Airway Pathologic Abnormalities in Symptomatic Children With Congenital Cardiac and Vascular Disease

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Objective: To identify the epidemiological profile of airway abnormalities in symptomatic children with cardiac or vascular anomalies.

Design: Retrospective medical chart review.

Setting: Tertiary referral pediatric hospital.

Patients: Children with airway-related symptoms and coexistent cardiac or vascular abnormality were included. The source for patient identification was a prospectively kept database.

Main Outcome Measures: Endoscopic airway diagnoses, presenting airway symptoms, cardiac diagnoses, other comorbid conditions and pertinent diagnoses, patient demographics, source of referral, treatments, and follow-up.

Results: The study population comprised 77 patients (45 male and 32 female; mean age, 18.2 months) treated between June 2002 and July 2006. Only 4 patients had no findings. The most common airway abnormality was laryngeal paralysis (n=32), followed by subglottic stenosis (n=18). Congenital and acquired lesions were equally encountered (n=70 and n=64, respectively). The most frequent presentation was intolerance to feed (n=51) (stridor and/or failure of extubation). Of the 77 patients, 32 (42%) required airway surgical intervention (open vs closed); 36 (47%) still require otolaryngologic follow-up; and 32 (42%) had a named syndrome or general multi-system condition.

Conclusions: At least 3% of all children with cardiac disease will harbor airway problems. Laryngeal paralysis was the most common problem encountered. Given the successes achievable in treating children with complex cardiac abnormalities, attention should be paid to concomitant and consequential airway problems. Counseling processes should acknowledge the role of early otolaryngologic involvement.

Arch Otolaryngol Head Neck Surg. 2007;133(7):672-676

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METHODS

STUDY POPULATION

The study population comprised patients presenting to a tertiary referral pediatric otolaryngology practice between June 2002 and July 2006 with airway-related symptoms and a coexisting history of cardiac disease or vascular abnormality. Only patients who had undergone airway examination were included. The initial source for patient identification was a prospectively kept database (Microsoft Access 2000; Microsoft Corp, Redmond, Washington) of surgical and endoscopic procedures, created and updated by the senior author (H.E.) on an ongoing basis. Other sources were hospital and practice medical charts. All data collected were confirmed and cross-checked at least 2 of these sources. Ethics approval was sought and granted by the health research ethics board of the University of Alberta, Edmonton. All the patients were treated in the same pediatric center (The Stollery Children’s Hospital, Edmonton), which houses the second largest pediatric cardiac program in Canada.

Information collected included patient’s demographics; source of referral; presenting airway symptoms; cardiac, airway, and other diagnoses; endoscopic findings; treatments; and follow-up. The following section contains specific details on these variables.

DEFINITIONS AND VARIABLES

Patients’ age was noted at the time of their first airway endoscopy. The patients’ source of referral (intensive care unit, ward, or clinic) was recorded, and their presenting airway symptoms (stridor, voice change, exercise intolerance, or failure to extubate) were noted in the referral document. Exercise intolerance was based on the presence of persistent cough, shortness of breath, or wheezing during physical activity. Patients in whom 2 or more attempts to extubate had failed or who did not meet the criteria for extubation were included as “failure to extubate.” Patients who experienced persistent or frequent respiratory symptoms during feeding, prolonged feeding time, or repetitive vomiting during feeding and required change in route, pattern, or consistency of feeds were considered to be intolerant of feeding.

The type and condition of anesthetic used (none, local, general with spontaneous respiration, or general with muscle paralysis) was documented. Airway pathologic abnormalities were documented, and their cause (congenital vs acquired) and whether they were static or dynamic lesions were noted. Otolaryngologic treatments were recorded as closed (endoscopic techniques and adenotonsillectomy) and open (tracheostomy, cricoid split, and laryngotracheal reconstruction) surgical techniques or nonsurgical management. The date of last otolaryngologic assessment was also recorded to determine length of follow-up, and it was noted whether patients were currently being followed up in the clinic.

All cardiac diagnoses were recorded for the patient to date. Cardiac treatments were documented as open vs closed cardiac surgery or nonsurgical management. Other lines of medical management recorded included use of alternate feeding route and use of pressure ventilation. Other relevant diagnoses that were documented included prematurity, syndromes, and relevant coexistent diagnoses.

OUTCOME MEASURES AND LITERATURE SEARCH

Outcome measures included the prevalence of airway lesions in total and by type; proportion of congenital vs acquired lesions; proportion of static vs dynamic lesions; and sources of referral for these patients. Descriptive statistics were used for all variables.

