IAPO-Interamerican Association of
Pediatric Otorhinolaryngology

Stridor in Children

Michael J. Rutter

Introduction

Stridor is a respiratory sound of variable pitch that is produced by turbulent airflow through a partially obstructed laryngeal or tracheal airway. It is not a diagnosis, but is rather a symptom of underlying airway pathology, and may be the first indication of a serious or life-threatening condition. This chapter outlines pertinent clinical and diagnostic information, and briefly describes many of the pediatric conditions in which stridor is present.

Clinical overview

- Unlike the harsh sound of snoring or the stertorous sound associated with pharyngeal obstruction, stridor has a characteristic musical quality.
- As stridor may be the initial symptom of serious airway pathology, it always warrants evaluation, including a thorough medical history and a diagnostic investigation.
- Whether the airway obstruction is fixed or dynamic, stridor is always present on inspiration. If the obstruction is fixed, stridor may also be present on expiration; this is termed ***biphasic stridor***.
- Because stridor is produced by turbulent airflow through the larynx or trachea, it is not normally seen in children who are intubated or tracheotomy dependent. If stridor is present in a child with a tracheotomy, it is of particular concern. This scenario indicates obstruction distal to or within the tracheotomy tube (eg, "mucous plugging") or that the tracheotomy tube is not lying in the trachea.
- If the etiology of stridor is supraglottic, glottic, or immediately subglottic, evaluation should include transnasal flexible laryngoscopy, as most etiologies are glottic or supraglottic and are diagnosable in an office setting. If flexible laryngoscopy is normal, formal bronchoscopic evaluation of the airway is indicated. This is carried out with the patient under general anesthesia.
- The most frequent causes of stridor are laryngeal (ie, laryngomalacia, vocal cord paralysis, and subglottic stenosis).
- Although stridor is the most blatant manifestation of airway obstruction, the degree of stridor does not necessarily reflect the severity of obstruction. Inspiratory retractions (whether suprasternal, intercostal, or substernal) allow for a more accurate evaluation of the degree of obstruction. In patients with severe obstruction, retractions may be marked whereas the associated stridor is comparatively subtle.

Diagnostic apprroaches

Laryngeal causes of stridor can generally be diagnosed in an awake and non-sedated child using transnasal laryngoscopy with a flexible endoscope; this examination can be performed in an office setting. This is the most effective approach for establishing a diagnosis and for triaging children who require formal bronchoscopic evaluation. If transnasal flexible laryngoscopy reveals no abnormality, the etiology of stridor may be subglottic or tracheal. In these patients, formal evaluation of the tracheal airway with rigid bronchoscopy is warranted and performed with the child under general anesthesia. This approach is preferable to flexible bronchoscopy, as it permits more adequate evaluation of the posterior glottis. Additionally, formal sizing of the airway can be performed.

Supraglottic causes of stridor

Laryngomalacia

Laryngomalacia is the most common cause of stridor in neonates. Stridor is generally mild, but is exacerbated by factors that increase airflow, such as crying, excitement, feeding, or lying in a supine position. In 50% of children with laryngomalacia, stridor worsens during the first 6 months of life. In virtually all cases, however, symptoms spontaneously resolve by 6 to 12 months of age.

In a subset of children with severe laryngomalacia (5%-10%), surgical intervention is required. Symptoms in these children may include apneic spells, cyanosis, severe retractions, and failure to thrive. In extremely severe cases, *cor pulmonale* is seen.

The diagnosis is confirmed by transnasal flexible laryngoscopy. Characteristic findings include short aryepiglottic folds, causing inspiratory supraglottic collapse and thus the presence of inspiratory stridor. In more than 50% of patients, significant arytenoid prolapse with the cuneiform cartilages prolapsing anteriorly into the airway can be seen. In up to 20% of patients, a tightly curled omega-shaped (Ω) epiglottis is also observed. Additionally, flexible laryngoscopy may reveal laryngeal irritation and edema. Gastroesophageal reflux disease is frequently associated with laryngomalacia, and is generally managed with either an H_2 antagonist or a proton pump inhibitor.[2]

Bronchoscopic evaluation is not indicated unless symptoms are severe enough to warrant intervention or the observed degree of laryngomalacia is disproportionate to the severity of the symptoms.

