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Laryngotracheal stenosis in children

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Abstract The diagnosis of laryngotracheal stenosis should be suspected in children with stridor, feeding difficulties, or atypical croup. Only half of the children with congenital laryngotracheal stenosis require tracheotomy, and many of these children can be decannulated following uncomplicated surgical therapy. In contrast, tracheotomy-dependent patients with acquired laryngotracheal stenosis require more extensive surgical intervention, which should be carried out as early as possible to provide the best opportunity for developing normal oral communication.

Key words Laryngotracheal stenosis · Intubation · Tracheotomy · Airway obstruction · Management

Introduction

Laryngotracheal stenosis is a congenital or acquired narrowing of the airway, representing a continuum of disease that may affect the glottis, subglottis, and/or trachea. Previously, the term "subglottic stenosis" was often used (inaccurately) as an umbrella term to describe stenosis occurring anywhere in the airway because the subglottis was the most frequent site of stenosis. Currently, rod lens telescopes are available, allowing the experienced endoscopist to describe precisely the site and type of airway pathology. This review will address the etiology, diagnosis, and management of laryngotracheal stenosis.

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Diagnosis

Diagnosis entails a complete history and physical examination, radiologic evaluation, and endoscopy. Presenting symptoms of laryngotracheal stenosis include stridor, restlessness, irritability, dyspnea, tachypnea, apnea, and cyanosis. Glottic stenosis may affect vocal cord mobility and present clinically as a weak, hoarse cry, aphonia, or aspiration with recurrent pneumonia. Respiratory distress with feeding may result in failure to thrive. Permanent pectus excavatum may result from long-standing increased negative pleural pressures during inspiration coupled with the increased compliance of the chest wall of the young child. The diagnosis may be confounded by concurrent pulmonary, cardiac, or neurologic disease.

Mild to moderate laryngotracheal stenosis may be occult until the child is 1–3 months of age, when the infant becomes capable of greater physical activity that requires increased ventilation [7]. In the majority of cases, the child with laryngotracheal stenosis will present with croup. "Atypical" croup refers to (1) multiple episodes of croup in the first year of life; (2) multiple hospital admissions for croup; (3) failure of prompt response to appropriate med-

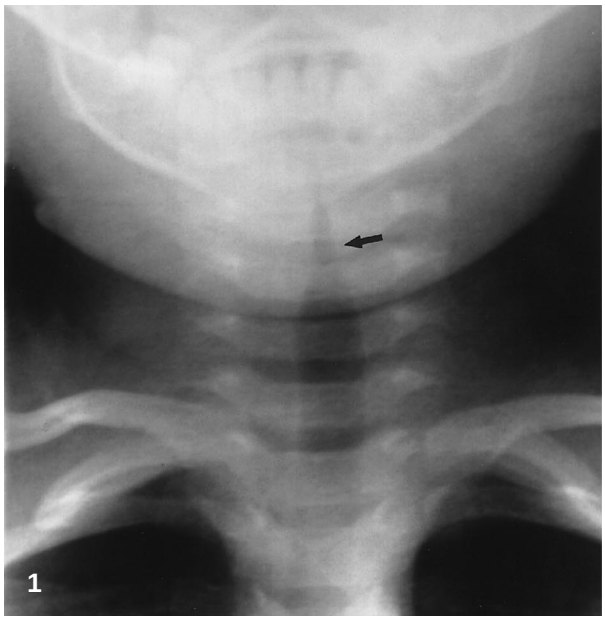
Fig. 1 Anteroposterior radiograph of airway demonstrating moderate subglottic stenosis with *(arrow)* absence of normal subglottic "shouldering" in a 22-month-old girl ▶

Fig. 2 a, b Airway fluoroscopy of a 4-year-old boy with congenital heart disease and tracheal stenosis involving *(arrow)* limited segment of thoracic trachea. **a** Inspiratory view; **b** dynamic component evident on expiratory view

