Editorial

Congenital tracheal stenosis: what should we look at for successful tracheoplasty?

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Provenance: This is a Guest Editorial commissioned by the Section Editor Xicheng Deng (Department of Cardiothoracic Surgery, Hunan Children's Hospital, Changsha, China).

Comment on: Cetrano E, Trezzi M, Secinaro A, *et al.* Bronchial Mismatch as a Predictor of Respiratory Failure After Congenital Tracheal Stenosis Repair. Ann Thorac Surg 2018;105:1264-71.

Submitted Jun 22, 2018. Accepted for publication Jul 03, 2018.

doi: 10.21037/tp.2018.07.01

View this article at: http://dx.doi.org/10.21037/tp.2018.07.01

Congenital tracheal stenosis (CTS) is often described as a rare but life-threatening condition. The reported incidence in one study was 1 in 6,400 live births (1); however, the true incidence is unclear because many infants with CTS may die without a diagnosis.

CTS is characterized by complete cartilaginous rings of the trachea without the membranous portion, resulting in various clinical manifestations. Affected patients are typically classified into three groups: those with minimal or no respiratory symptoms, those with respiratory distress in the neonatal period, and those who present with respiratory symptoms (usually caused by a respiratory infection in late infancy) (2).

More than half of patients with CTS have other congenital anomalies, particularly cardiovascular disease such as left pulmonary artery sling, patent ductus arteriosus, ventricular septal defect, and more complex congenital heart diseases. Moreover, CTS is often associated with pulmonary agenesis or hypoplasia, which also complicates the management of CTS.

Diagnosis is usually made by contrast-enhanced computed tomography (CT) imaging and confirmed by bronchoscopy, which reveals complete cartilaginous tracheal rings. Rigid bronchoscopy under general anesthesia is the gold standard for making the definitive diagnosis and assessment of length and diameter of CTS. However, it is sometimes difficult to completely visualize the stenotic portion. Flexible bronchoscopy is a useful alternative to visualize the narrow segment of the trachea. Because minor mucosal edema at the narrow segment of the trachea could result in severe airway obstruction, great care should be taken when inserting the bronchoscope into the extremely narrow airway. Bronchoscopy should be performed by experienced bronchoscopists with detailed knowledge of CTS.

Respiratory failure, extubation failure, and acute life-threatening events are definitive indications for surgery in patients with CTS. Resection and end-to-end anastomosis for short-segment CTS and slide tracheoplasty for long-segment CTS are currently the procedures of choice. Slide tracheoplasty was first reported by Tsang *et al.* (3) and modified by Grillo (4). This procedure can be performed for all types of CTS (5) with minimal postoperative complications in most patients. However, the mortality rate after slide tracheoplasty is still significant, and the reported mortality rate ranges from 5% to 20% (1,6-8). Postoperative restenosis, tracheal granulation, and tracheobronchial malacia may increase mortality.

Conservative management of CTS is advocated for patients with mild symptoms (8,9). The stenotic trachea may grow, and patients may become free of respiratory symptoms over time. However, common viral upper airway infections deteriorate the airway obstruction. In their systematic review, Ywakim and El-Hakim (8) reported a 5% mortality rate among studies involving conservative treatment.

To optimize the therapeutic strategy for each patient with CTS, it is significantly important to consider the risk factors associated with surgical interventions. A younger age and smaller body weight at the time of the operation, longer duration of cardiopulmonary bypass, longer segment

of stenosis, carina involvement, and longer duration of preoperative and postoperative mechanical ventilation have been shown to have a negative impact on outcomes (5,6,10,11). Hewitt *et al.* (7) reported that preoperative bronchomalacia and bronchial stenosis significantly increased mortality after tracheoplasty.

