Congenital Tracheal Stenosis

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Overview

Compromised tracheal airflow in neonates is a rare but potentially life-threatening occurrence. Management of these infants is always subservient to the etiology of the tracheal compromise, and this compromise may be a consequence of any one of the following clinical scenarios: (1) extrinsic compression of the trachea (e.g., vascular compression); (2) a problem of the tracheal exoskeleton without the presence of stenosis (e.g., tracheomalacia); (3) a problem of the tracheal exoskeleton with stenosis (e.g., complete tracheal rings (CTRs), sleeve trachea, absent tracheal rings); and (4) an intraluminal obstruction with an adequate exoskeleton (e.g., congenital tracheal web). We will limit our discussion to an overview of the etiology, clinical presentation, and management of the three conditions that fall within the classification of problems of the tracheal exoskeleton with stenosis. We will also present a detailed description of the slide tracheoplasty procedure, which is now considered the operation of choice for the management of these conditions.

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Epidemiology, Etiology, and Pathogenesis

Congenital tracheal stenosis encompasses a wide range of anomalies resulting from aberrant embryogenesis of the respiratory system. It is infrequently seen and has an estimated incidence of 1 in 64,500 births [1]—representing only 0.3–1 % of all laryngotracheal stenoses [2]. A frequently cited article by Hoffer et al. [3] describes the associated embryologic processes and timing of the developmental aberrations that lead to the various manifestations of tracheal narrowing. As these authors point out, lung development begins during weeks 3 and 4 of gestation. The developing lung buds give rise to the trachea, infraglottis, and the glottic opening. By week 8 of gestation, the mesenchymal rudiments of the tracheal cartilages are present. During weeks 9 and 10, the cartilages form fibroelastic tissue, and smooth muscle is incorporated into the trachea. Hoffer and his colleagues postulate that (1) aberrant development in week 4 of gestation affects the developing respiratory and hepatic primordia and cause the more severe forms of the disease, which are associated with anomalies such as heart and skeletal malformations and that (2) aberrant development between gestational weeks 8 and 10 is confined to the developing cartilages and their supporting tissues. Abnormalities during this second critical time point likely result in less severe stenosis with fewer associated anomalies.

Clinical Presentation

Infants with compromised tracheal airflow classically present with biphasic stridor within the first weeks of life. Over the first few months of life, symptoms exacerbate, with retractions, dying spells, and severe deterioration occurring 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; this pattern is referred to as "washing machine breathing."

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Because the growth of the trachea is not commensurate with the growth of the infant over the first few months of life, decompensation frequently occurs around 4 months of age [4]. Important to note, however, even a neonate with a life-threatening tracheal stenosis may have surprisingly few symptoms.

Complete Tracheal Rings

CTRs are the most common etiology of congenital tracheal stenosis. In a normal trachea, luminal support is provided by C-shaped cartilaginous rings with a posterior sheet of muscle (the trachealis) completing the ring (Fig. 1a–c). In the infant with CTRs, the cartilaginous rings are circular and may affect varying lengths of the trachea. In addition, the trachealis muscle is absent. The diameter of the affected tracheal segment is always smaller than the trachea above that segment; however, the degree of stenosis may range from mild to severe. A classification system developed by Speggiorin et al. [5] delineates a number of recognizable morphological patterns. Of these patterns, the most frequently seen are: (1) CTRs that are of reasonable size proximally, but that cone down to a small distal ring close to the carina; (2) the "stovepipe" airway with a long segment of CTRs of similar diameter; (3) a short-segment stenosis, often in the mid-trachea; and (4) CTRs associated with a high tracheal (or pig) bronchus. More than 75 % of patients also have other congenital anomalies that may be severe. A relatively recent study conducted at Cincinnati Children's Hospital indicated that 60 % of patients with CTRs had cardiovascular abnormalities, particularly a pulmonary artery sling (21 %) [6].