Fig. 3 Radiograph obtained immediately after birth of male infant demonstrating laryngeal atresia with tracheoesophageal fistula. Note endotracheal tube tip *(black arrow)* contacting atresia plate with upper esophagus *(white arrows)* distended by air

Fig. 4 Acquired circumferential laryngotracheal stenosis of moderate severity. The scar in this patient was mainly a soft (membranous) stenosis

Fig. 5 Congenital laryngotracheal stenosis. The stenosis is usually anterior, involving the immediate subglottic area



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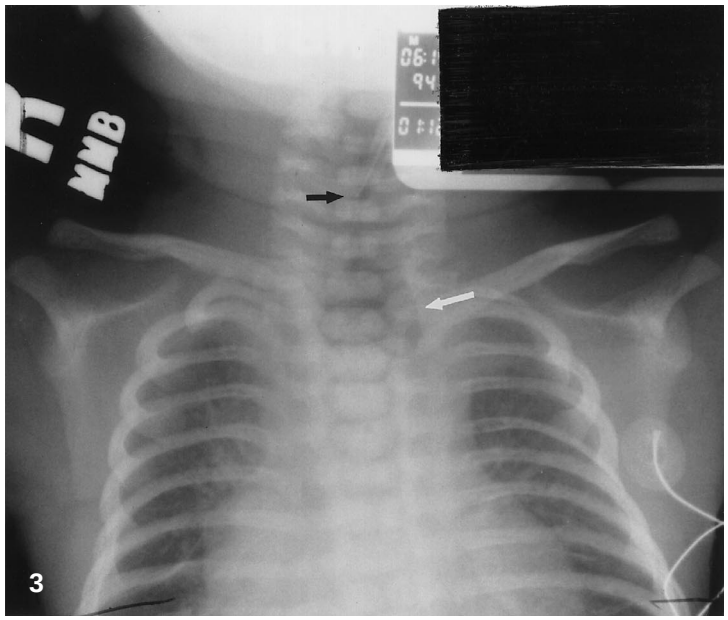

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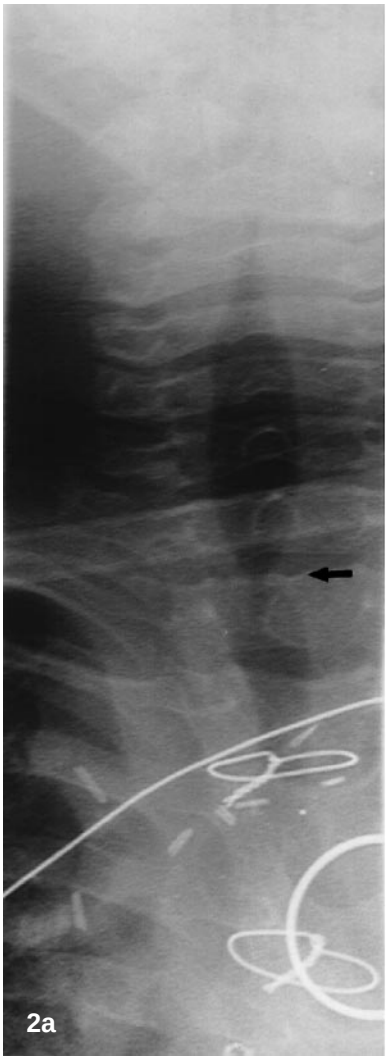

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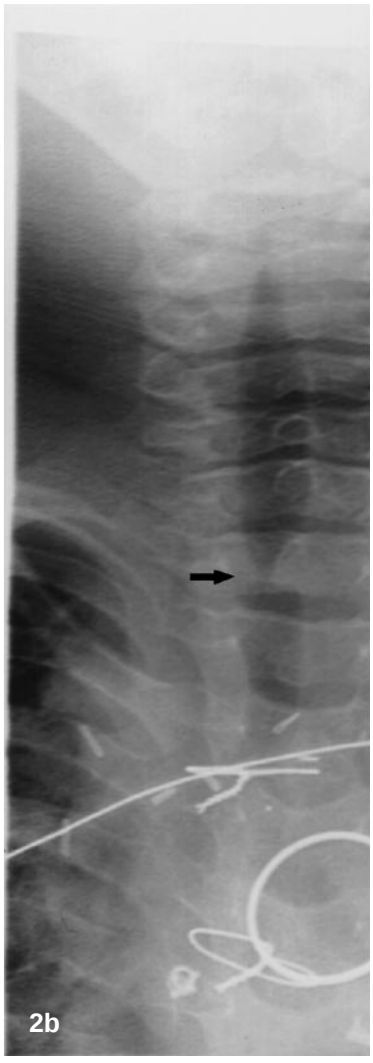

