



A child with congenital tracheal stenosis presenting as recurrent wheezing

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Introduction

Congenital tracheal stenosis is a rare and potentially fatal congenital disorder which usually presents in the first few weeks of life. It is caused by focal or diffuse complete tracheal cartilage rings resulting in fixed tracheal narrowing. Many patients also have other associated congenital anomalies, commonest being pulmonary artery sling (35%), congenital heart diseases (22%) and anomalous right upper lobe bronchus (22%).¹ In severe cases patients present with respiratory distress shortly after birth. Patients with milder disease may present later in life with recurrent bronchiolitis or croup. A patient with congenital tracheal stenosis was presented here with recurrent wheezing during infancy.

Case report

A six-month-old girl was admitted to our hospital because of cough, stridor, wheeze and fever for several days. She was born at full term with birth weight of 2.3 kg. At one-month, she was admitted to a hospital in China because of pneumonia and was hospitalised for 2 months during that episode.

Physical examination on admission revealed that she had failure to thrive with head circumference at 3rd percentile and body weight and height below 3rd percentile. She was febrile with inspiratory stridor and mild respiratory distress. There was one 2 cm haemangioma over scalp and one over left upper arm. Examination of the respiratory system revealed tachypnoea, supra-sternal insucking and bilateral crepitations and rhonchi over the lungs. Cardiovascular examination revealed a continuous murmur below left clavicle and no heart failure.

Chest X-ray showed a mildly hyper-inflated lung with increased peri-hilar bronchial markings and no

consolidation. Sputum culture grew *H. influenzae*. Echocardiogram and cardiac catheterisation revealed a small patent ductus arteriosus. Flexible bronchoscopy showed signs compatible with laryngomalacia. She was treated with antibiotics and bronchodilators and showed good response.

In the next few months she developed recurrent episodes of respiratory distress and was admitted repeatedly with various diagnoses including pneumonia, acute bronchiolitis and croup. Further investigations including 24-hour esophageal pH, barium swallow and immunoglobulin levels and IgG subclasses and secretory IgA level were all normal.

A repeat flexible bronchoscopy was done when patient was 1 year old. This time there was no sign of laryngomalacia but tracheal stenosis was diagnosed because of the circular-shaped trachea and the absence of the posterior membranous portion of trachea. A bronchoscope with 3.5 mm external diameter could not be passed through the stenotic segment. The stenotic segment was passed with a 1.9 mm bronchoscope. CT thorax confirmed tracheal stenosis with complete cartilaginous rings over stenotic segment (Figure 1). The axial diameter of upper trachea was 7 mm and the axial diameter of the distal stenotic trachea above carina was 4 mm. The findings were compatible with long segment distal tracheal stenosis.

The patient was referred to the cardio-thoracic unit for assessment. She was offered operative management for the tracheal stenosis but the parents refused because of the high risks associated with the operation.

Subsequently she had several CT thorax to assess the growth of the stenotic trachea. As shown in Table 1, there was no growth over the stenotic segment as measured from the CT scan. However, our patient improved clinically as she grew older. She had frequent admissions due to respiratory problems in the first 2 years of life but her condition gradually improved and she did not have any pneumonia or wheezing attack in the past 2 years.

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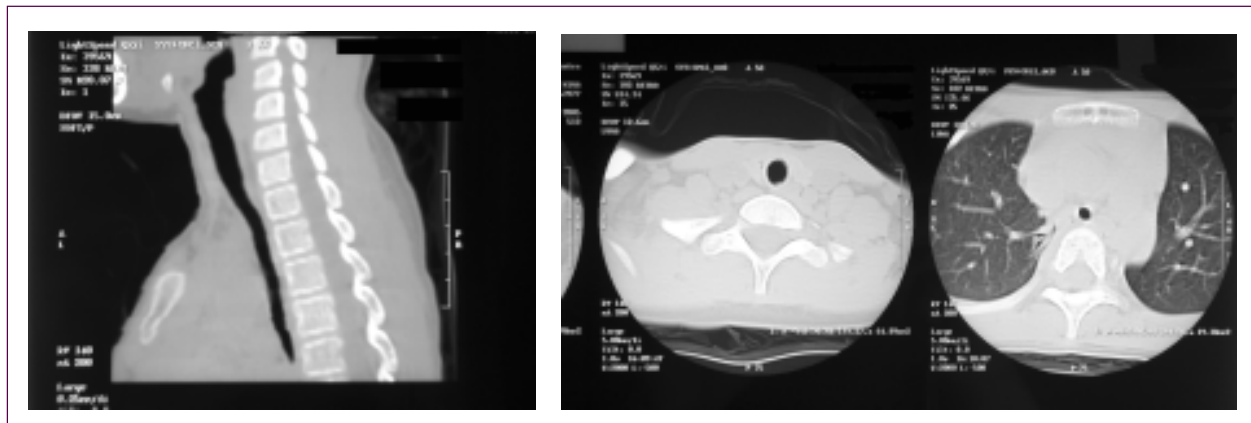


Figure 1. Left: CT trachea showing narrowed lower trachea. Right: cross section of trachea showing normal upper trachea (left) narrowed lower trachea (right).

