Objective: To determine the incidence and character of clinically significant laryngotracheal anomalies in pediatric patients undergoing surgical repair of congenital cardiac defects at a tertiary care center.

Design: Single-center retrospective review.

Patients: The charts of pediatric patients who required surgical treatment for congenital heart disease over a 4-year period were reviewed. Forty-eight of 1957 patients were seen in inpatient consultation by the otolaryngology service. The parameters studied included cardiac diagnosis, reason for consultation, findings on examination, and follow-up.

Results: There were 16 (33%) cases of subglottic stenosis, which were graded according to the Cotton-Myer classification system as follows: grade 1 (n=8); grade 2 (n=3); and grade 3 (n=5). Three of the 16 patients with subglottic stenosis required tracheotomy and 4 required laryngotracheal reconstruction. Nine (19%) of the 48 patients were diagnosed as having unilateral true vocal cord paralysis and 3 (6%) as having bilateral paralysis. With the exception of 1 patient, all patients with true vocal cord paralysis on the left side had undergone repair of the aortic arch.

Conclusions: Pediatric patients with congenital cardiac disease are predisposed to laryngeal anomalies owing to (1) frequent intubation, (2) prolonged ventilatory support, and (3) recurrent laryngeal nerve injury. In our patients, subglottic stenosis was the most common laryngeal abnormality. When recognized early, in the eschar phase, most of these cases can be managed with sequential endoscopic debridement, which is conceptually similar to debridement that is performed after functional endoscopic sinus surgery. Established stenosis requires more vigorous intervention, the invasive degree of which depends on the length and circumference of the narrowing. Unilateral vocal paralysis tends to be a self-limited problem, while an elegant solution to bilateral paralysis remains elusive.


CME course available at www.archoto.com

CONGENITAL AIRWAY ABNORMALITIES are responsible for a significant percentage of referrals to otolaryngologists for respiratory symptoms.¹ These abnormalities include laryngomalacia, congenital laryngeal web, congenital subglottic stenosis, vocal cord paralysis, and laryngeal cleft. Patients with concomitant cardiac anomalies have demonstrated a higher incidence of congenital laryngeal anomalies.²,³ Although this association has been frequently noted, there is a paucity of literature that characterizes these children.

In conceiving this study, it was hypothesized that pediatric patients with congenital heart disease have several predisposing factors for airway anomalies. In addition to the documented association of congenital laryngotracheal and cardiac anomalies, such patients are at risk for subglottic stenosis and iatrogenic vocal cord paralysis owing to multiple intubations, prolonged ventilation, and surgical procedures. We investigated the incidence of airway anomalies occurring after surgery for correction of cardiac anomalies.

METHODS

After obtaining institutional review board approval, diagnosis codes indicating simultaneous diagnoses of congenital cardiac disease and laryngotracheal anomalies in pediatric patients over a 4-year period were compiled into

Author Affiliations: Cleveland Clinic Foundation, Head and Neck Institute, Cleveland, Ohio (Drs Khariwala and Lee); and Division of Pediatric Otolaryngology, Stanford University School of Medicine, Stanford, Calif (Dr Koltai). Financial Disclosure: None.
Subglottic stenosis and unilateral vocal cord immobility were the most common diagnoses in this series. The other otolaryngologic diagnoses are summarized in Figure 1. There were 16 cases of subglottic stenosis, which were graded according to the Cotton-Myer (C-M) classification system as follows: grade 1 (n=8 [50%]), grade 2 (n=3 [19%]), and grade 3 (n=5 [31%]). Patients in the early eschar phase (C-M grade 2) underwent 3 weekly serial endoscopic debridements with concomitant application of mitomycin. Patients with established subglottic stenosis were treated with 1 of 2 modalities: (1) serial dilation with application of mitomycin or (2) laryngotracheal reconstruction with cartilage augmentation. The 3 patients with stenotic eschar completely resolved their airway obstruction. The airway obstruction was resolved in the 8 patients who were treated with serial dilation and mitomycin for mild stenosis (C-M grade 1). Five patients with C-M grade 3 subglottic stenosis underwent cartilage graft laryngotracheal reconstruction. Their airway obstruction resolved and they underwent decannulation. One other patient underwent augmentation cartilage graft tracheal reconstruction for treatment of a tracheotomy-related stenosis.

