Retrospective review of tracheoplasty for congenital tracheal stenosis

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ABSTRACT

Introduction: Congenital tracheal stenosis is a rare but life-threatening obstructive airway disease. It usually presents in early infancy and requires surgical intervention. This study is a review of our experience in the management of congenital tracheal stenosis in children at KK Women's and Children's Hospital, Singapore.

Methods: All children who had undergone tracheoplasty for congenital tracheal stenosis between January 1999 and December 2008 were included. The patients' medical records were retrieved from our database, and the demographic data, comorbidities, clinical presentation and management, surgery performed, postoperative complications, final outcomes and follow-up were reviewed.

Results: A total of 11 children aged 12 days to six years underwent surgery for congenital long-segment tracheal stenosis, of which ten (90.9 percent) had associated cardiac and vascular anomalies and nine (81.8 percent) had left pulmonary artery sling predominance. Five (45.4 percent) children had associated bronchopulmonary abnormalities. All the patients underwent slide tracheoplasty with concomitant repair of congenital heart defects. Overall mortality was 45.4 percent (n is 5), including one late mortality due to an unrelated cause. With the exception of one, the follow-up of all patients was complete at this writing. One patient had mild residual tracheal stenosis and another had bilateral bronchomalacia.

Conclusion: This is the largest case series of congenital long-segment tracheal stenosis reported in Southeast Asia to date. Slide tracheoplasty with concomitant repair of cardiac lesions is currently the preferred management for long-segment stenosis. Management of such patients remains a challenge for clinicians and the patients' families, and requires a multidisciplinary approach.

Keywords: airway obstruction, congenital tracheal stenosis, infant, Southeast Asia, trachea

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INTRODUCTION

Tracheal stenosis is a very rare but often life-threatening structural obstructive airway disease. In congenital tracheal stenosis, complete cartilaginous tracheal rings are often present and the posterior membranous trachea is absent. A wide variety of morphological variants have been described, with no agreed-upon international classification to date.

Congenital tracheal stenosis is often associated with congenital cardiac defects and vascular anomalies, in particular, a left pulmonary artery sling. This is known as the ring-sling complex, where the anomalous left pulmonary artery arises from the right pulmonary artery and continues leftward to encircle the lower trachea, causing external compression of the distal trachea, and occasionally, the right and left mainstem bronchi.[1] Affected children often present early in infancy with stridor, severe respiratory distress and/or recurrent chest infections requiring non-invasive or invasive mechanical ventilation.[1-6] Some children may even present *in extremis* with cardiac arrest. Surgical options available for treatment of congenital long-segment stenosis include resection with primary end-to-end anastomosis, patch tracheoplasty, slide tracheoplasty and tracheal homograft.[2-14] Over the last decade, slide tracheoplasty has become the preferred option for management of congenital long-segment tracheal stenosis, resulting in an improvement in mortality.[2,3,5,6]

Given the rarity of this condition, there is a lack of large-scale studies on its management, and the majority of published studies are single-centre series. To date, in Southeast Asia, there have only been one published case series of four infants in Thailand in 2002 and one case report in 1998.[15,16] This current case series reviews the outcomes for children with congenital

long-segment tracheal stenosis treated at our institution over a nine-year period.

METHODS

This study was approved by the Institutional Review Board as a retrospective case series, with waiver of informed consent. All patients who underwent surgery for congenital long-segment tracheal stenosis in KK Women's and Children's Hospital, Singapore between January 1999 and December 2008 were included in this study. Those who did not undergo surgery for congenital long-segment tracheal stenosis were excluded. Patient data was obtained from our department's anaesthesia audit database between January 1999 and December 2008. This database keeps a record of all children who had surgery under general or regional anaesthesia at our institution. The patients' medical records were then retrieved and reviewed for the demographic data (gestational age, gender, race, age at diagnosis, age and weight at the time of surgery), comorbidities, clinical presentation and its management, including the need for preoperative ventilator support, type of tracheal stenosis and type of surgery performed, postoperative complications, final outcomes and follow-up.

RESULTS

A total of 11 children underwent surgery for congenital tracheal stenosis (Table I). Five out of the 11 patients (45.4%) presented within the neonatal period. Of these five neonates, three presented with signs of respiratory distress at birth and stridor. Two neonates were found to have tracheal stenosis during a workup for congenital heart disease, which revealed a left pulmonary artery sling.