Diagnostic Workup

The initial evaluation should include plain airway films, as these may indicate that congenital tracheal narrowing is present. Although imaging studies may be very useful, bronchoscopic evaluation remains the gold standard for definitive assessment. This should be performed with extreme care, as instrumentation in an area of stenosis may cause enough swelling to convert a narrow airway to a critical airway, necessitating abrupt intervention. If the smallest bronchoscope available (whether rigid or flexible), cannot be easily passed through the area of stenosis, it is better to identify the proximal extent of the stenosis without fully evaluating the distal airway. Good communication with anesthesia colleagues is essential, and to facilitate the bronchoscopy, the child should be spontaneously breathing. We recommend the use of sevoflurane and propofol; steroid administration is also advisable. The trachea is then suctioned with a soft 6 Fr catheter and the patient is preoxygenated. Next, a further propofol bolus is delivered to temporarily halt respiratory efforts during the bronchoscopy.

Ideally, the aims of endoscopic evaluation are: to confirm the diagnosis of tracheal stenosis; establish whether this is due to CTRs; estimate the size of the smallest ring; estimate the percentage of the airway involved with CTRs as well as the position of the rings within the trachea; and evaluate the bronchial anatomy.

In patients with distal tracheal stenosis, an adequate evaluation cannot be made with a ventilating bronchoscope, as even the smallest ventilating bronchoscope is too large for most complete rings. The Hopkins rod telescope (removed from the bronchoscope) should thus be introduced into the airway to assess the stenosis. The initial bronchoscopic view is often sufficient to establish the diagnosis, thereby avoiding the risk of airway edema.

Estimating the size of the airway is valuable. In a normal full-term neonate, the narrowest point of the airway is at the cricoid, and this should measure 4.5–5.5 mm. Subglottic stenosis is defined as an airway diameter of <4.0 mm (by comparison, a 2.5 mm endotracheal tube has an outer diameter of 3.6 mm). Although the trachea should be of greater capacity than the cricoid, the airway in patients with CTRs may be too small to easily permit safe passage of any form of instrumentation. For comparative purposes, the smallest Storz 2.5

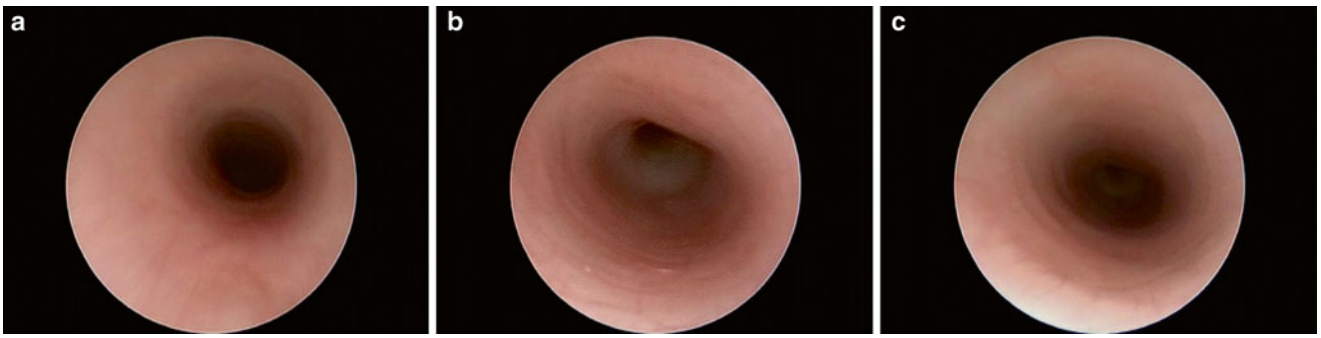

Fig. 1 Endoscopic views of a child with long-segment complete tracheal rings. (**a**) View showing beginning of rings; (**b**) view showing mid-section of rings; (**c**) view showing distal segment of rings

ventilating bronchoscope has an outer diameter (OD) of 3.7 mm. The smallest 20017 Hopkins rod telescope (as found in the Storz 2.5 ventilating bronchoscope) has an OD of 1.9 mm.