The decision as to whether or not to intervene surgically depends more on the severity of symptoms than on the endoscopic appearance of the larynx. For children who require surgical intervention, supraglottoplasty with division of the short aryepiglottic folds, and if indicated, removal of the cuneiform cartilages (if they are excessively mobile), is extremely effective.[3] If there is a risk of edema, overnight intubation is warranted.

When obstructive symptoms continue postoperatively, the possibility of underlying neurologic problems should be considered. Although such problems may be quite subtle initially, they may become more evident with time. This small subset of children is far more likely to require tracheotomy placement.

Tumors

The most frequently seen pediatric tumors affecting the larynx are **papillomas and lymphatic malformations**. **Papillomatosis** is discussed in the section on glottic obstruction. Lymphatic malformations that involve the larynx are usually seen as a spectrum of disease that also involves the tongue base, pharynx, floor of mouth, and parotid regions.[4] The degree to which the larynx is affected by a **lymphatic malformation** may vary significantly; however, because the lesion tends to expand rapidly in response to bacterial or viral infection, a low threshold for tracheotomy placement is advisable. Lymphatic malformations have a predilection for the epiglottic and false vocal cord areas and progression to subglottic and subglottic disease is unusual. In children with significant upper airway involvement, pharyngeal involvement generally takes precedence over laryngeal involvement. As such, laryngeal surgery should not be considered until pharyngeal disease is controlled, is no longer considered the primary cause of obstruction, and does not require maintaining a tracheotomy. If this is achievable, then debulking of disease in the supraglottic and epiglottic larynx may be considered. As recurrence is frequent, a significant period of quiescent disease is indicated before decannulation is attempted.

Cysts

Supralaryngeal cysts comprise lingual thyroglossal duct cysts, vallecular cysts, laryngoceles and saccular cysts. These cysts usually present in the neonatal period with a muffled cry and apneic spells that may be life threatening.

Lingual thyroglossal duct cysts classically occur in tongue base near the epiglottis. Although they are frequently asymptomatic in older children, they may be life threatening in neonates.[5] Excision is indicated and a transoral cyst excision is a straightforward and effective technique for managing the disease. In neonates, special care should be taken on induction of general anesthesia, as the airway is susceptible to complete obstruction during induction.

In contrast to thyroglossal duct cysts, which tend to be mid-line and deep to the mucosa, vallecular cysts are thin walled cysts near the glottic surface of the epiglottis. In general, these cysts are easily marsupialized.

Laryngoceles and saccular cysts may also cause severe upper airway compromise and occur when the laryngeal ventricle is obstructed.[6] Although endoscopic marsupialization may be attempted, recurrence is frequent and placement of a tracheotomy is often required. An open surgical approach with dissection of the cyst through the thyrohyoid ligament is more effective. This enables complete cyst removal and is thus curative.

Transnasal flexible laryngoscopy suggests the diagnosis of all of these cysts, and formal bronchoscopic airway evaluation confirms it. Radiographic evaluation with a computed tomography (CT) scan with contrast enhancement helps to ensure that a lingual thyroid is not mistaken for a lingual thyroglossal duct cyst and also delineates the extent of disease.

Supraglottic Infection

The introduction of effective immunizations has almost eliminated supraglottic causes of stridor. Two decades ago, epiglottitis was considered one of the most

significant causes of acute airway obstruction, with the most common infective organism being *Haemophilus influenza* type b. Because of the Hib vaccine, epiglottitis is no longer a significant disease process. Although it still occurs, it is more frequently a disease of older age groups and is associated with less acute airway obstruction, or it is caused by less virulent organisms.

Glottic causes of stridor

Laryngeal Webs

Laryngeal webs may be congenital or acquired. **Congenital webs** are rare and result from a failure of recanalization of the glottic airway in the early weeks of embryogenesis. Anterior glottic webs comprise more than 95% of congenital cases. Associated congenital anomalies occur in up to 60% of these children, and there is a strong association between anterior glottic webs and velocardiofacial syndrome.[7]

Acquired webs are **generally post-traumatic in origin**, either iatrogenic in nature or the result of direct trauma or inhalational injuries.

Congenital anterior glottic webs present with varying degrees of glottic airway compromise. The severity of symptoms correlates with the size and position of the web. Some webs are gossamer thin, though most are thick and are associated with a subglottic "sail" that compromises the subglottic lumen. Thin webs may elude detection, as neonatal intubation for airway distress may lyse the web, which is curative. Moderate to severe webs in infants may manifest in biphasic stridor and retractions, particularly when feeding or upset. These symptoms become increasingly evident as the infant grows.