RESULTS

A total of 77 patients satisfied the inclusion criteria (45 male and 32 female). The mean age was 18.2 months (range, 1 day–10.3 years). The patients were treated by the same pediatric otolaryngologist (H.E.) between June 2002 and July 2006 at the same institution.

The most common cardiac diagnosis was patent ductus arteriosus, diagnosed in 42 patients, followed by septal defect(s), diagnosed in 33 patients, of whom 15 had an atrioventricular septal defect, 8 had a ventricular septal defect, 6 had an atrial septal defect, and 4 had both an atrial septal and a ventricular septal defect. Hypoplastic left heart was diagnosed in 11 patients, tetralogy of Fallot in 7 patients, and an aberrant vessel in 5 patients, and there were 10 other miscellaneous diagnoses in 18 patients. Of the 77 patients, cardiopulmonary bypass and open cardiac surgery was performed in 49 (64%), closed heart surgery in 22 (29%), and nonsurgical management in 6 (8%).

Of the 77 patients, 30 (39%) from the neonatal intensive care unit and 27 (35%) from the pediatric intensive care unit were referred for otolaryngology consultation. Twenty patients (26%) were referred for urgent consultation by pediatricians, pulmonologists, or family physicians. The most common presenting airway symptom was intolerance to feed, experienced by 51 patients (66%), followed by 32 patients (42%) with stridor, 25 (32%) with voice change, 20 (26%) with failure to extubate, and 14 (18%) with exercise intolerance (Figure 1).

An English language literature search of indexed articles published between 1966 and 2006 was conducted. We searched PubMed using pediatric and English language search limitations and the following MeSH terms: cardiovascular system diseases and cardiovascular surgical procedures, each in combination with otolaryngology, pharynx, larynx, trachea, bronchi, bronchoscopy, and laryngoscopy. We checked the bibliographies of pertinent articles for other articles relevant to our study.
Laryngoscopy (flexible and rigid) was performed in 73 patients, bronchoscopy (rigid) in 63 patients, and suspension laryngoscopy in 14 patients (patients commonly had >1 type of endoscopy). The most common type of anesthetic used was general anesthetic with spontaneous respiration (n=41 [53%]), followed by general anesthetic with muscle paralysis (n=64 [82%]), no anesthetic (n=6 [8%]), and topical anesthetic (n=4 [5%]).

Of the 77 patients, 73 (94%) had positive airway findings. Over the same 4-year study period, our cardiac program performed approximately 2460 pediatric cardiac surgical procedures (open and closed). Considering that 73 patients had positive airway findings, this yields an overall prevalence of airway lesions (both congenital and acquired) of 3% in this population.

In total, we documented 134 airway findings in these 73 patients. Of these airway findings, 75 (56%) required dynamic assessment for diagnosis. Congenital and acquired airway pathologic lesions were encountered equally. There were 64 acquired airway problems, the most common being vocal cord paralysis (n=32 [27 unilaterally and 5 bilaterally]). The second most common acquired lesion was subglottic stenosis (n=18), followed by laryngeal granulations (n=11). An additional 3 acquired conditions were diagnosed, namely plastic bronchitis (n=2) and dislocated arytenoids (n=1) (Table 1).

There were 70 congenital pathologic cases, which included tracheomalacia (n=12), obstructive sleep apnea (n=11), bronchomalacia (n=10), and laryngomalacia (n=8), velopharyngeal insufficiency (n=4), bronchial stenosis (n=3), micrognathia (n=3), vocal cord cysts (n=3), tracheoesophageal fistula (n=3), choanal stenosis (n=3), hypotonic larynx and pharynx (n=3), microlarynx (n=2), lung collapse due to secretions (n=2), congenital long-segment tracheal stenosis (n=2), and abnormal bronchial branching (n=1) (Table 1).

Thirty-two patients required airway surgical intervention. Thirty closed or endoscopic procedures were used in 28 patients (36%), and 10 open techniques were used in 8 patients (10%); note that 4 patients received both open and closed interventions (Table 2). Seventy-five patients (97%) received nonsurgical management, which included acid suppression (n=54 [70%]), inhaled steroids (n=45 [58%]), systemic steroids (n=33 [43%]), and botulinum toxin type A (n=2 [3%]). Fifty-four patients received more than 1 of these forms of nonsurgical management. Alternate routes of feeding were used in 51 patients (66%), many of whom required more than 1 type during the course of their treatment. Pressure ventilation was used in 27 patients (35%).

Sixty-six of the patients attended otolaryngologic follow-up for a mean of 12.4 months (range, 6.0 days–3.7 years). Of the remaining 11 patients, 8 did not require follow-up after the initial otolaryngologic assessment, and the remaining 3 patients were lost to follow-up (n=1) or moved away (n=2). However, 36 patients (47%) were still being followed up at the time of the data collection.

