great success in terms of improved airway and swallowing function, with studies by Watters (2003) and Rahbar (2006) showing success rates ranging from 80-90%. More recently, the endoscopic approach has yielded promising results for select type 3 clefts as well (Sandu 2006).

Type 3 clefts are generally repaired via an open anterior approach. The anterior approach was first first described by Jahrsdoerfer in 1967. It entails performing a thyrotomy, cricoidotomy, and tracheofissure of the first and second rings to provide access to the posterior cleft and enable microsurgical repair. After gaining exposure of the posterior airway, the mucosal edges of the cleft are incised and a repair is completed in 2 layers. This approach offers excellent visualization and minimizes risk to neighboring neurovascular structures. Risks of open repair on laryngeal instability and long-term laryngeal growth have been raised but not substantiated.

More extensive clefts extending into the thoracic trachea necessitate a lateral phayngotomy with a right thoracotomy or an anterior approach with a median sternotomy to be used. The pharynx is entered by incising the inferior constrictor muscle above the level of the pyriform sinus. A two layer repair is then undertaken in a manner identical to that previously described. This approach offers more limited exposure and places the recurrent laryngeal nerve at great risk.

Complications are noted to occur in approximately 50% of open repairs and typically require revision. Most commonly, a dehiscence of sutures at the site of cleft repair occurs. This could be secondary to GER, coughing, and/or pressure caused by an endotracheal or nasogastric tube. Risk of failed repair has been found to increase with both the severity of the cleft and the presence of concomitant anomalies (Walner 1999). Other complications with open repair include laryngeal nerve injury, granulation tissue formation, esophageal stricture, and late development of TEF.

The use of interposition grafts has been advocated to decrease the risk of dehiscence. Costal cartilage, pleura, and sternocleidomastoid muscle have been

used (Prescott 1995). More recently, the use of tibial periosteum has been described (Garabedian 1998). The authors advocate the use of tibial periosteum due to its resistance to necrosis, osteogenic properties, and pliability.

Less invasive methods of cleft repair have recently been performed. These include the use of the injectable bulking agents gelfoam and bioplastique in order to build up the posterior glottic wall (Ahluwalia 2004). Long-term outcomes on greater numbers of patients are needed in order to flesh out the utility of these substances.

Several factors must be addressed in the perioperative period in order to increase the likelihood of a successful repair. Patients' pulmonary and nutritional status must be optimized. Avoidance of aspiration from gastroesophageal reflux is critical. Gastroesophageal reflux should be evaluated and treated with medical and/or surgical therapies. Medical therapy includes treatment with acid suppression medications. Surgical intervention may include gastrostomy and/or Nissen fundoplication. Nutritional status must be optimized through either enteral or parenteral feeding.

Morbidity and mortality rates remain significant despite evolution in technique and improved awareness leading to an earlier age of presentation. Postoperative complications are noted to occur approximately 50% of cases, with almost half of these patients requiring revision (Kubba 2005). Though mortality rates were as high as 40-50% in the 1980s (Roth), rates of 6-14% are still reported (Kubba 2005, Evans 1995). Deaths are more common with more severe clefts and are often due to complications resulting from concomitant congenital anomalies.

Continued improvement in the evaluation and management of children with laryngeal cleft demands that pediatric specialists caring for children in question have a heightened index of suspicion. Once diagnosed, further evaluation and management should occur with a multidisciplinary pediatric team, consisting of otolaryngology, gastroenterology, pulmonary, and speech and swallowing specialists familiar with treating cleft larynx children.

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