Because 50 % of children with CTRs 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 (2.0 mm inner diameter [ID]; 2.9 mm OD) and the smallest tracheotomy tube (2.5 mm ID; 3.9 mm OD) cannot pass through the stenotic segment without severe damage to mucosa or tracheal rupture. As the stenosis usually extends to the carina, bypassing the stenosis risks bronchial intubation. This may leave extracorporeal membrane oxygenation (ECMO) as the only viable alternative for stabilizing the child in the event of decompensation following bronchoscopy. In an effort to avoid ECMO in a child who is decompensating and poorly ventilating, intubation proximal to the complete rings is preferable. The endotracheal tube should be sized to the cricoid, with the Murphy eye just below vocal fold level. Given that it is unusual for the proximal two tracheal rings to be complete, shallow intubation is achievable in most children with CTRs. A nasal intubation to allow tube stabilization (as it is so shallow) is advisable. Ventilation requires a long inspiratory phase and an even longer expiratory phase to allow air to pass the stenosis. Higher than typical ventilator pressures may be tolerated, as the stenosis ensures that the lungs are not exposed to the same pressures as the subglottis. Maintenance of high humidity levels is crucial, as mucus accumulation may be lethal and is often heralded by rising CO_2 rather than low oxygen saturation. In a crisis, 1 mL of 1:10,000 epinephrine delivered down the endotracheal tube may assist ventilation. If possible, extubation is desirable, as most children with CTRs maintain ventilation more effectively themselves than on a ventilator. To prevent mucus accumulation, saline may be regularly nebulized if required.

In view of the high proportion of patients with other congenital anomalies, a thorough diagnostic investigation should include a contrast-enhanced computed tomography (CT) scan of the chest with three-dimensional reconstruction, and an echocardiogram. These tests will identify any coexisting cardiovascular pathology, which should be repaired concurrently with the tracheal repair. As mentioned earlier in this chapter, our clinical experience indicates that a pulmonary artery sling is the most common cardiovascular anomaly; intracardiac anomalies and the presence of a persistent left superior vena cava draining to the coronary sinus, with absence of the innominate vein are, however, also frequently seen. Other anomalies may be incidental (e.g., limb or central nervous system anomalies) or also affect the airway. Nearly one third of children with CTRs have a tracheal (or pig) bronchus, and subglottic stenosis is present in approximately 20 % [6]. Rarely, CTRs may coexist with a tracheoesophageal fistula or laryngeal cleft. Pulmonary anomalies may also occur, with subsegmental branching patterns almost universally aberrant, though of limited significance. Pulmonary hypoplasia or agenesis, more commonly affecting the right lung, is more significant, as it may cause mediastinal shift and aortic compression of the already stenotic trachea.

Although most children with CTRs require early tracheal reconstruction, some (10–15 %) have sufficient tracheal growth to avoid the need for reconstruction. A further 10–15 % eventually outgrows their trachea and requires late repair. The recommended surgical technique is the slide tracheoplasty (discussed later in this chapter). This approach yields significantly better results than any other tracheal reconstruction technique and is applicable to all anatomic variants of CTRs [6, 7].

Most children with CTRs have distal tracheal involvement. If the distal one-third of the trachea is involved or if there are coexistent cardiovascular anomalies that require repair, we recommend repair utilizing cardiopulmonary bypass. More than 90 % of children requiring slide tracheoplasty for CTRs fall into this category. If only the upper or mid-trachea is involved, repair may be performed with routine anesthesia through a cervical approach. Hyperextension of an infant's neck over a shoulder roll allows good access to the upper two thirds of the trachea through a cervical approach. Exposure can be further enhanced through a limited upper sternotomy if required.