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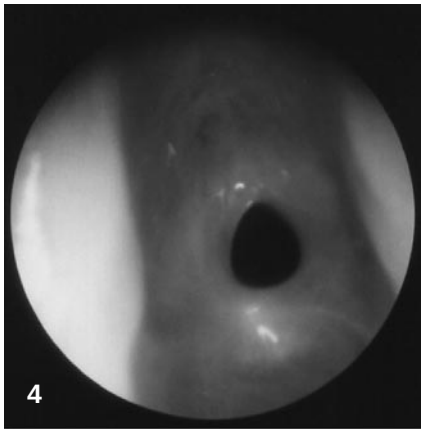

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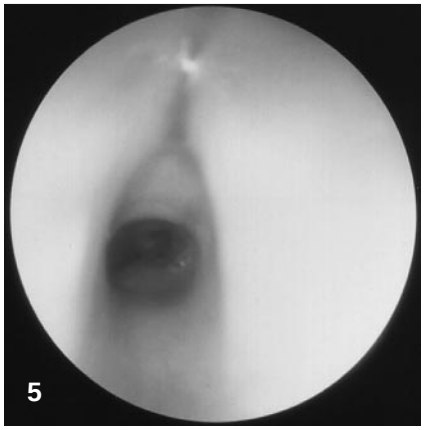

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ical management; and (4) croup requiring endotracheal intubation.

Children with severe laryngotracheal stenosis have significant respiratory distress that persists in the absence of upper respiratory tract infection. Severe congenital laryngotracheal stenosis may be symptomatic at birth. The latent period for acquired laryngotracheal stenosis is generally 2–4 weeks from the time of airway injury, and the degree of respiratory distress is often progressive over time.

High kilovoltage anteroposterior and lateral airway radiographs (Fig. 1) should be obtained unless the severity of respiratory distress precludes radiologic evaluation. If true inspiratory and expiratory views are difficult to obtain, airway fluoroscopy may be useful (Fig. 2). Although barium esophagram may be indicated to assess feeding problems, it rarely rules out external compression of the trachea. Computed tomography may be useful in assessing the length of the stenotic segment. However, as sedation is contraindicated in the child with a compromised airway, this latter study may be difficult to obtain. Preoperative voice analysis and pulmonary function tests are useful in older children [23].

Flexible fiberoptic endoscopy is performed to assess supraglottic and glottic pathology. On occasion, the subglottis can be visualized with flexible endoscopic examination. Congenital laryngeal webs may be partial or complete (laryngeal atresia) and are characterized by a firm fibrous membrane connecting the vocal cords (Fig. 3). An immobile vocal cord may be evidence of posterior glottic stenosis with cricoarytenoid joint fixation [1], and laryngeal electromyography should be performed to assess vocal cord innervation.

At present, the standard for diagnosis is direct laryngoscopy and bronchoscopy under general anesthesia. The outer diameter of the largest bronchoscope that can pass through the stenosis should be noted. Congenital subglottic stenosis is defined as a subglottic diameter of 4 mm or less in a full-term neonate, or a diameter less than 3.5 mm in a premature neonate [21]. The length and location of the stenotic segment are noted. The use of the rod lens telescope without the bronchoscope allows dynamic assessment of the cartilaginous support.