In patients with laryngeal paralysis (n=32; mean age, 13.4 months), the most common presenting symptoms were voice change (n=20) and stridor (n=19). These were patients who underwent patent ductus arteriosus ligation (n=24) and/or surgery directly proximal to, or involving, the aortic arch (n=30).

For patients diagnosed as having subglottic stenosis (n=18; mean age, 8.36 months), their most common presenting symptoms were stridor (n=11) and failure to extubate (n=7). Of these 18 patients, 12 underwent open cardiac surgery; all 12 patients had repair of septal defects, 3 of whom had tetralogy of Fallot repairs and 2, hypoplastic left heart repairs. Five patients underwent closed cardiac repair for patent ductus arteriosus, and 1 was treated nonsurgically.

In the group of patients who had tracheobronchomalacia (n=18; mean age, 12.5 months), the most common presenting symptoms were failure to extubate (n=10) and stridor (n=9). Of these patients, 7 were diagnosed as having external compression (4 with an aberrant vessel, 1 with cardiomegaly, 1 with double aortic arch, and 1 with bilateral superior vena cava). Of the 18 patients, 14 underwent open cardiac surgery, 1 underwent closed surgery, and 3 required no cardiac management.

Of the 77 patients, 31 (40%) were born prematurely, and chronic lung disease was documented in 30 (39%). Thirty-two patients (42%) had another major named condition or syndrome: 12 had Down syndrome, 7 had cardiofacial syndrome, and 2 had Robin sequence/chromosome abnormalities. Other miscellaneous congenital conditions were diagnosed (1 of each of the following: cutis aplasia congenita, CHARGE [coloboma, heart disease, atresia choanae, craniofacial anomalies, genital anomalies, and hearing loss], and 11 other miscellaneous congenital conditions).
nae, retarded growth and development, genital hypoplasia, and ear anomalies], cerebral palsy, Turner syndrome, arthrogryposis multiplex, chromosome 22 duplication, VACTERL [vertebral, anal, cardiac, tracheal, esophageal, renal, and limb anomalies], chromosome 9q34 deletion, Moebius-Poland syndrome, achondroplasia, and thrombocytopenia–absent radius [TAR syndrome] (Figure 2).

The literature search resulted in 1336 hits; 37 abstracts were reviewed, of which 21 articles were used. The following specific airway and feeding problems have been investigated in pediatric cardiac patients: feeding problems after cardiac surgery, vocal cord paralysis, failure of extubation, laryngeal edema, requirement of tracheostomy, laryngeal web, subglottic stenosis, and hoarseness.5,7,10,12,16 One group explored the specific cardiac anomaly tetralogy of Fallot for associated tracheal anomalies, and another group investigated laryngopharyngeal dysfunction after the Norwood procedure.11,16 Three recent studies studied the prevalence of airway abnormalities and their morbidity associated with pediatric cardiac surgery and congenital heart disease.5,7,16

COMMENT

Our results indicate that nearly all children with heart disease or vascular abnormalities, presenting with an airway symptom, will have a major positive finding on endoscopy (94% [n = 73] in our series). We found an almost equal distribution of congenital and acquired airway pathologic anomalies, but laryngeal paralysis was by far the most common problem, followed by subglottic stenosis, tracheomalacia, obstructive sleep apnea, laryngeal granulations, and bronchomalacia. As expected, our main sources of referral were the pediatric and neonatal intensive care units (n = 57 [74%]). However, a considerable number of patients presented beyond the stage of intensive management and were seen on the pediatric ward or in the outpatient clinic instead (n = 20 [26%]). It should also be noted that the most common presenting symptom was actually intolerance to feed, followed by stridor, voice change, and failure to extubate. Thirty-two patients required surgical intervention, most being endoscopic techniques. However, nearly all patients received some form of medical treatment. As a result, 36 patients (47%) still require otolaryngologic follow-up, which may be attributable in part to other otolaryngologic problems, since 32 patients (42%) had a named syndrome or general multisystem condition.

Some of this information is different from the available English language literature on the topic. Only 3 broad studies were undertaken to describe the epidemiologic characteristics in this area. Many researchers were concerned with specific clinical problems in this group of patients (eg, failing to extubate and requirement for tracheostomy) and, naturally, were colored by that in addition to their primary area of expertise. Khariwala et al,1 a group of pediatric otolaryngologists, claimed that subglottic stenosis is the most common problem encountered after cardiac surgery. The study was retrospective, and the methods section documented few of the characteristics of the patient and none of the source and reason of the referral.3 A study by Lee et al3, a team of pediatricians, was aimed at all children with congenital heart disease presenting with airway obstruction. Their findings were biased toward lower airway pathologic disorders. While they share with Khariwala et al the characteristics of design, there is disparity between the 2 groups’ results. This probably stems from their exclusive use of flexible bronchoscopy (possibly a proportion performed through an indwelling tracheostomy or endotracheal tube) and lack of formal laryngeal examination.3 A study by Pfammatter et al shared most of the flaws mentioned and excluded patients with acquired pathologic disorders and patients with external airway compression. Based on the congenital lesions they found, they determined an overall prevalence of upper airway anomalies in pediatric cardiac surgical patients of 1.5%.