There were 14 cases of vocal cord immobility in our study, 11 of which were unilateral. The left vocal cord was affected in 10 (91%) of the unilateral cases. None of the 11 patients with unilateral vocal cord immobility required operative treatment, and 9 (82%) of them eventually demonstrated full return of movement. Return to function generally occurred by 3 months (range, 8 weeks to 10 months). Of the 3 patients with bilateral vocal cord paralysis, 1 underwent decannulation after a posterior cricoid augmentation cartilage spreader graft, 1 died after cardiac surgery, and 1 is tracheotomy dependent. The study patients also had a wide range of congenital cardiac anomalies, including tetralogy of Fallot, transposition of great vessels, hypoplastic left heart, atrial septal defect, ventricular septal defect, and DiGeorge syndrome (Figure 2).

An analysis of respiratory embryology helps to explain the association between congenital airway and cardiac anomalies. The respiratory system begins to develop as the respiratory primordium at 25 to 28 days of gestation. This is an epithelial thickening along the ventral aspect of the foregut. As the respiratory primordium develops, the respiratory diverticulum (an outpouching of the foregut lumen) grows into the respiratory primordium. When the respiratory diverticulum extends inferiorly, the primordial heart also proliferaes and is separated from the respiratory apparatus by the septum transversum. Because of the temporal and physical proximity of early cardiac and respiratory development, a single dismorphicogenic perturbation can involve both systems.

Several studies have documented the presence of associated congenital cardiac disease in pediatric patients with congenital laryngeal anomalies. Cohen et al observed that 7 of 51 patients with congenital laryngeal web had associated cardiac anomalies. Congenital laryngeal web has been noted to occur in 1% to 5% of patients with the 22q11 deletion (DiGeorge syndrome and velocardiofacial syndrome [VCFS]). McElhinney et al studied the frequency at which this deletion occurs in patients with congenital laryngeal web. They found that 25% of patients who underwent surgical intervention for congenital laryngeal web also had a 22q11 deletion. Miyamoto et al demonstrated an association of anterior glottic web (AGW) with the diagnosis of VCFS. In their study, 11 of 17 patients with AGW were positive for the chromosome 22q11 deletion. Of the 11 patients, 5 showed only subtle clinical signs of VCFS. The diagnosis of VCFS in these 5 patients was made purely on
the basis of high clinical suspicion in the setting of AGW. The authors recommended that all patients found to have AGW undergo chromosomal analysis for the 22q11 deletion. Furthermore, Gay et al\textsuperscript{9} described 4 patients from 1 family with laryngeal web, 3 of whom also had a ventricular septal defect. Shearer et al\textsuperscript{9} also reported a similar association.

We initially hypothesized that unilateral vocal cord paralysis would be the predominant diagnosis seen in our patient population. It seems likely that every patient who undergoes surgical repair of a cardiac anomaly is a risk for this problem owing to exposure of one or both recurrent laryngeal nerves during surgery. Surprisingly, subglottic stenosis was the most common problem. Unilateral vocal cord paralysis following congenital cardiac surgery is well recognized.\textsuperscript{10} Based on our experience, in cases in which full recovery was the usual outcome, stretching of the nerve(s) was the probable mechanism of injury, and the best predictor of return of function was the certainty of the cardiac surgeon about the integrity of the recurrent nerves at the end of the cardiac procedure.

The consistent presentation of unilateral paralysis is a weak cry after extubation, and the diagnosis is best made by means of bedside fiberoptic laryngoscopy. To confirm the presence of safe pharyngeal function, a modified barium swallow is obtained in these cases, and if there is any indication of aspiration, then thickening of the feedings, particularly of liquids, is initiated under the supervision of our speech therapy colleagues. Despite the occurrence of laryngeal penetration or aspiration in the perioperative period, swallowing appears to return to normal after several weeks to several months, even without return of full vocal cord motion, and the need for thickening of liquids resolves. Similarly, as compensation occurs, the voice gradually strengthens. Bilateral paralysis is much less common, although the consequences are highly problematic if it persists. Of our 3 patients with this devastating complication, 1 underwent decannulation after a posterior glottic cartilage spacer graft, 1 died after cardiac surgery with the tracheotomy still present, and 1 is tracheotomy dependent.