Membranous subglottic stenosis (sometimes called "soft tissue stenosis") may be due to submucosal fibrosis, submucous gland hyperplasia, or granulation tissue. Most commonly, the stenosis is circumferential and symmetric, with the smallest diameter 2–3 mm below the vocal cords (Fig. 4). Frequently, posterior glottic stenosis is continuous with stenosis of the posterior subglottis [12]. Cartilaginous stenosis presents as a deformity of the cricoid cartilage projecting into the airway lumen from the posterior, anterior, or lateral walls [13]. A small cricoid of otherwise normal shape will present as a symmetric stenosis. Causes of asymmetric congenital cricoid narrowing include an elliptical or oval-shaped cricoid, isolated enlargement of either the anterior or posterior cricoid lamina, or incomplete laryngeal cleft. A trapped first tracheal ring is a type of cartilaginous stenosis characterized by a nondistensible, complete tracheal ring [8].

Etiology and pathophysiology

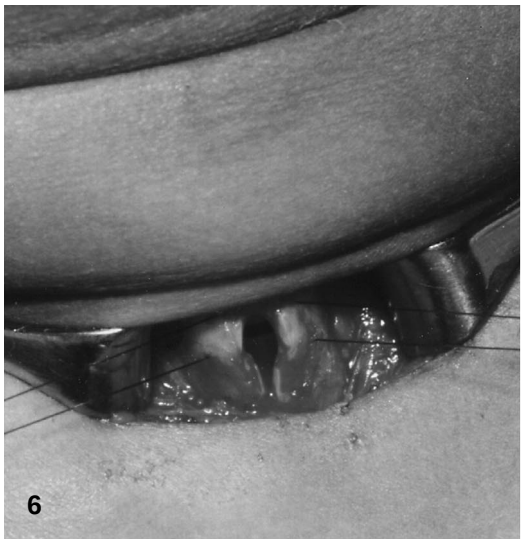
Laryngotracheal stenosis is considered congenital when there is no previous documentation of a normal airway, and risk factors for acquired stenosis are absent (Fig. 5). The most common cause of acquired stenosis is endotracheal intubation, accounting for approximately 90% of mature laryngotracheal stenoses [12]. Histopathologic studies of the airway have shown that even brief periods of endotracheal intubation can cause damage to the thin respiratory epithelium of the subglottis [11]. The subglottis is the narrowest region of the airway in children, and the cricoid cartilage is the only complete circumferential cartilaginous ring in the airway. The loose areolar tissue of the submucosa readily develops edema, which extends inward at the expense of the airway lumen.

Endotracheal intubation causes mucosal compression when the pressure from the tube exceeds the capillary pressure of the mucosa [10, 12]. Mucosal edema, ischemia, and ulceration lead to perichondritis and chondritis. Healing occurs by secondary intention with granulation tissue proliferation and deposition of fibrous tissue. A weakened cartilage framework and a firm scar narrowing the lumen of the airway result.

External compression may contribute to the loss of cartilage support, which often persists long after surgical relief of the compression. The differential diagnosis includes vascular anomalies, such as innominate artery compression or double aortic arch, and congenital masses, such as bronchogenic cyst, duplication cyst, teratoma, cystic hygroma, and hemangioma. Systemic illnesses such as sepsis, dehydration, anemia, avitaminosis, hypoxia, neutropenia, and immunosuppression also influence the development of laryngotracheal stenosis through alteration of perfusion, nutrition, and resistance to infection.

The most important risk factor for the development of laryngotracheal stenosis is the duration of intubation. Although 5–10 days of intubation are generally considered to be an acceptable length of time for intubation in adults, there is no definite "safe" period in children. In general, premature infants may be intubated for longer periods, since airway cartilages in the neonate are more yielding and pliable [10]. The size of the tube should allow an air leak at an inspiratory pressure of 20 cm H_2O. The rate of laryngotracheal stenosis has been reported as very low when small (2.5 mm) endotracheal tubes are used in infants weighing less than 2500 g [4]. However, because of the intensive nursing care required to keep the tubes patent, larger tubes are often used in North America. The type of endotracheal tube is rarely debated, now that Silastic and polyvinyl chloride tubes without cuffs are used almost uniformly.