Delayed diagnosis of congenital tracheal stenosis was noted in two children. They presented at ages 21 months and six years, with recurrent episodes of stridor and chest infection since infancy, and had subsequently defaulted follow-up. The six-year-old child had episodes of wheezing precipitated by upper respiratory tract infections, which had been diagnosed as asthma prior to her initial presentation at our institution. Seven of the 11 children (63.6%) required either continuous positive airway pressure or invasive positive pressure ventilation prior to surgery. In particular, one patient required heliox for worsening respiratory distress and stridor, while another suffered a hypoxic cardiac arrest prior to surgery.

Associated anomalies were present in ten of the 11 children (90.9%, Table II). A left pulmonary artery sling was found in nine (81.8%) patients. One neonate had an antenatal diagnosis of complex congenital heart disease with a double outlet right ventricle, large ventricular septal defect, pulmonary atresia with patent ductus arteriosus and left pulmonary artery sling. Respiratory anomalies included a laryngeal web (n = 1), right lung sequestration with Scimitar syndrome (n = 1) and right lung hypoplasia in a child with long-segment tracheal stenosis extending into the right main bronchus.

Diagnosis of congenital tracheal stenosis was reached in all cases using a combination of computed tomography of the chest, with or without rigid bronchoscopy. All patients underwent two-dimensional echocardiography for diagnosis of concomitant congenital heart disease and vascular anomalies. The median age of the patients at the time of surgery was 14 weeks (range two weeks to six years), while the median weight was 4.0 (range 1.3–27) kg. Slide tracheoplasty was performed in all cases. Relocation of the left pulmonary artery sling and repair of other congenital cardiac lesions were performed in the same setting for all cases. In addition,

Table I. Patient demographics.

Demographic	No. of patients (%)
Gender	
Male	1 (9.1)
Female	10 (90.9)
Age at presentation	
0–28 days	5 (45.4)
1–6 mths	4 (36.4)
> 6 mths	2 (18.2)
Age at surgery	
0–28 days	1 (9.1)
1–6 mths	7 (63.6)
> 6 mths	3 (27.3)
Clinical presentation	
Stridor	5 (45.4)
Respiratory distress	5 (45.4)
Infection	3 (27.3)
Cyanosis	2 (18.2)
Need for preoperative ventilation	
Yes	8 (72.7)
No	3 (27.3)

Table II. Associated anomalies.

Type of anomaly	No. of patients (%)
Left pulmonary artery sling	9 (81.8)
Atrial septal defect	4 (36.4)
Ventricular septal defect	2 (18.2)
Patent ductus arteriosus	3 (27.2)
Pulmonary atresia	1 (9.1)
Double outlet right ventricle	1 (9.1)
Hypoplastic right lung	1 (9.1)
Right lung sequestration with Scimitar syndrome	1 (9.1)

one child had a right main bronchus pericardial patch, where the tracheal involvement had extended into the right main bronchus. Surgical repair was performed via a median sternotomy in eight (72.7%) cases. In three patients, slide tracheoplasty and posterior translocation of the left pulmonary artery sling were repaired via a right thoracotomy. Cardiopulmonary bypass was used in nine of the 11 cases (81.8%). All patients were directed postoperatively to the intensive care unit with ventilatory support. The duration of postoperative ventilation was 2–36 days, while the length of hospital stay was 22–302 days for survivors.

Early in our series, two patients died in the immediate perioperative period. Both had undergone slide tracheoplasty as an emergency surgery due to progressive respiratory failure despite sedation, paralysis and high ventilating pressures. The causes of death were postoperative pneumonia with bronchopleural fistula and pulmonary haemorrhage in one patient, and right lung hypoplasia with severe pulmonary hypertension in the other. Subsequent mortality after 2002 was 37.5% (three deaths out of eight cases). Overall mortality was 45.4% (five deaths out of 11 cases), with one late mortality due to an unrelated cause. The causes of death included postoperative pneumonia, severe acute respiratory distress syndrome, pulmonary haemorrhage and tracheal dehiscence. Four out of the five patients who died had co-existing congenital heart disease and concomitant surgical repair at the time of slide tracheoplasty.

Five out of the six surviving patients were followed up till 2010. One patient defaulted follow-up two years after the initial surgery. Of the survivors, two were active and free from stridor till the time of this writing. Three patients had recurrent admissions for exacerbations of asthma and chronic lung disease. One patient had residual tracheal stenosis 1 cm from the carina, with mild narrowing of the right main bronchus, while another had bilateral bronchomalacia requiring long-term non-invasive ventilation. The last patient was on a tracheostomy with speaking valve, with mild collapse of the left main bronchus demonstrated on postoperative microlaryngobronchoscopy.

