Subglottic Stenosis

Gabriel Marshak, Kennet M. Grundfast

(Pediatric Clinics of North America, Vol 28, No 4, November 1981)

Subglottic stenosis refers to a narrowing of the space bounded inferiorly by the inferior margin of the cricoid cartilage and superiorly by the inferior surface of the vocal cord. This space is the narrowest portion of the upper airway, the lumenal diameter in a full-term newborn baby being 4.5 to 5 mm. This key area is predisposed to injury because of certain inherent anatomic features: it is a circular structure surrounded by a rigid, intact, cartilaginous ring abutting the spinal column that is exposed to external trauma. In addition to these factors, this area is the narrowest portion of the airway lined by respiratory epithelium that is prone to mechanical trauma and submucosa of loose connective tissue (which is capable of edematous swelling), thus rendering it susceptible to internal (endolaryngeal) trauma

Congenital Subglottic Stenosis

Subglottic stenosis is considered to be congenital in the absence of a history of endotracheal intubation or other apparent cause. It is the most common laryngeal disorder that might produce serious airway obstruction requiring tracheotomy in infants under one year of age. Congenital laryngotracheal anomalies, such as tracheoesohageal fistula, tracheal stenosis, and vocal cord paralysis, occur in 7 per cent to 10 per cent of patients with subglottic stenosis, and congenital anomalies not related to the larynx and trachea have been reported in 10 per cent of these patients.

The narrowing of the lumen may be caused by both cartilaginous and soft tissue deformities. Cartilaginous ring has been described to be unusually small in diameter or elliptical in shape; in rare cases, the first tracheal ring is "trapped" or "telescoped" insice the anterior cricoid arch. The soft tissue abnormality results from thickness of the submucosa that is caused by either connective tissue or by hyperplastic dilated mucous glands. The narrowing is circumferential or could occupy only part of the ring. The symptoms and their severity, and age at onset are clearly related to the amount of narrowing of the subglottic space. In severe cases, respiratory distress associated with stridor will be present at birth; this was true for half of the subjects in the largest series of patients with congenital subglottic stemosis reported by Hollinger. Milder cases become evident during the first weeks or months of life, presenting with either a prolonged episode of croup or recurrent croup. Laryngeal inflammation can precipitate severe respiratory distress since the limiting cricoid cartilage only permits inward swelling of tissues at the expense of the airway. Thus an airway already compromised by congenital subglottic stenosis will become inadequate by the slight edema associated with a viral infection. Ten per cent of these children will have recurrent pneumonitis. A barking or brassy cough may be the only symptom of minimal subglottic stenosis. The most important sign, if present, and that in no way is pathognomonic, is stridor, which is often both inspiratory and expiratory, and of low pitch.

The diagnosis of congenital subglottic stenosis is strongly suggested by the history and clinical presentation, assuming that a high index of suspicion exists. Lateral and anteroposterior neck films, laryngeal tomograms, and lateral xeroradiograms may provide

further evidence, but the ultimate proof is the endoscopic diagnosis. Subglottic stenosis is confirmed in a full-term newborn infant if a 3 mm infant bronchoscope (having an outer diameter of 4 mm by 4.5 mm) cannot pass readily through the subglottic region. A complete evaluation of the tracheobronchial tree is mandatory to rule out other concomitant breathing anomalies.

The management of congenital subglottic stenosis is based on the experience that children outgrow the disorder. In our experience, less than half of the patients will require tracheostomy as an alternative airway, most of which will be decannulated within two years. Only a small number will require surgical correction. The smallest tracheostomy tube that will permit adequate ventilation should be used in order to allow preservation of the function of the larynx and to permit the child to phonate. With the newly developed tracheostomy tube, adequate care can be administered at home after the parents receive appropriate instructions. The mortality rate in this group of patients, which is directly associated with tracheostomy, is still about 2 per cent. Periodic endoscopic dilatations of the stenotic area appear to help, but no control studies have been reported.

Acquired Subglottic Stenosis

The wide variety of mechanisms that potentially porduce subglottic stenosis is often surprising to those unfamiliar with this subject. Although it may be caused by chemical or thermal inhalation, benign or malignant neoplasm, or even connective tissue disorders such as lupus erythematosus, there has been a definite increase inthe incidence of chronic subglottic stenosis in children caused by blunt external laryngeal trauma. This is partly the result of an increasing number of vehicular accidents in which the extended neck (caused by the acceleration/deceleration events involved in car accidents) hits the dashboard or the front seat, resulting in closed laryngeal fractures. The other major cause of external blunt trauma to the larynx is the "clothesline" injury: a child who is riding a minibike or trailbike can be hit in the anterior neck by a branch or a clothesline, sustaining a laryngeal fracture that may later result in subglottic stenosis. Such injuries may require emergency care but occasionally may present later with progressive upper respiratory obstruction caused by subglottic (and/or laryngeal) stenosis.