Thick webs necessitate open reconstruction of the anterior commissure or placement of a laryngeal keel.[8] Intervention with the CO_2 laser is not advised, as it usually leads to recalcitrant web re-formation. The presence of thick membranous webs may require temporary placement of a tracheotomy to allow growth before elective laryngeal repair is performed. Repair is generally undertaken before the child reaches school age.

Vocal Cord Paralysis

Vocal cord paralysis is the second most common cause of neonatal stridor. It may be **congenital or acquired and unilateral or bilateral**. **Bilateral paralysis is usually congenital**, whereas **unilateral paralysis is commonly an acquired condition caused by damage to the recurrent laryngeal nerve**. Because of the length and course of the left recurrent nerve, it is far more likely to be damaged than is the right recurrent laryngeal nerve. As **such acquired paralysis generally affects the left vocal cord**. The risk factors for acquired paralysis are patent ductus arteriosus repair, the Norwood cardiac repair, and esophageal surgery, especially tracheoesophageal fistula repair. In older children, thyroid surgery is an additional risk factor.

Congenital vocal cord paralysis is usually idiopathic, but may also be seen with central nervous system pathology (eg, hydrocephalus and Chiari malformation of the brainstem). When the underlying cause is successfully treated, paralysis may be reversible. Although most children with bilateral paralysis exhibit significant airway compromise, they have excellent voice quality. Unless associated central

nervous system abnormalities are present, aspiration generally does not occur. Up to 90% of these infants ultimately require tracheotomy placement. In contrast, children with unilateral vocal cord paralysis generally have an acceptable airway, but a breathy voice. As well, they have a slightly higher risk of aspiration.

In an infant with stridor and retractions resulting from bilateral vocal cord paralysis, placement of a tracheotomy is indicated. Stabilization can be achieved with intubation, continuous positive airway pressure (CPAP), or high-flow nasal canula as an alternative temporizing measure. As up to 50% of children with congenital idiopathic bilateral vocal cord paralysis have spontaneous resolution of their paralysis by 1 year of age,[9] surgical intervention to achieve decannulation is usually delayed until this time. Similarly, children with acquired bilateral vocal cord paralysis may have spontaneous recovery several months after recurrent laryngeal nerve injury if the nerve is stretched or crushed but otherwise intact.

Surgical intervention is not required in most children with unilateral vocal cord paralysis. For those with bilateral paralysis, several surgical options have been used, since no particular approach offers a universally acceptable outcome.[10] Surgery is aimed at achieving an adequate decannulated airway while maintaining voice and avoiding exacerbation of aspiration. Surgical options include laser cordotomy, partial or complete arytenoidectomy (endoscopic or open), vocal process lateralization (open or endoscopically guided), and posterior cricoid cartilage grafting. In a child with a tracheotomy, it is often prudent to maintain the tracheotomy to ensure an adequate airway prior to decannulation. In a child without a tracheotomy, a single-stage procedure can be performed.

Acquired bilateral vocal cord paralysis is usually more recalcitrant to treatment than idiopathic cord paralysis, and more than one operative intervention may be required to achieve decannulation. In patients who have undergone any such procedures, post-extubation stridor may respond to CPAP or high-flow nasal cannula. A child's postoperative risk of aspiration should be assessed by a video swallow study prior to resuming a normal diet. During the initial weeks following surgery, there is sometimes an increased aspiration risk with certain textures, particularly thin fluids.

Posterior Glottic Stenosis

Posterior glottic stenosis may occur as an isolated entity or may exist along with subglottic stenosis. It is frequently misdiagnosed and confused with bilateral true vocal cord paralysis. The most common etiology of posterior glottic stenosis is prolonged intubation. Older children are at greater risk than the neonate. Patients who are not already tracheotomy dependent present with stridor and exertional dyspnea.

Awake transnasal flexible laryngoscopy confirms the lack of cord abduction. In contrast to vocal cord paralysis, there is considerable cord movement. Definitive diagnosis requires assessment with rigid bronchoscopy, which confirms the presence of posterior glottic scarring. Bronchoscopic evaluation should include assessment of the subglottis, which also may be involved with scarring. Because joint fixation is sometimes a coexistent pathology, an assessment of arytenoid mobility is required. Formal sizing of the airway utilizing endotracheal tubes may

be misleading. Flexible bronchoscopy provides a poor view of the posterior glottis and cannot be relied on to establish a definitive diagnosis.