DISCUSSION

Tracheal stenosis can be defined as a reduction in the anatomical luminal diameter of the trachea by greater than 50% compared with the remaining normal trachea.[17] It may be congenital or acquired following trauma or prolonged intubation. Congenital tracheal stenosis in children can occur in isolation or secondary to vascular ring abnormalities. In our series, 81.8% of children had an associated left pulmonary artery sling and other congenital cardiac anomalies, compared to the studies in the international literature, which reported an incidence of 40%–60%.[1,4] In long-segment congenital tracheal stenosis, more than two-thirds of the length of the trachea is affected.[17] This often occurs in association with complete cartilaginous tracheal rings and an absence of the normal posterior membranous trachea. The complete tracheal rings may extend into the bronchi, or a tracheal bronchus may occur. Cantrell and Guild described three general morphologies of congenital tracheal stenosis as 'generalised', 'funnel-shaped' and 'segmental' stenosis.[18] However, these do not encompass the wide variety of tracheal and bronchial types of stenosis. A more useful classification regarding the severity of tracheal stenosis has been described by Elliott et al,[2] where the four elements contributing to the severity of disease include: (1) the narrowness of the trachea; (2) the extent of tracheal involvement; (3) the involvement of the bronchi; and (4) the presence or absence of complete tracheal rings.[4]

Depending on the severity of the stenosis, affected children may present with recurrent stridor, chest infections or near-death episodes requiring cardiopulmonary resuscitation. Children with a left pulmonary artery sling without long-segment tracheal stenosis can also present with stridor due to compression of the trachea. In our series, the neonates presented with more severe symptoms of stridor and severe respiratory distress requiring positive pressure ventilation preoperatively. Six (54.5%) children underwent tracheoplasty as an emergency surgery due to progressive respiratory failure despite mechanical ventilation. Older infants and children presented with recurrent chest infections and stridor, often with a delayed diagnosis of congenital tracheal stenosis. The diagnosis of congenital tracheal stenosis is often reached with a combination of computed tomography with three-dimensional reconstruction and microlaryngobronchoscopy. The latter is also used to delineate the anatomy and extent of the lesion. Two-dimensional echocardiography is performed to check for associated cardiac anomalies.

The four main surgical techniques for congenital long-segment tracheal stenosis include tracheal resection with end-to-end anastomosis, slide tracheoplasty, autologous patch tracheoplasty and tracheal homograft.[5] Primary tracheal resection with end-to-end anastomosis is better suited for short-segment tracheal stenosis. Excessive tension at the anastomotic site makes it less suited for long-segment tracheal stenosis, as it carries the risk of leakage and recurrent stenosis.

Over the last decade, slide tracheoplasty has emerged as the preferred treatment for long-segment tracheal stenosis, along with concurrent repair of cardiac anomalies.[2-6] With slide tracheoplasty, the circumference of the trachea is doubled, and the cross-sectional area quadrupled. As native tracheal tissue is used, granulation tissue is minimal, and tracheal growth has been shown to occur following slide tracheoplasty.[19] Patch tracheoplasty is suited for very long segments of tracheal stenosis and in cases where bronchial involvement is noted. Autologous pericardium is most commonly used. Other autologous tissues used include the rib cartilage, tracheal autograft or allograft and carotid artery.[3-5,7,19,20] A recent study evaluating the long-term outcomes of anterior pericardial tracheoplasty for long-segment tracheal stenosis reported 19% mortality in 26 patients over a 26-year period.[7] However, patch tracheoplasty is often complicated by frequent granulation tissue formation and postoperative recurrent tracheal stenosis, which may require repeat bronchoscopic removal of granulomas, stent placement or surgery.[3-5,7,19,20] Postoperative complications are often related to prolonged cardiopulmonary bypass, repair of cardiac lesions, recurrent tracheal stenosis due to formation of granulation tissue and tracheomalacia, which may subsequently be unmasked following repair of tracheal stenosis.[2,5,6,19]

In conclusion, severe long-segment tracheal stenosis remains a great challenge for clinicians, patients and their families. Affected children tend to present early in infancy, with inspiratory stridor and severe respiratory distress requiring invasive positive pressure ventilation. Ventilation strategies in small babies are a challenge, and helium-based ventilation may be required. Near-death episodes are also frequent. At present, slide tracheoplasty with concomitant repair of cardiovascular lesions is the gold standard for treatment of long-segment tracheal stenosis. Management of children with congenital tracheal stenosis is complex and requires a multidisciplinary approach involving cardiac surgeons, anaesthetists, intensivists and physiotherapists in order to ensure optimal management.

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