Laryngotracheal stenosis may result from external or surgical trauma through a similar mechanism of edema, granulation, and fibrosis. Severe glottic stenosis may result from blunt laryngeal trauma, particularly when the injury is not repaired promptly, and the stenosis often extends to the supraglottis and subglottis. Caustic ingestion



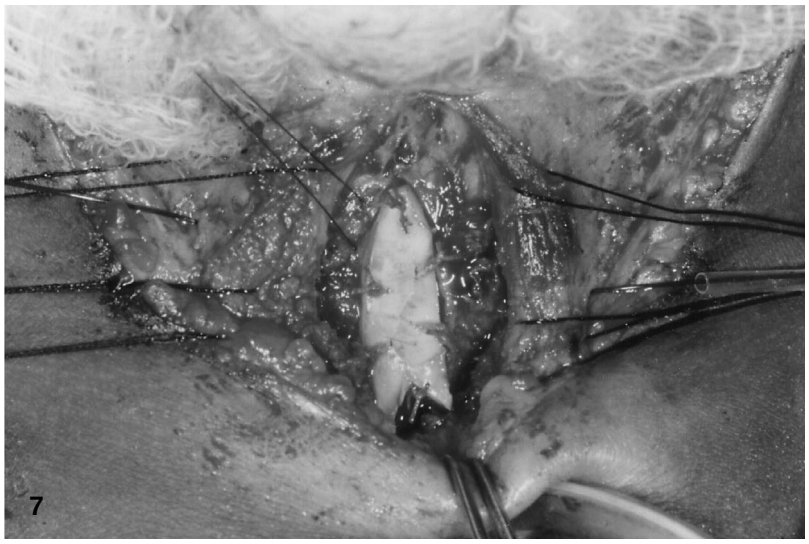


Fig. 6 Anterior cricoid split procedure. Note the endotracheal tube in the subglottic lumen and the distraction of the segments of the cricoid cartilage after the anterior split

Fig. 7 Anterior costal cartilage graft (modified boat shape) used to reconstruct anterior laryngotracheal stenosis

and thermal burns can cause severe mucosal damage that results in stenosis. Glottic and subglottic stenosis may result from a high tracheotomy, cricothyrotomy, or endoscopic laser procedures [12, 17]. Anterior glottic stenosis is most commonly a consequence of endoscopic procedures involving both true vocal cords at one session [17]. A 20% incidence of subglottic stenosis has reported following laser treatment of subglottic hemangioma [15].

Although many patients with laryngotracheal stenosis have demonstrable physiologic gastroesophageal reflux (GER), there are no accepted clinical criteria to confirm the presence of pathologic GER or reflux disease. A prospective study of patients undergoing laryngotracheal reconstruction revealed no differences in outcome whether or not patients underwent preoperative testing for GER, had evidence of GER, or were treated for GER [25]. Surgical technique (discussed below) rather than the presence or treatment of GER is likely the most important factor in successful treatment of laryngotracheal stenosis.

Excessive suctioning, multiple intubations, and tube changes should be avoided in order to minimize trauma to the airway mucosa. A child with a history of laryngotracheal stenosis may sustain significant re-injury to the airway while undergoing a brief general anesthetic if the anesthesiologist is not informed of the need to use a smaller tube. Adequate sedation of the intubated patient will reduce the shearing action of the tube caused by swallowing or volitional motion of the patient. Meticulous hygiene is critical, as secondary bacterial infection increases inflammation and the deposition of scar tissue [14]. Concomitant use of a nasogastric tube increases the risk of secondary infection because of local reaction to the foreign body and increased stasis of secretions.