Placement of a posterior costal chondral graft is the mainstay of management and is a highly effective way of achieving an adequate glottic airway.[11] This procedure is best performed through an anterior approach, traditionally through a complete laryngofissure, division of the posterior cricoid plates, and placement of the cartilage graft.

This procedure may be performed without a complete laryngofissure. If the anterior airway incision is carried up to the true vocal cords but not through them, there is usually sufficient access to perform a safe posterior cricoid split and allow insertion of a flanged graft. Such a graft is required as there is not adequate access to comfortably place sutures. This technique is more difficult to perform in small children.

Although endoscopic techniques may also be effective, they are usually not as reliable as the open approach. A laser posterior cordotomy with or without partial arytenoidectomy carries a significant chance of restenosis. To prevent this, the use of Mitomycin C and placement of a temporary transglottic stent should be considerations. Other described techniques for the endoscopic management of posterior glottic stenosis include mucosal advancement flaps, micro-trapdoor flaps, vocal cord lateralization, and Botox injections.

Both open and endoscopic techniques are associated with a restenosis rate ranging from 10% to 20%. This rate is higher in children who have had thermal injury to the posterior glottic stenosis. If a child has failed an endoscopic procedure, then open reconstruction is appropriate. Conversely, if a child has failed an open procedure, endoscopic management may be appropriate. In a very recalcitrant airway, it is possible to place a second posterior costal cartilage graft if required.

Papillomatosis

Recurrent respiratory papillomatosis (RRP), also known as juvenile laryngeal papillomatosis, is the most common infective lesion of the larynx in children. The average age at diagnosis of RRP is 4 years, and nearly 75% of cases are diagnosed by age 5. The etiology of RRP is infection of the upper airway with human papillomavirus (HPV) types 6 and 11, and less commonly, types 16 and 18.[12] The pathogenesis of RRP is frequently associated with transplacental transmission of maternal HPV; however, contact with active cervical HPV during delivery is also considered a causal factor. It is nevertheless important to note that although genital papillomas are extremely common, RRP is extremely rare and the relative risk of acquiring RRP is low. In view of this low risk and because transmission may be transplacental, maternal HPV should not be used as a justification for performing a Cesarian delivery. Similarly, a Cesarian delivery is unjustified for the delivery of a sibling of a child with RRP, as the risk to the sibling of developing RRP approaches zero.

RRP is often misdiagnosed as asthma, recurrent croup, or bronchitis. Stridor due to airway obstruction is common, and often precipitates otolaryngologic referral. Initial evaluation is carried out with flexible transnasal laryngoscopy, which may reveal a laryngeal mass. Subsequent microlaryngoscopy and bronchoscopy with

biopsy of the papillomas is required for a definitive diagnosis and to serotype lesions for prognostic purposes. Serotypes 16 and 18 are associated with more aggressive disease and a higher risk of malignant transformation.

Although the course of the disease is both variable and unpredictable, RRP tends to recur locally and, in severe cases, spreads throughout the respiratory tract. Surgical intervention should be based on debulking gross disease without attempting complete removal of the affected tissue, so to avoid laryngeal scarring or stenosis. The most widely used surgical procedure is suspension laryngoscopy with tumor removal using the CO_2 laser, microforceps, or the microdebrider. In patients with extensive disease, surgery should be aimed at reducing the tumor burden, decreasing the spread of disease, creating a patent airway, improving voice quality, and lengthening the intervals between surgical interventions. In children with severe RRP, tracheotomy placement may be required; however, this is often at the cost of disseminating disease beyond the glottis.

Numerous attempts at finding medical alternatives for controlling RRP have been made, with repeated cycles of hope tempered by reality. Unsuccessful therapeutic trials have included antibiotics, steroids, the topical antiviral agent podophyllin, and antimetabolites (5-fluorouracil and methotrexate). The current therapy of choice is direct lesional injection of Cidofovir — an antiviral agent with a broad spectrum of activity against a wide variety of DNA viruses, including HPV. However the best long term hope for management of this debilitating disease is the development of a therapeutic vaccine.