Sleeve Trachea

Albeit extremely rare, tracheal sleeve is the second most common condition falling within the general classification of problems involving the tracheal exoskeleton with stenosis. In infants with this condition, the trachea consists of a single sheet of cartilage rather than 15–20 separate tracheal rings; this cartilaginous sheet may extend proximally into the cricoid and distally into the bronchi. Sleeve trachea is universally associated with a craniosynostosis syndrome, with Pfeiffer, Crouzon, and Apert syndromes, represented in that order [8]. Nonetheless, only a small number of patients with craniosynostoses have a sleeve trachea, and even fewer have associated tracheal obstruction with the posterior aspects of the tracheal cartilage overlapping, effacing the trachealis muscle (Fig. 2). In these cases, a slide tracheoplasty is still an effective reconstructive option, although technically more challenging than a straightforward repair of CTRs. Of note, most children with sleeve trachea have multiple levels of airway obstruction from the choana to the bronchi. Although a slide tracheoplasty may therefore not prevent the need for a tracheotomy, in some children, it does, however, allow for the safe placement of a tracheotomy.

Absent Tracheal Rings

Although compromised tracheal airflow in exoskeletal problems with stenosis usually results from abnormalities in cartilage structure, an infant may also present with stenosis or collapse resulting from an absence of cartilage [9]. This extremely rare condition generally presents in an otherwise normal child as an isolated segment of trachea (usually just proximal to the carina) that is missing cartilage in a two- to three-ring segment (Fig. 3a, b). Clinical presentation is similar to that in infants with complete tracheal rings; however, bronchoscopically the stenotic segment lacks cartilage and is therefore distensible. While most affected children are otherwise normal, we have seen associated congenital left vocal fold paralysis as well as esophageal atresia.

Management

Endoscopic techniques such as balloon dilation rarely have a role in the treatment of congenital tracheal stenosis. More specifically, they are contraindicated in the management of CTRs, as the risk of tracheal rupture is high. Balloon dilation may, however, have a role after reconstruction if restenosis occurs.

Although a number of approaches for repairing congenital tracheal stenosis have historically been used (i.e., pericardial patch, cartilage grafts, resection, and autografts), these approaches have been replaced by the slide tracheoplasty. This operation marks a clear turning point in the management of tracheal stenosis, as outcomes have dramatically improved since its inception. Although three decades ago the diagnosis of tracheal stenosis carried a mortality rate of 50–80 %, the survival rate currently exceeds 90 %. In essence, a patient's prognosis is now less about the compromised tracheal airway than about concomitant congenital anomalies.