The reported incidence of laryngotracheal stenosis in intubated patients is approximately 1–5% [6, 10]. In the 1960s, prolonged endotracheal intubation of neonates became widely accepted as an alternative to tracheotomy [Brandstater 1962, unpublished work]. During this time, the incidence of laryngotracheal stenosis was 12–20% [9]. The reduction in the incidence over the last 20–30 years is likely due to improved education of health care personnel leading to increased awareness of the factors discussed above. However, the exact incidence rate of laryngotracheal stenosis is unknown. Many children are initially asymptomatic and will not be diagnosed until they are referred for rigid endoscopy much later in life. In addition, severely ill children may die of their primary illness before laryngotracheal stenosis is recognized. Finally, although staging systems for laryngotracheal stenosis have been described, none are universally applicable and useful. The Cotton staging system is useful for mature, firm, circumferential stenosis confined to the subglottis [7].

Management

Unlike acquired stenoses, congenital subglottic stenoses carry an excellent prognosis. Respiratory distress associated with upper respiratory tract infections can be treated with aggressive medical management. In mild cases, the airway will increase in diameter as the child grows, and surgery will not be necessary. Tracheotomy is required to relieve airway obstruction in less than 50% of cases of congenital laryngotracheal stenosis. If reconstructive surgery is postponed or declined, most patients remain tracheotomy-dependent for 2–5 years [12].

Cases of increased severity require surgical intervention. In our experience, the majority of patients with acquired laryngotracheal stenosis or severe congenital stenosis require tracheotomy. Given that the mortality of tracheotomy is still 2–5% per child per year, there is no role for “wait-and-see” management in acquired or severe laryngotracheal stenosis. In general, the mortality of tra-

cheotomy is particularly related to the status of the residual airway above the tracheotomy tube. However, surgical correction of the airway is contraindicated in the patient who would still require a tracheotomy for mechanical ventilation, supplemental oxygen, or management of other pulmonary or neurologic disease.

Subglottic cysts and very thin, limited regions of stenosis may be amenable to endoscopic resection with the carbon dioxide laser [16]. However, stenosis may become dramatically worse after laser therapy. Circumferential laser treatment will often lead to cicatricial contracture of the airway lumen. We believe that the failure of laser therapy should not be considered a prerequisite for laryngotracheal reconstruction, as many stenoses will not be amenable to such treatment at any time.

Anterior cricoid split was first described by Cotton and Seid [8] for the treatment of acquired subglottic stenosis. The indications were expanded to include patients with congenital subglottic stenosis that would otherwise require tracheotomy or endotracheal intubation [9]. Anterior cricoid split is indicated in children with a small, normally shaped cricoid ring, extensive submucosal fibrosis, or for decompression of subglottic cysts.

After induction of general anesthesia with endotracheal intubation, the laryngotracheal complex is exposed through a cervical incision. The cricoid cartilage and the first and second tracheal rings are split in the midline anteriorly to allow expansion of the cricoid ring (Fig. 6). The endotracheal tube is replaced as a stent for 7 days. Severe congenital stenosis, marked deformity of the cricoid cartilage, or failure of the anterior cricoid split procedure are indications for tracheotomy and laryngotracheal expansion surgery.

The concept of laryngotracheal expansion surgery is to divide an existing scar and distract the edges of a stenotic site with an interposed graft to widen the airway lumen [7, 12]. The scar is not resected, since creating a surface denuded of mucosa will inevitably heal with recurrent formation of scar. The operation is customized for each patient, involving choices regarding the graft (anterior, posterior, or both), the stent (discussed below), the type of cricoid incision (anterior, posterior, or four-quadrant split), and the need for laryngofissure.

The autogenous costal cartilage graft is the most useful for laryngotracheal reconstruction [5]. The advantages of cartilage include less resorption than bone grafts, viability without a vascular pedicle, and ease of carving. Donor site morbidity is minimal. Unlike bone grafts, cartilage grafts will maintain their bulk despite a lack of functional stress. The graft is harvested with the inner perichondrium in situ and placed with the perichondrium facing the lumen as a barrier to infection [22]. The boat-shaped graft used for posterior grafts is modified for anterior grafts (Fig. 7) to include outer flanges in order to prevent prolapse into the lumen, gain more distraction, and avoid the need to bevel the graft.