Subglottic causes of stridor

Subglottic Cysts

Subglottic cysts are typically a consequence of prolonged intubation of a premature infant. They may be superficial and thin walled or may lie deep in the submucosal layer, and are often multiple. Although the pathogenesis of subglottic cysts is unlike that of subglottic stenosis, both problems may coexist. Management includes de-roofing the cyst using microlaryngeal instrumentation, powered instrumentation, CO_2 laser, or Bugbee electrocautery.[13] Because subglottic cysts tend to recur, followup bronchoscopy is essential. Removal may need to be performed on several occasions before complete resolution is attained.

Subglottic Hemangioma

Nearly all hemangiomas occurring within the tracheobronchial tree are in the subglottis. The natural history of these lesions mirrors that of cutaneous hemangiomas. A phase of proliferation is followed by a phase of spontaneous involution; however, lesions expand and involute more rapidly than cutaneous lesions. Involution typically begins by 12 months of age and is completed by 18 to 24 months of age. Fifty percent of children with subglottic hemangiomas also have cutaneous lesions. Subglottic hemangiomas are seen more frequently in patients with cervicofacial hemangiomas that cover a beard distribution including the chin, jawline, and preauricular areas.[14] These patients should therefore be closely monitored for airway involvement. Most infants present at 2 to 4 months of age, and the earlier the presentation of symptoms, the more likely it is that the child will require surgical intervention. Characteristic symptoms include

progressive stridor and retractions. Initial evaluation with transnasal flexible laryngoscopy may allow for visualization of the compromised subglottis, though more importantly, it should exclude other causes of neonatal stridor, particularly laryngomalacia and vocal cord paralysis. A child with progressive stridor and a normal glottic laryngeal examination on flexible laryngoscopy requires a formal laryngoscopic and bronchoscopic evaluation. Preoperative plain (high kilovolt) airway films are advisable to evaluate the subglottic airway. Typically, a subglottic hemangioma will cause an asymmetric narrowing of the subglottis. Airway imaging may be of value in selected cases, particularly if mediastinal extension is suspected.

Traditional management has been placement of a tracheotomy tube, with the expectation of decannulation between 1 and 2 years of age. In children with mild to moderate symptoms, high-dose systemic steroids may cause involution of the lesion and prevent the need for other surgical intervention; however, their prolonged use is strongly discouraged. A number of alternatives to tracheotomy placement have been used, including intralesional steroids, prolonged intubation, CO_2 and KTP laser ablation, and microdebrider resection.[15] Drawbacks of these techniques include inadequate removal of the hemangioma and the development of subglottic stenosis. In the author's experience, open surgical resection of the subglottic hemangioma is the intervention of choice in children with moderate to severe symptoms.[16] This is due to the association of subglottic hemangioma with mild to moderate congenital subglottic stenosis in most cases. Whereas other treatment modalities carry a significant risk of inducing an acquired on congenital subglottic stenosis, open surgical resection allows both removal of the hemangioma and simultaneous repair of the congenital subglottis stenosis if present.

Subglottic Stenosis

Subglottic stenosis can be either congenital or acquired. Congenital subglottic stenosis is thought to result from failure of the laryngeal lumen to recanalize, and is one of a continuum of embryologic failures that include laryngeal atresia, stenosis, and webs. Although this condition is uncommon, it is the third most frequent cause of congenital stridor in infants. In the neonate, subglottic stenosis is defined as a lumen 4.0 mm in diameter or less at the level of the cricoid. Acquired subglottic stenosis is more common and is generally a sequela of prolonged intubation of the neonate. A useful and practical guide is that the outer diameter of a 3.0 endotracheal tube is 4.3 mm, and if air leaks around the tube at less than 20 cm of subglottic pressure of water, then the subglottis is not stenotic.__

Levels of subglottic stenosis severity are graded according to the Myer-Cotton grading system (grades I to IV), with grade I ranging from no obstruction to 50% obstruction and grade IV being no detectable lumen.[17] Mild subglottic stenosis may manifest in recurrent upper respiratory infections (often diagnosed as croup) in which minimal subglottic swelling precipitates airway obstruction. More severe cases may present with acute airway compromise at delivery. If endotracheal intubation is successful, the patient may require intervention before extubation. When intubation cannot be achieved, tracheotomy placement at the

time of delivery may be life saving. Important to note, infants typically have few symptoms, and because growth of the child exceeds growth of the airway, even those with grade III stenosis may not be symptomatic for weeks or months.