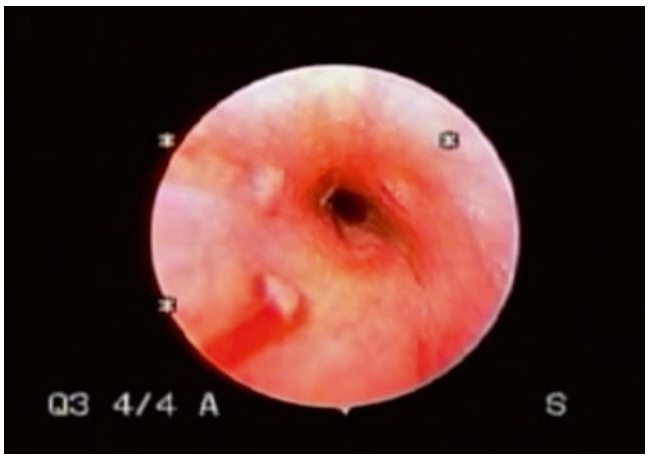

Fig. 2 Endoscopic view of sleeve trachea

Slide Tracheoplasty

Conceived by Goldstraw [10] in the 1980s and popularized by Grillo [11] in the 1990s, the slide tracheoplasty was originally designed as an operation to repair congenital tracheal stenosis caused by CTRs. Although this procedure may be performed using ECMO or jet ventilation, we prefer to use cardiopulmonary bypass to facilitate the repair [6, 7, 12]. Aside from the advantage of not requiring ventilation during the procedure, access is also enhanced, as the lungs and heart may be relatively "deflated." Typically, a sternotomy allows for exposure of the trachea, placement of atrial and aortic cannulae, and repair of any coexisting cardiovascular anomalies. Removal of the carinal lymph nodes facilitates tracheal exposure and mobilization. The extent of the tracheal stenosis is then assessed. The assessment usually requires bronchoscopic examination of the airway while a 30-gauge needle is placed into the trachea from the mediastinal side; this allows the proximal and distal extent of the stenosis to be precisely identified within the chest. The length of the stenosis is then measured and its midpoint is marked. Next, the trachea is transected at or just proximal to the midpoint of the stenosis, with the transection being slightly bevelled (anterior proximal to posterior distal). The transected trachea is

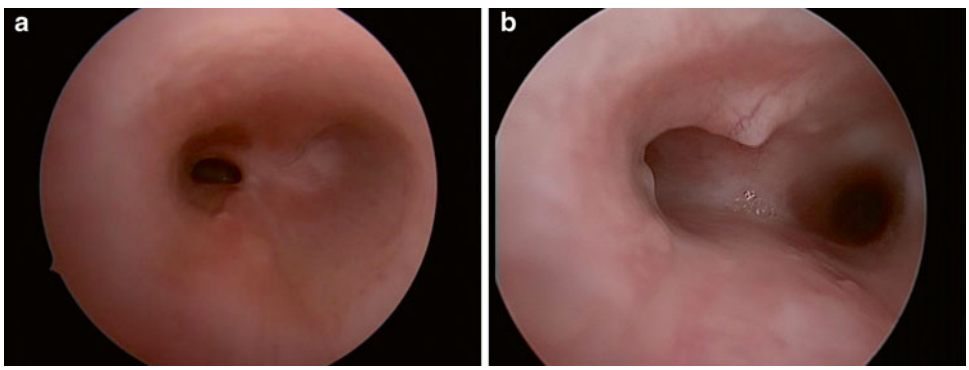

Fig. 3 Pre- and postoperative views of absent cartilage. (**a**) preoperative view; (**b**) postoperative view

mobilized by dissecting free the soft tissue attachments between the trachea and the esophagus of both the proximal and distal segments. Care is taken to preserve some lateral attachments to maintain a blood supply as well as to protect the vagus and recurrent laryngeal nerves. The distal segment is split posteriorly through all complete rings (to carina or down a bronchus if required) and the proximal segment is split anteriorly through the area of stenosis and into normal trachea. At the split, the trachea may be trimmed to round off either end at the transection margins to facilitate the closure. The anastomosis is commenced from distal posterior (carinal) in a running fashion using appropriately sized double-armed PDS sutures (usually 6–0 PDS in infants). Four to six throws of the suture are generally placed at the carina and tightened with a nerve hook. The anastomosis is then continued up the left and right sides of the trachea, with the sutures placed through cartilage and mucosa, therefore being exposed intraluminally. An effort is made to evert the lateral sides of the anastomosis to prevent internal bunching of the anastomotic lines (a "figure 8" trachea). Before the anastomotic suture lines rejoin in the midline at the proximal anterior aspect of the repair, the trachea is suctioned clear. The anastomosis is completed with a single proximal knot being thrown, leak tested (to 35 cm water pressure), and marked with Ligaclips applied to the proximal and distal ends of the anastomosis (for aid in radiographic positioning of the endotracheal tube). Fibrin glue is then applied to the anastomosis. The patient is reintubated and taken off cardiopulmonary bypass, and the chest is closed. At completion of the procedure, the airway is re-evaluated with a flexible bronchoscope to ensure that the repair is adequate and that blood and secretions are suctioned. A 2.8 mm flexible bronchoscope with a suction port can be placed into a 3.5 mm endotracheal tube and still allow for ventilation during the evaluation. If the patient's cardiovascular status permits, extubation is usually achieved within 24 h [12].