Endotracheal Intubation

By far, however, the leading cause of chronic subglottic stenosis in children is prolonged endotracheal intubation. This noninvasive modality, first advocated by McDonald and Stocks in 1965 to provide an airway and as an adjunct to artificial ventilation, is being used with increasing frequency in the management of the critically ill neonate and child. The reported incidence of chronic subglottic stenosis following prolonged endotracheal intubation in neonates and infants ranged from 12 to 20 per cent in the late 1960s and early 1970s, to from 4 to 8.5 per cent at present. Five per cent of neonates intubated for respiratory distress syndrome have been reported to have chronic subglottic stenosis. The actual incidence is probably higher since these data include only patients who had survived their primary illness. All reports also include only patients who demonstrated subglottic stenosis while still hospitalized or shortly thereafter, but fail to report an extended follow-up which might unveil subglottic insufficiency only during a "cold" or following a surgical procedure requiring a short endotracheal intubation. Thus, as neonatal and infant survival is improved in the newly

developed neonatal and pediatric intensive care units, more cases of subglottic stenosis secondary to prolonged endotracheal intubation are expected to be recognized.

The patient may require endotracheal intubation to ensure an adequate airway for weeks and sometimes months, resulting in continuous trauma to the larynx that may ultimately result in chronic subglottic stenosis. Several factors contributing to this trauma have been elucidated: (1) direct trauma to the mucous membrane by traumatic intubation or by repeated intubations; (2) the piston action of the respirator and the abrasive action of the tube against the mucosa during swallowing or in restless patients; (3) indirect trauma to the mucosa and cartilage from the continuous pressure of a large tube or the inflated cuff, causing ischemic necrosis of the mucous membrane and cartilage; (4) superimposed bacterial infection compounding the mechanical trauma; (5) chemical irritation of tubes made from rubber or plastic other than polyvinyl chloride, as well as toxic residues of sterilization with ethylene oxide gas; and (6) prolonged periods of intubation.

Development of Stenosis

The histopathologic sequence in the development of stenosis in neonates and children begins with mucosal edema and hemorrhage, followed by ulceration of the mucosa. With time, infection, or both, the ulceration becomes more extensive, penetrating into the perichondrium and cartilage and causing perichondritis and frank chondritis. Healing may occur by epithelial growth inthe early stage with various degrees of subepithelial fibrosis (even without removal of the tube) or by exuberant granulation tissue, which indicates injury to the cricoid cartilage, its nutrient perichondrium, or both. Frank necrosis of the cricoid may then occur in severe cases, resulting in loss of cartilaginous support and collapse of the upper airway. This stage of injury with its proliferativ phase may result in "soft" stenosis and a compromised airway, and is usually managed by tracheotomy. The reparative process may resolve following the establishment of an adequate upper airway, or may progress to the stage of "hard" cicatricial stenosis, which is generally but not inevitably circumferential. This end stage of subglottic injury is responsible for the permanent, crippling airway obstruction that requires long-term by-pass with tracheotomy.

The development sequence of subglottic stenosis secondary to endotracheal intubation in human beings has been reconstructed by the examination of a larg number of autopsy specimens of neonates, children, and adults. However, it is obvious that although these studies contributed significant insight into the process, they have not precisely defined the role of many variables that affect the development of chronic subglottis stenosis. All factors being equal, the duration of intubation is crucial indeed; however, even the definition of prolonged intubation differs in the opinion of several authors, ranging from 48 hours to six days. Hawkins has shown that the neonatal cricoid cartilage is hypercellular with a scant, gel-like matrix rendering the cartilage more pliable and yielding to pressure. With growth, the matrix increases and becomes less hydrated, more fibrous, more rigid, and considerably more susceptible to pressure-related injury. This is substantiated by the fact that neonates can tolerate intubation for a longer period of time (measured in weeks) compared with older infants and children (measured in days). In spite of these favorable conditions, signs of subglottic injury in the larynges of 15 of 16 neonates who were intubated for six days or longer were found at autopsy.

The role of infection in the development of a stenosing lesion, which is simultaneously a destructive and a reparative process, was recently investigated by Saski et al using dogs. They concluded that tracheotomy-related infection of a mucosal injury of the subglottic lesion can progress to chondritis (and later stenosis) that may be prevented by the administration of systemic antibiotics or by meticulous stomal care. These experimental findings are in agreement with those of Strong and McGovern et al, who reported that subglottic stenosis is twice as common in children when intubation is followed by tracheotomy. The latter studies, although retrospective and uncontrolled, were substantiated and may have far-reaching consequences in the management of pediatric respiratory insufficiency, since tracheotomy is almost always carried out when extubation is difficult or when, using a vague or arbitrary definition of time span, intubation is considered to be too prolonged.