Congenital subglottic stenosis is often associated with other congenital head and neck lesions and syndromes (eg, a small larynx in a patient with Down syndrome). After initial management of subglottic stenosis, the larynx will grow with the patient and may not require further surgical intervention; however, if initial management requires intubation, there is considerable risk of developing an acquired subglottic stenosis in addition to the underlying congenital subglottic stenosis.

Radiologic evaluation of an airway that is not intubated may give the clinician clues about the site and length of the stenosis. The single most important investigation is a high-kilovoltage airway film. This is taken not only to identify the classic "steepling" observed in patients with subglottic stenosis, but also to identify possible tracheal stenosis.

Evaluation of subglottic stenosis (whether it is congenital, acquired, or a combination of both), requires endoscopic assessment; ideally, this is done with a Hopkins rod rigid telescope. Precise evaluation of the endolarynx should be carried out, including grading of the subglottic stenosis. Stenosis caused by scarring, granulation tissue, submucosal thickening, or a congenitally abnormal cricoid can be differentiated from subglottic stenosis with a normal cricoid, but endoscopic measurement with endotracheal tubes or bronchoscopes is required for an accurate evaluation.

The greatest risk factor for developing acquired subglottic stenosis is prolonged intubation with an inappropriately large endotracheal tube. The appropriate endotracheal tube size is not the largest that will fit, but rather the smallest that allows for adequate ventilation. Ideally, the tube should leak air around it, with subglottic pressures below 25 to 30 cm of water. Other cofactors for the development of acquired subglottic stenosis include gastroesophageal reflux and eosinophilic esophagitis.

Children with mild acquired subglottic stenosis may be asymptomatic or minimally symptomatic. Observation rather than intervention may thus be appropriate. This is often the case for children with grades I or II disease. Those with more severe disease are symptomatic, with either tracheal dependency or stridor and exercise intolerance. Unlike congenital subglottic stenosis, acquired stenosis is unlikely to resolve spontaneously and thus requires intervention.

In children with mild symptoms and a minor degree of subglottic stenosis, endoscopic intervention may be effective. Endoscopic options include radial laser incisions through the stenosis and laryngeal dilatation. More severe forms of disease are better managed with open airway reconstruction. Laryngotracheal reconstruction using costal cartilage grafts placed through the split lamina of the cricoid cartilage is reliable and has withstood the test of time.[18,19] Costal cartilage grafts may be placed through the anterior lamina of the cricoid cartilage, the posterior lamina of the cricoid cartilage, or both. These procedures are usually performed as a two-stage procedure, maintaining the tracheal tube

and temporarily placing a suprastomal laryngeal stent above the tracheal tube. Alternatively, in selective cases, a single-stage procedure may be performed, with removal of the tracheal tube on the day of surgery and with the child requiring intubation for a 1- to 14-day period.[20] More recently, better results have been obtained with cricotracheal resection than with laryngotracheal reconstruction for the management of severe subglottic stenosis; nevertheless, this is a technically demanding procedure that carries a significant risk of complications.[21]

Reconstruction of the subglottic airway is a challenging procedure and the patient should be optimized before undergoing surgery. Preoperative evaluation includes assessment and management of gastroesophageal reflux, eosinophilic esophagitis, and low-grade tracheal infection, particularly methicillin-resistant *Staphylococcus aureus* (MRSA) and *Pseudomonas aeruginosa*.

Croup

Croup is a viral condition caused primarily by parainfluenza virus types I and II, though other viruses have also been implicated. The age range of affected children is generally from 3 months to 5 years of age, with a peak at age 2 years. The condition presents in late fall and early winter, and transmission occurs by direct contact. Children with croup generally have a low-grade fever, rhinorrhea, and a characteristic barking cough, with accompanying inspiratory stridor and retractions. Symptoms rarely last longer than three days. In mild cases, investigations are not warranted. In more severe episodes, confirmation of the diagnosis is made radiographically. Airway films show characteristic "steepling" of the subglottis. Flexible laryngoscopy confirms that no other supraglottic exists and that there is an edematous subglottic airway. Treatment is usually supportive, with humidity and cool air being useful in the home setting. In more severe cases, systemic steroids or racemic epinephrine are effective. Rarely, intubation may be required.[22] Bronchoscopy is indicated only when croup is atypical – this is, when it is severe, when it occurs at an atypical age, or in cases of crescendo croup, with each episode being worse than the previous episode. In some children croup is more severe due to a mild underlying congenital subglottic stenosis.