The slide tracheoplasty is an extremely versatile procedure [6]. If necessary, it is possible to slide: the entire length of the trachea; own a bronchus; into the anterior cricoid; or past a tracheal (pig) bronchus. The intrathoracic slide tracheoplasty may also be used to repair stenosis associated with absent tracheal rings, a sleeve trachea, or distal tracheoesophageal fistula repair. In addition, it may be used to repair an acquired tracheal stenosis.

The success of the intrathoracic slide tracheoplasty for long- or short-segment tracheal stenosis prompted us to use this technique to manage tracheal stenosis involving the more proximal trachea [13]. The upper half to two thirds of the trachea is accessible through a cervical approach, and the slide tracheoplasty is an effective method of repairing acquired upper tracheal stenosis, often in conjunction with partial resection of the most severely stenotic tracheal segment. The technique is similar to the intrathoracic slide, with the anastomosis commencing at the distal posterior aspect of the repair. In older children with a more proximal stenosis, the risk of developing a "figure 8" trachea is higher, and a temporary intrathoracic silicone stent may be placed for a week or more if required.

A child is typically extubated 24–96 h postoperatively. During this time we try to minimize positive pressure ventilation so as not to endanger the anastomosis. Chest drains are ideally left in place until after the extubation. Preoperatively, some children may present unstable and ventilated; rarely, some may be on ECMO support. In these circumstances, postoperative ECMO may be required. The aim is to establish endotracheal ventilation and remove the child from ECMO as rapidly as possible.

Most complications associated with the slide tracheoplasty have no long-term consequence. The "figure 8" trachea (lateral bunching of the anastomosis) seen in the majority of patients tends to spontaneously resolve over subsequent months and rarely causes obstruction or requires intervention. Although recurrent laryngeal nerve palsy occurs in less than 20 % of patients, it is usually unilateral and transient. Restenosis is rare, as is anastomotic dehiscence. Restenosis is more likely to occur at the proximal end of a slide tracheoplasty when a tracheal bronchus is present at this apex; it may be prevented by extending the slide two or three rings higher, proximal to the tracheal bronchus, into normal trachea.

Multidisciplinary Considerations

In view of the complexity of infants with the conditions described above and the likelihood that they may have serious coexisting conditions, syndromes, and/or anomalies (e.g., bronchial stenosis, lung agenesis or hypoplasia, craniosynostosis, cardiac disease), we cannot overemphasize the importance of a multidisciplinary team approach to management. The success rate at our own institution reflects the close collaborative efforts of a team comprising experienced pediatric subspecialists in otolaryngology, anesthesiology, pulmonology, critical care, and cardiothoracic surgery. The slide tracheoplasty is an operation with a steep learning curve, and research clearly shows that optimal outcomes are achieved through a team approach at a center of excellence [6, 14].

Future Considerations

Although slide tracheoplasty has markedly improved outcomes for children with congenital stenosis, there are circumstances in which this procedure does not provide a stable, viable, long-term solution. The two most common circumstances are (1) children with partial or total tracheal

agenesis and (2) children in whom previous tracheal reconstruction has failed, with resultant damage to the tracheal exoskeleton that may be caused by infection. In these clinical scenarios, the ideal solution is to replace the trachea.

Historically, tracheal homografting was fraught with problems, frequently related to long-segment grafts with delayed reepithelialization. Tracheal homografting with pre-epithelialized or pre-vascularized segments of homograft trachea, zenograft trachea, or synthetic biologic scaffolds seeded with a patient's own respiratory epithelium does, however, offer promise. Pre-epithelialized homograft tracheal replacement remains an area of research that is being refined. Despite highly publicized cases, this is not yet a reality.

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