Air hunger is almost a unique feature of subglottic stenosis; it is manifested by low-pitched stridor that is inspiratory but could be biphasic. Although hoarseness is not a symptom of the pure subglottic lesion, a weak voice, caused by the small expiratory volume reaching the vocal chords, may be present.

Although the radiation dosage is relatively high, xeroradiography is the current radiologic technique of choice in the evaluation of chronic airway disease in the young child, as the tissue-air interface is better defined than in tomography. The best technique is the endoscopic evaluation, although it is still deficient in respect to defining the thickness of the stenosis, its extent, and the adequacy of airway diameter.

Management of Chronic Subglottic Stenosis

The management of chronic subglottic stenosis secondary to prolonged endotracheal intubation is both controversial and frustrating, as manifested by the wide spectrum of therapeutic procedures proposed in the last decade, all of which have been far from ideal, especially in children. The basic principle in treating this form of airway obstruction is to bypass the obstruction with tracheotomy, then launch a therapeutic attack on the stenotic area. The method of treating stenosis with dilatation has been applied to acquired subglottic stenosis, since the results of this therapeutic modality in congenital subglottic stenosis were encouraging. The application of this modality to acquired stenosis secondary to intubation appears to be costly. Fearon and Cotton reported a 24 per cent mortality rate from causes directly related to tracheotomy and 26 per cent of the children in their series still had a tracheotomy several years later. Similar results have been reported by Hollinger et al, who had the largest series of patients with acquired subglottic stenosis: 11 per cent died and 28 per cent still had a tracheotomy after several years of treatment. A negative impression regarding the value of dilatation is also conveyed by others; however, it appears that in mild cases early recognition of the possibility of developing stenosis (granulations, ulcerations, or difficult extubation) improves the chances that dilatation will be successful.

Since the basic principle in treating chronic subglottic stenosis is to control and reduce fibrosis, systemic administration of steroids has been tried, with varying results: Skolnick believes that steroids are not efficacious, while Hawkins employed them successfully after every dilatation. Intralesional injection of steroids has been reported by several authors. This method was used in a small number of patients "early" in the disease process with conflicting results; it requires weekly injections under general anesthesia for about three months.

Successful laser excision of granulations, subglottic webs, and severe cicatricial stenosis in a small number of children has recently been reported. It has been our experience in a small number of children that laser excision is useful in thin, web-like, not extensive narrowing of the subglottic stenosis; in severe cases, this treatment modality alone cannot establish an adequate airway. If this method proves successful in a controlled study, it will offer many advantages: (1) precise control, (2) protection of surrounding structures with preservation ofanatomy and juncture, (3) preservation of the airway without tracheotomy in some patients, (4) rapid, excellent healing with minimal stricture formation or functional disturbance, (5) the possibility of repeated surgery without trauma and edema, and (6) superior cost effectiveness.

Several other surgical methods have been described for treatment of the stenosis in its cicatricial phase. Prolonged stenting for at least six weeks was successful in six of seven children between the ages of two and nine years, with one death related to the management. All patients in this series appear to have sustained trauma to the larynx secondary to intubation, although this was not specifically stated.

When laryngotracheal cartilages are split with stenting or lumen maintainers, the lower thyroid cartilage as well as several tracheal rings are divided anteriorly; the cricoid is split anteriorly and, sometimes, posteriorly. This allows expansion of the stenotic area (with or without "coring out" the stenosis and with or without grafting) that is maintained by either a stent or by homologous bone (hyoid), cartilage grafts, and composite flaps. These methods, with minor variations, have yielded an airway that is 60 to 90 per cent "adequate" so that decannulation was possible two to four months after the operation. In one series in which decannulation was successful in 70 per cent of patients who underwent an operation, a roughly comparable group of patients was not operated on; however, most reports have lacked a control group. In addition, the studies have been retrospective and have not provided a clear definition of the disease or information (such as extent, thickness, and duration) on the condition of the stenosis.

Future Goals

In spite of the voluminous literature on acquired subglottic stenosis, the basic sequential development of this injury and reparative process has not been delineated. Is perichondrium or cartilage injury essential to trigger the cicatricial stenosis, and how important is infection and/or tracheotomy? What is the contribution of the general hemodynamic status of the child to the presumed ischemic necrosis of the cricoid?

Regarding the precise diagnosis of the vertical extent of the stenosis, which is crucial to the treatment and outcome, we still fall short of the optimum. As yet no satisfactory method exists to determine the character of the keystone of the problem, the cricoid ring, except by exposing it through an external surgical approach.

There is an urgent need for a unified method to report pretreatment and treatment outcomes and to develop objective methods for demonstrating the subglottic area and the stenosis. A clinical tool to measure the fundamental status of this area will certainly facilitate this objectivity and will help to define a true subglottic stenosis.

These and many other questions cannot be answered in a clinical setting because of the large number of confounding variables. Thus, the development of an animal model of chronic subglottic stenosis secondary to prolonged intubation should be a top priority.