Stents are used to restore distorted anatomy, maintain a lumen, counteract keloid formation, and create a stable framework in the absence of cartilage support. The choice of stent and duration of stenting depends on the purpose of the stent. If the stent is placed only to keep repaired areas in place (i.e., a posterior costal cartilage graft), only a short duration of stenting is required. Conversely, months of stenting are required if the function of the stent is to counteract scar formation. Stent materials include Teflon (Aboulker), Silastic (Montgomery stent and “Swiss roll” stent), polyvinyl chloride (endotracheal tube), and a finger cot stent. Rubber stents should not be used. The Aboulker stent is most commonly used, as Teflon is the least reactive stent material. Oral antibiotics are administered while the stent is in place to prevent infection and colonization of the granulation tissue.

The long Aboulker stent extends inferiorly from the tip of the tracheotomy tube to the arytenoid cartilages superiorly [19]. Since the long stent is wired to the tracheotomy tube, the outer cannula of the tracheotomy tube cannot be changed for the duration of stenting. The short Aboulker stent is used if the stenosis is confined to the glottis and if the duration of stenting will be short (4–5 weeks). The short stent is secured with a suture, since the inferior aspect of the short stent is above the tracheotomy tube. The disadvantage of the short stent is the potential for granulation tissue formation between the stent and the tracheotomy tube. The average time to decannulation after stent removal is 4 weeks. The position of the stent is monitored with flexible laryngoscopy for stent displacement or granulation tissue. Glottic stenosis may recur if the stent does not extend superiorly enough to distract the vocal cords. Complications of stenting include granulation tissue with subsequent stenosis, wound infection, stent breakage, and stent migration.

Anterior cricoid split with anterior costal cartilage grafting is indicated for anterior tracheal wall collapse or isolated anterior subglottic stenosis. The anterior costal cartilage graft is placed such that it extends inferiorly to repair the previous tracheotomy site. “One-stage laryngotracheoplasty” refers to procedures performed on these relatively mild cases of laryngotracheal stenosis that do not require stenting for longer than 1 week.

Indications for posterior costal cartilage grafting include posterior glottic and subglottic stenosis, complete or circumferential stenosis, vocal cord fixation, and bilateral subglottic shelves. In order to place posterior grafts, the cricoid must be split in the midline both anteriorly and posteriorly; therefore, a rigid posterior cricoid lamina must be present. Laryngofissure may be necessary in the presence of posterior glottic stenosis in order to visualize the superior extent of the stenosis. An anterior graft is used in addition to the posterior graft if the anterior cricoid cannot be closed around the stent.

The indications for a four-quadrant cricoid split without grafting are continuous, complete scarring from glottis to trachea, severe distortion of the anatomy, or the absence of a stable cartilage framework. The cricoid cartilage is split anteriorly, posteriorly, and laterally on both sides. A long duration (i.e., 6 months) of stenting is required.

Resection of the stenotic segment with primary end-to-end anastomosis is indicated for isolated subglottic or up-

per tracheal stenosis, provided that a normal lumen extends at least 10 mm below the glottis. The vertical length of the stenosis must be such that after resection, the cricoid and tracheal ends can be approximated without tension. Laryngeal release will provide an extra 1–3 cm of mobilization. Stenting is not required, and the tracheotomy tube usually can be removed by 4 weeks postoperatively. Partial cardiopulmonary bypass may be necessary to accomplish the repair.

Outcome of treatment

The ultimate goal of laryngotracheal reconstruction is decannulation. Restoration of the normal functions of the airway at the earliest possible age will allow optimal development of speech and articulation. Success rates of 80–90% have been reported [20]. Although a functional voice can be restored in most patients, persistent voice disorders are common [3]. The postoperative voice is characterized by lower than optimal pitch and a restricted pitch range [24]. As surgical management must be individualized, comparison of results requires consideration of (1) the etiology of stenosis; (2) the number of previous (failed) endoscopic procedures; (3) the location and extent of scar and the degree of anatomic disruption in the entire airway, including the glottis; (4) the mobility of the vocal cords; (5) the stability of the laryngotracheal framework and the integrity of cartilaginous support; and (6) the patient's general medical condition.

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