Although rare, congenital tracheal stenosis is important to consider in children with cardiac abnormalities, and patients with both conditions are well served by early diagnosis of the tracheal stenosis.\textsuperscript{11} A detailed review of the child’s history may reveal symptoms of airway obstruction that develop as the child outgrows the capacity of the narrowed airway, indicating the need for a diagnostic bronchoscopy. Unfortunately, the symptoms of congenital cardiac disease and tracheal stenosis may be similar, and in focusing on correcting the cardiac problem, the tracheal lesion can be overlooked. Some patients with complete tracheal rings can be treated nonsurgically, allowing the airway to grow and sparing unnecessary airway reconstruction.\textsuperscript{12} On the other hand, most children with complete tracheal rings do require intervention with pericardial patch or with slide tracheoplasty, which necessitates sternotomy and cardiopulmonary bypass.\textsuperscript{13} Early diagnosis of the tracheal stenosis allows repair of both lesions at the time of primary sternotomy. In our series, the patient with complete tracheal rings was first diagnosed as having a D-transposition of the great vessels. After correction of the cardiac anomaly, inability to extube led to a diagnostic bronchoscopy and the correct diagnosis. The tracheal stenosis was then corrected via a second sternotomy and slide tracheoplasty.

Subglottic stenosis was the more common problem among our children. While a predisposition for acquired subglottic stenosis may exist in children with cardiac disease as a result of congenitally smaller airway diameters, we found only 1 child with obvious congenital subglottic narrowing. Based on this finding, we conclude that the stenoses that developed were the result of the airway trauma that was sustained from intubation for the anesthetic or during the postoperative period, while the child was receiving ventilatory support. Pereira et al\textsuperscript{14} described 8 patients with congenital cardiac disease who developed subglottic stenosis and postulated that the stenoses were the result of endotracheal tube trauma. Koh et al\textsuperscript{15} described 19 of 181 patients who were found to have laryngeal edema after correction of congenital cardiac defects.

Our patients with airway obstruction due to stenosis had a variety of clinical presentations. Six failed extubation and underwent tracheotomy (4 patients) or immediate augmentation cartilage laryngotracheal reconstruction (2 patients). Three had persistent stridor immediately after extubation. These children were all found to have soft eschar narrowing the subglottis, and all responded to gentle serial debridement and treatment with mitomycin. The remaining 7 patients presented with progressive stridor irrespective of the length or diameter of their stenosis. In our study, membranous C-M grade 1 lesions and some short C-M grade 3 lesions were treated with serial dilation and mitomycin. We found this to be an efficacious and less invasive treatment modality. Children with long (≥ 5 mm) C-M grade 3 lesions require laryngotracheal reconstruction for consistently predictable success at resolving the obstruction.

**CONCLUSIONS**

Pediatric patients with congenital cardiac disease are predisposed to laryngeal abnormalities owing to the need for anesthetic intubation, prolonged ventilation, and surgery, which puts the recurrent laryngeal nerve at risk. In our patients, subglottic stenosis was the most common laryngeal abnormality. When the condition is recognized in the early eschar phase, most cases can be managed with gentle endoscopic debridement. Conceptually, this treatment is similar to postoperative nasal endoscopic debridement, which is performed after functional endoscopic sinus surgery. Established stenosis requires more vigorous intervention, the invasive degree of which depends on the length and circumference of the narrowing, but can be consistently and successfully treated with currently available techniques.

Unilateral vocal fold immobility was the second most common anomaly found in our patients. The left vocal cord was most commonly affected, and in these cases,
the patients demonstrated a high rate of spontaneous resolution. Therefore, expectant management of an immobile unilateral vocal cord after pediatric open heart surgery is generally well tolerated and recommended. An elegant solution to persistent bilateral vocal cord paralysis remains elusive.

Submitted for Publication: October 5, 2004; accepted December 16, 2004.

Correspondence: Peter J. Koltai, MD, Division of Pediatric Otolaryngology, Stanford University School of Medicine, 801 Welch Rd, Stanford, CA 94305 (koltai@stanford.edu).

REFERENCES