The present study avoids some of the mentioned pitfalls. Though retrospective in design, through searching a prospectively kept database (for procedures in and out of the operating room) and setting broad inclusion criteria, our results are more likely to reflect the real scope of the pathologic abnormalities and their distribution and certainly suffered from less attrition of data. The profile of the patients, demographics, secondary diagnoses, and near-complete information on follow-up reflects well on the plan and execution of the method. To our knowledge, this is the largest study in the English language literature.

Our determined overall prevalence of airway lesions is nearly 3%, double that of Pfammatter et al.3 However, there are 2 pediatric otolaryngology practices at our center. Assuming an almost equal contribution, the cited figure is probably a conservative one, if not an underestimation. This is bolstered by the fact that these children presented most commonly with feeding intolerance, which was prevalent in 18% to 29% in similar populations,7 many of whom do not necessarily get referred to an otolaryngologist.

Our assertion that laryngeal paralysis is the most common finding in this group of patients rests on several reasons, aside from anecdotal impressions of real-life experience. Namely, that previous work has indicated that...
unilateral laryngeal paralysis will occur in 8.8% of patients as a complication secondary to patent ductus arteriosus ligation.\textsuperscript{15} Also, preliminary research data from our center indicates it may occur more commonly than suggested, especially in extremely premature neonates, with greater associated morbidity than believed.\textsuperscript{20} Laryngeal paralysis, among other dynamic airway lesions (tracheomalacia, bronchomalacia and obstructive sleep apnea) will not be appreciated fully if spontaneous respiration is not preserved by the anesthetic technique. In terms of numbers, nearly half the lesions diagnosed in the present study would have been missed if a paralytic agent were used.

To our knowledge, we demonstrated for the first time that those airway problems acquired secondary to cardiac management were almost equal in prevalence to congenital ones. Just more than 40% required an active surgical method to treat the condition. Thus, it behooves us to inform the parents that a considerable number of children will require a major contribution from another surgical discipline for a long period, in part owing to the management of the original problem. Frustrating but true, laryngeal paralysis and tracheobronchomalacia are not always amenable to noninvasive treatment, nor do they have a well-delineated natural history.

The past 2 decades have witnessed higher rates of salvage in prematurely born infants, increased availability of better fiber optic instruments, and more sophisticated anesthetic techniques. Together, these factors deliver more patients with even more findings for pediatric surgeons in various disciplines. A recent multicenter study by Welke et al\textsuperscript{22} using data from 2001 to 2004 showed overall in-hospital mortality for congenital cardiac surgery to be 2.9%, a significant decrease from the 7.3% mortality rate reported by Jenkins et al\textsuperscript{22} using data from 1994 to 1996. With this in mind, aggressive otolaryngologic management would be well warranted to improve the quality of life of these children, as well as for further research and efforts into more timely and functional solutions. It would not be unreasonable to suggest a universal involvement from the outset, at least in children with major named syndromes and conditions. Most of these children would have head and neck problems that require our efforts regardless of their cardiac issues.

In conclusion, it appears that children with congenital heart disease have more airway insults than was previously thought, and a good proportion is secondary to their surgical and intensive care management. Dynamic airway lesions (most commonly laryngeal paralysis) play a considerable role and can be missed if the conditions of examination are not optimal. Just over 40% would need surgical correction, and nearly half of those affected require long-term follow-up by a pediatric otolaryngologist. There is a strong case for involving these specialists early on in the course of their cardiac management.

Submitted for Publication: December 16, 2006; final revision received March 18, 2007; accepted March 19, 2007. Correspondence: Handy El-Hakim, FRCS(Ed), FRCS (O.R.L.), Pediatric Otolaryngology, 2C.3.57 Walter MacKenzie Health Science Centre, 8440-112 St, Edmonton, AB T6G 2B7, Canada (haelhakim@cha.ab.ca).

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Financial Disclosure: None reported.

Previous Presentation: This study was presented at the 2006 American Academy of Otolaryngology–Head Neck Surgery Foundation Annual Meeting and OTO EXPO; September 19, 2006; Toronto, Ontario.

REFERENCES