Tracheal causes of stridor

Complete Tracheal Rings

Complete tracheal rings are a rare but life-threatening anomaly. They present with insidious worsening of respiratory function over the first few months of life and with stridor, retractions, and marked exacerbation of symptoms during intercurrent upper respiratory tract infections. Children with distal tracheal stenosis usually have a characteristic biphasic wet-sounding breathing pattern that transiently clears with coughing. The risk of respiratory failure increases with age. Over 80% of children with complete tracheal rings have other congenital anomalies; these anomalies are generally cardiovascular in origin.

Although diagnosis is made with rigid bronchoscopy, an initial high-kilovolt airway film may warn of tracheal narrowing. Bronchoscopy should be performed with great caution, using the smallest possible telescopes, as any airway edema in the region of the stenosis may turn a narrow airway into an extremely critical airway. The location, extent, and degree of stenosis are all relevant; however, if

the airway is exceptionally narrow, it may be more prudent just to establish the diagnosis rather than to risk causing post-traumatic edema by forcing a telescope through a stenosis. Because 50% of children have a tracheal inner diameter of approximately 2 mm at the time of diagnosis, the standard interventions for managing a compromised airway are not applicable. More specifically, the smallest endotracheal tube has an outer diameter of 2.9 mm and the smallest tracheotomy tube has an outer diameter of 3.9 mm; as such, the stenotic segment cannot be intubated. Extracorporeal membrane oxygenation (ECMO) may thus be left as the only viable alternative for stabilizing the child. This situation is best avoided by performing bronchoscopy with the highest level of care. Over 80% of children with complete tracheal rings have other congenital anomalies, which are generally cardiovascular in origin. In view of frequent cardiovascular anomalies, investigation should include a high-resolution contrast-enhanced CT scan of the chest and an echocardiogram.

Most children with complete tracheal rings require tracheal reconstruction.[23] The recommended surgical technique is the slide tracheoplasty.[24] This approach yields significantly better results than any other form of tracheal reconstruction and is applicable to all anatomic variants of complete tracheal rings.

Bacterial Tracheitis

Bacterial tracheitis is an uncommon but sometimes life threatening condition. Unlike croup, a number of organisms are implicated in bacterial tracheitis;[25] these include *Staphylococcus aureus,* and less commonly, alpha-hemolytic streptococcus, *Haemophilus influenzae, Moraxella catarrhalis,* and *Streptococcus pneumoniae.* Affected children are usually otherwise healthy and between 6 and 11 years of age. Children present with a severe sore throat that is exacerbated by coughing and swallowing. This in turn restricts fluid intake and results in increasingly thick tenacious tracheal secretions. Marked subglottic edema is typically observed and presentation is with stridor and retractions. In advanced cases, the child may be febrile and appear toxic. Airway films show tracheal membranes and flexible laryngoscopy shows thick subglottic secretions that are difficult for the patient to cough out. In mild cases, rehydration with intravenous fluids, analgesia, broad spectrum antibiotics and encouragement to cough may be sufficient to manage the disease. In most cases, however, bronchoscopic clearing of the airway is required. Bronchoscopic findings are of subglottic and tracheal edema, with thick tenacious secretions in the trachea, but sparing of the bronchi. Occasionally the secretions are so tenacious that they cannot be adequately suctioned, and removal with foreign body forceps may be required. Postoperative intubation for several days is frequently required.