Table 1. Serial CT measurement of cross-sectional area of normal trachea (1st rib) and stenotic trachea (just above carina) at different age

Age	Tracheal area at 1st rib (mm ²)	Tracheal area above carina (mm ²)
1	31	18
2	42	20
5	52	17
8	65	19

She is now an 8 years old primary 3 student. She leads a relatively normal life and is described to be an active child. She can walk 6 flights of stairs with shortness of breath afterwards. Her growth is satisfactory and her body weight and height is presently on the 25th percentile, which corresponds to her parents' height.

Lung function test and cardio-pulmonary assessment were done to assess our patient's pulmonary and cardiac status. Spirometry revealed normal forced vital capacity (FVC) of 1.32 L (91% predicted), decreased forced expiratory volume in 1 second (FEV1) of 0.8 L (60% predicted), which is compatible with airway obstruction. The maximal voluntary ventilation (MVV) is decreased to 36% predicted. However the 6 minutes walk test was within normal range. Cardio-pulmonary assessment with Bruce protocol was done using treadmill with patient having good effort and achieving a maximal heart rate (HR) of 178/min (85% of predicted maximal HR). Oxygen saturation was normal throughout the test. Her maximal oxygen consumption (VO₂max) was slightly decreased to 74% predicted. There was also a marked decrease in ventilatory reserve with maximal minute ventilation

(VE_{max}) equals to 89% of MVV (normal $\leq 50\%$), indicating a primary ventilatory limitation to exercise and VO₂max.

Discussion

Congenital tracheal stenosis is a rare anomaly where there is narrowing of trachea due to fusion of the normal C-shaped tracheal cartilage posteriorly, replacing the posterior membranous portion of the trachea. The severity of symptoms depends more on the diameter of the trachea than the length of the stenosis, as airway resistance is only linearly proportional to length of stenosis, whereas resistance increases fourfold as the luminal radius decreases. Congenital tracheal stenosis is classified into long and short segment, with long segment type having a stenotic segment greater than 50% of total tracheal length.

Patients with congenital tracheal stenosis can present with different symptoms depending on the patient's age, degree of stenosis and associated anomalies. In general, the earlier the presentation, the more severe is the stenosis. The most severely affected patients usually present with respiratory distress soon after birth. If intubated, these patients fail extubation repeatedly. For those patients presenting later in life, symptomatology could be variable. Common presentation includes stridor, recurrent bronchiolitis and recurrent croup. The majority of patients present in infancy or early childhood. However, there was one case report of an adult patient with congenital tracheal stenosis presenting as asthma.²



High-kilovoltage neck radiographs show a narrowed airway. Bronchoscopy, bronchogram and CT can confirm the diagnosis of congenital tracheal stenosis by showing the complete tracheal ring and narrowing of the trachea. Other investigations including echocardiogram, barium esophagram will help to diagnose associated congenital anomalies.

Literature generally suggested that patients with congenital tracheal stenosis had grave outcome in the absence of surgical correction.³ It was widely believed that there was no growth potential for the complete cartilaginous rings. However, this view was challenged by Manson et al⁴ who reported two patients with complete cartilaginous ring tracheal stenosis who subsequently showed clinical improvement and growth of trachea with conservative management.

The rarity of congenital tracheal stenosis has not allowed sufficient experience for the development of a standard surgical approach. Several surgical techniques were used, including resection and anastomosis,^{1,5,6} slide tracheoplasty,^{1,7} patch tracheoplasty,^{1,8} tracheal autograft¹ and balloon tracheoplasty with stenting.⁹ For short segment tracheal stenosis, resection and anastomosis was the treatment of choice and the average mortality from published studies was 8%.¹ The choice of surgical techniques for long segment tracheal stenosis varied. Backer et al¹ reported their 18-year experience of tracheal surgery in children and reviewed the literature. They found that the average mortality for patch tracheoplasty was 28%, average mortality for slide tracheoplasty was 20% and mortality for tracheal autografts in their own centre was 8%.

Our patient has long segment tracheal stenosis so resection and re-anastomosis is not possible. Although slide tracheoplasty was done in infants as young as 2-week-old,⁷ in general the younger the patient the more difficult is the surgery. For patients with severe diseases it is obvious that surgery needs to be done as soon as possible. It is difficult to decide on optimal timing of surgical intervention in milder cases. Our patient's

parents refuses operative management because of the high risks associated with the operation and because the patient is apparently leading a normal life at present. However cardio-pulmonary assessment showed that she had impaired function with decreased FEV₁, decreased MVV and decreased VO₂max. At present she can still compensate for the impaired function as illustrated by the normal six minutes walk test and normal daily activities. However, she should be closely monitored for any deterioration of cardio-pulmonary function. With the absence of growth of the complete tracheal ring shown in the serial CT scan, the patient will most likely need surgical intervention in the future.

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