In a recent paper published in the *Annals of Thoracic Surgery*, Cetrano *et al.* (12) described their single-institute retrospective study of 16 patients who underwent tracheoplasty. The aim of their study was to investigate risk factors associated with postoperative chronic respiratory failure, which was defined as prolonged mechanical ventilation and the need for surgical reintervention. The authors found that bronchial mismatch (BM) [1 - (smaller bronchus/larger bronchus diameter)] of >20% on preoperative CT images was significantly associated with postoperative chronic respiratory failure. Four of five (80%) patients with BM of >20% developed chronic respiratory failure, two required bronchial reconstruction, and two required permanent tracheostomy. The remaining 11 patients, all of whom had BM of <20%, recovered well without chronic respiratory failure (although 3 required multiple tracheal dilatation after tracheoplasty for recurrent stenosis). No mortality had occurred by the time of discharge. In this series, chronic respiratory failure was not influenced by weight, age at surgery, or absolute diameter of the bronchi. BM of >20% was the only factor associated with an increased risk of prolonged mechanical ventilation and bronchial intervention or tracheostomy (P=0.002).

The authors showed that although no rational explanation was provided in this paper, the difference in diameter between the right and left bronchi (rather than the absolute diameter of the bronchi) was associated with respiratory outcomes after tracheoplasty. A difference in diameter between the two bronchi might affect the configuration of the carina (malacia or flattened) and could be associated with poor postoperative respiratory outcomes (13). A hypoplastic unilateral lung, which is usually associated with a hypoplastic bronchus, may result in the need for persistent mechanical ventilation postoperatively. In fact, the authors reported that two patients with BM of >20% developed a hypoplastic right lung associated with Goldenhar syndrome and required an additional surgical intervention after tracheoplasty.

Notably, two patients with BM of >20% underwent successful reoperation after slide tracheoplasty. Both underwent bronchial reconstruction (tracheal and right main stem bronchial reconstruction and bilateral bronchial reconstruction) with autologous rib cartilage grafts. A patient with BM of <20% but with hypoplastic bilateral main stem bronchi underwent resection of localized CTS with preemptive bilateral bronchial reconstruction using fragments from the resected autologous tracheal tissue and successfully recovered without mechanical ventilation support. The authors concluded that proactive bronchial reconstruction may be justified in patients with BM of >20% to achieve successful respiratory outcomes.

In this study, as well as in other studies (8), the length or degree of the stenosis did not affect the surgical outcomes. Although preoperative contrast-enhanced CT is a powerful tool to determine the type of CTS, relationships between the great vessels and trachea, presence of anomalous vessels, and presence of pulmonary hypogenesis, the diameter of the trachea or bronchus on CT imaging is not always accurate. Secretions trapped in the narrow trachea and/or bronchi or the presence of a malacia tracheobronchial wall make the diameter appear to be narrower on CT than the real diameter. Additionally, swelling of the tracheal wall with airway inflammation, often at the time CT is performed, significantly reduces the airway diameter. Thus, BM should preferably be calculated on CT images when the patient is not experiencing severe respiratory symptoms or at multiple time points to ensure accuracy.

In the patient with a tracheal bronchus, the authors defined BM as mismatch between the bridging right bronchus and the left main bronchus. One patient with this "porcine type" CTS underwent modified side-slide tracheoplasty. A tracheal bronchus is not a rare arborization pattern in patients with CTS. Butler *et al.* (11) reported that CTS presented with an anomalous right upper lobe bronchus in 13% of patients in their series. Side-slide tracheoplasty was reported by Wang *et al.* (14) as a useful modification in patients with long-segment CTS with a tracheal bronchus. However, this technique is associated with potential risks of anastomotic complications due to damage to the lateral blood supply and severe tracheobronchial malacia due to the transverse figure-of-eight configuration (15). Manning *et al.* (5) reported that no specific modification with slide tracheoplasty was necessary for most patients with a right upper tracheal bronchus.