References

1. Holinger L. Etiology of stridor in the neonate, infant, and child. Ann Otol Rhinol Laryngol 1980;89:397.
2. Giannoni C, Sulek M, Friedman EM, et al. Gastroesophageal reflux association with laryngomalacia: A prospective study. Int J Pediatr Otorhinolaryngol 1998;43:11-20.
3. Denoyelle FM, Mondain M, Gresillon N, et al. Failures and complications of supraglottoplasty in children. Arch Otolaryngol Head Neck Surg 2003;129:1077-80.
4. Azizkhan RG, Rutter MJ, Cotton RT, Lim HY, et al. Lymphatic malformations of the tongue base. J Pediatr Surg 2006;41:1279-1284.
5. Bayard RW, Bourne AJ, Silver MM. The association of lingual thyroglossal duct remnants with sudden death in infancy. Int J Pediatr Otorhinolaryngol 1990;20:107-112.
6. Ward, RF, Jones J, Arnold JA. Surgical management of congenital saccular cysts of the larynx. Ann Otol Rhinol Laryngol 1995;104 (9 Pt 1):707-710.
7. Miyamoto RC, Cotton RT, Rope AF, Hopkin RJ, et al. Association of anterior glottic webs with velocardiofacial syndrome (chromosome 22q11.2 deletion). Otolaryngol Head Neck Surg 2004;130:415-417.
8. Wyatt ME, Hartlley BE. Laryngotracheal reconstruction in congenital laryngeal webs and atresias. Otolaryngol Head Neck Surg 2005;132:232-238.
9. Miyamoto RC, Parikh SR, Gellad W, et al. Bilateral congenital vocal cord paralysis: a 16-year institutional review. Otolaryngol Head Neck Surg 2005;133:241-245.
10. Hartnick CJ, Brigger MT, Willging JP, et al. Surgery for pediatric vocal cord paralysis: a retrospective review. Ann Otol Rhinol Laryngol 2003;112:1-6.
11. Rutter MJ, Cotton RT. The use of posterior cricoid grafting in managing isolated posterior glottic stenosis in children. Arch Otolaryngol Head Neck Surg 2004; 130:737-739.
12. Pransky SM, Kang DR. Tumors of the larynx, trachea, and bronchi. In: Bluestone CD, Stool SE, Alper CM et al, eds. Pediatric Otolaryngology, chap 91. Philadelphia, PA: Saunders, 2003.
13. Johnson LB, Rutter MJ, Shott SR, Cotton RT. Acquired subglottic cysts in preterm infants. J Otolaryngol 2005;34:75-78.
14. Orlow SJ, Isakoff MS, Blei F. Increased risk of symptomatic hemangiomas of the airway in association with cutaneous hemangiomas in a “beard” distribution. J Pediatr 1997;131:643-646.
15. Pransky SM, Canto C. Management of subglottic hemangioma. Curr Opin Otolaryngol Head Neck Surg 2004;12:509-512.
16. Vijayasekaran S, White DR, Hartley BE, Rutter JM, Elluru RG, Cotton RT. Open excision of subglottic hemangiomas to avoid tracheostomy. Arch Otolaryngol Head Neck Surg 2006;132:159-163.

17. Myer CM, O'Connor DM, Cotton RT. Proposed grading system for subglottic stenosis based on endotracheal tube sizes. Ann Otol Rhinol Laryngol 1994;103:319-323.
18. Cotton RT, Gray SD, Miller RP. Update of the Cincinnati experience in pediatric laryngotracheal reconstruction. Laryngoscope 1989;99:1111-16.
19. Cotton RT. The problem of pediatric laryngotracheal stenosis; a clinical and experimental study on the efficacy of autogenous cartilaginous grafts placed between the vertically divided lamina of the cricoid cartilage. Laryngoscope 1991;101:1-34 (suppl 56).
20. Gustafson LM, Harley BE, Liu JH, et al. Single-stage laryngotracheal reconstruction in children: a review of 200 cases. Otolaryngol Head Neck Surg 2000;123:430-434.
21. White DR, Cotton RT, Bean JA, et al. Pediatric cricotracheal resection: surgical outcomes and risk factor analysis. Arch Otolaryngol Head Neck Surg 2005;131:896-9.
22. Leung AK, Kellner D, Johnson DW. Viral croup: A current perspective. J Pediatr Health Care 2004;18:297-301.
23. Rutter MJ, Cotton RT. Tracheal stenosis and reconstruction. In: Mattei P, ed. Surgical Directives: Pediatric Surgery, chap 24. Philadelphia, PA; Lippincott, Williams & Wilkins, 2003.
24. Rutter MJ, Cotton R, Azizkhan RG, et al. Slide tracheoplasty for the management of complete tracheal rings. J Pediatr Surg 2003;38:928-34.
25. Salamone FN, Bobbitt DB, Myer CM, et al. Bacterial tracheitis reexamined: Is there a less severe manifestation? Otolaryngol Head Neck Surg 2004;131:871-876.