The authors described another patient with short-segment CTS and an abortive tracheal bronchus along with bilateral hypoplastic bronchi who underwent bronchial plasty using an autologous tracheal fragment from the resected tracheal tissue, and this patient successfully recovered without mechanical ventilation. Quite a few

reports have described the performance of bronchial plasty, mostly using rib cartilage (16,17). The use of autologous tracheal fragments was reported by Backer *et al.* (18) for treatment of long-segment tracheal stenosis and by Huang *et al.* (19) for bridging of the bronchus and distal bronchi to augment the lumen. However, bronchial plasty with slide tracheoplasty can be extremely challenging, and it is not always applicable (20,21).

The authors excluded patients with postoperative tracheal granulation or restenosis that were treated by balloon dilatation from those with chronic respiratory failure. Manning *et al.* (5) reported that gentle balloon dilatation was useful for a figure-of-eight deformity after slide tracheoplasty and that a single dilatation was not defined as a significant airway reintervention in their outcome measures. Butler *et al.* (11) reported that 45% of children required balloon dilatation, and among them, almost half required subsequent stenting. Although balloon dilatation is often effective to treat granulation and restenosis, the damage to the tracheal wall caused by balloon dilatation results in laceration and/or perforation and may cause significant sequelae; additionally, patients need tracheostomy for long-term mechanical ventilation or stenting (5,6). The need for more than a single balloon dilatation should be carefully considered because of the risk of serious postoperative complication.

As the authors attempted to demonstrate factors associated with postoperative outcomes, information is needed on which patients can benefit from tracheoplasty. The mortality rate associated with CTS has significantly decreased in recent reports since the advent of slide tracheoplasty and improvements in preoperative and postoperative management by multidisciplinary teams that comprise cardiovascular surgeons, cardiologists, otolaryngologists, pediatric surgeons, anesthesiologists, pulmonologists, intensivists, and respiratory nurse specialists. Moreover, due to improvements in contrast-enhanced three-dimensional CT imaging, CTS can be diagnosed in local institutions before patients develop respiratory failure. However, the preoperative respiratory status significantly affects the postoperative course. The need for preoperative mechanical ventilation or extracorporeal membranous oxygenation has been shown to be a prognostic factor for prolonged extubation after slide tracheoplasty (5,7,10,11). Surgical intervention before respiratory symptoms deteriorate due to airway infection would be advantageous for patients with CTS, while conservative treatment would be advocated for patients with mild symptoms. Thus, the risk factors affecting the postoperative course should be clarified to define the optimal treatment strategy for individual patients with CTS.

Since CTS is a rare and potentially critical condition, the limitations of studies including the study by Cetrano *et al.* (12) that have assessed the prognostic factors in patients with CTS are far from trivial. The numbers of patients included in these cohorts are usually too small to reach statistically significant results. Because the wide range of anatomical and clinical presentations necessitates individualized management, it is usually difficult to make uniform preoperative assessments in terms of timing and modalities. Concomitant congenital anomalies, especially the presence of severe cardiovascular diseases, may significantly influence the management of patients with CTS. Identification and follow-up of patients with mild or moderate symptoms without the need for surgical interventions is not always possible; thus, it would be difficult to use this cohort as a control for comparison with patients who require surgical interventions. Each surgeon's experience as well as the use of various surgical modifications, especially for complicated cases (22), would affect the outcomes. Hence, it is ideal to establish a multicenter registry system for data collection and analysis, which would allow us to validate whether BM of >20% and other factors are significant prognostic factors.

In conclusion, although we undoubtedly need further investigations involving larger and preferably prospective series to determine whether BM of >20% is a predictor of chronic respiratory failure after tracheoplasty, this manuscript has highlighted some important factors to consider when preoperatively assessing the anatomy of patients with CTS to achieve a successful tracheal intervention.

Acknowledgements

None.

Footnote

Conflicts of Interest: The author has no conflicts of interest to declare.

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Cite this article as: Yokoi A. Congenital tracheal stenosis: what should we look at for successful tracheoplasty? Transl Pediatr 2018;7(3):229-232. doi: 10.21037/tp.2018.07.01