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## Titanium plating system with autologous rib graft sternoplasty in the treatment of thoracic inlet compression



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#### ABSTRACT

Narrowing of the thoracic inlet leading to airway compression is a rare and challenging condition in the pediatric population. Reports in the literature have described this variant related to multiple conditions including double crush phenomenon following repair of pectus excavatum, anterior spinal displacement, and straight back syndrome. Underlying genetic conditions such as Marfan's Syndrome and Hurler's Syndrome have also been reported to contribute to clinically significant airway compression independent of dynamic tracheal collapse such as tracheomalacia. The borders of the thoracic inlet are anatomically bound by the body of the first thoracic vertebrae (T1) posteriorly, the posterior surface of the manubrium anteriorly, and the medial aspects of the first ribs on either side laterally. Relief of tracheal compression in this location is complicated by the rigidity of the boney thoracic inlet and limited space for lifting procedures such as anterior aortopexy. Several operative approaches to treat this condition have been described including manubrial/sternal resection, first rib resection, and reconstruction of the thoracic inlet. Described here are three patients where successful reconstruction of the thoracic inlet was achieved using autologous rib graft sternoplasty and a titanium sternal plating system to widen the thoracic inlet and eliminate external compression on the trachea.

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Narrowing of the thoracic inlet leading to airway compression is a rare and challenging condition in the pediatric population [1]. This unusual phenomenon has been associated with conditions including anterior spinal displacement, straight back syndrome, and double crush phenomenon [2-4] as well as various genetic diseases [5-8]. Due to the complexity of the anatomy, several surgical approaches have been described including sternal resection, first rib resection, and reconstruction of the thoracic inlet [7,9-12]. Here we describe three patients in whom tracheal compression was successfully relieved by utilizing autologous rib graft and titanium sternal plating to expand the anterior boundaries of the thoracic inlet and relieve tracheal compression.

### 1. Case reports

Patient 1 was a 14-year-old boy with a history of hypoxic ischemic encephalopathy and spastic cerebral palsy. Six months following a laparoscopic fundoplication with gastrostomy tube for gastroesophageal reflux disease, his family described worsening cough, gagging, and choking. He was evaluated by otolaryngology and underwent a tonsillectomy and adenoidectomy for upper airway obstruction. Two weeks following this procedure, he experienced a hypoxic arrest at home. He was intubated and transferred to our institution. He failed extubation twice due to repeated acute life threatening events secondary to airway obstruction. Diagnostic bronchoscopy revealed non-dynamic external compression of the distal trachea. The left mainstem bronchus was not able to be intubated due to external compression. Chest computed tomography showed there was significant compression of the trachea and left mainstem bronchus by the innominate artery and aortic arch (Fig. 1A). On sagittal images, the distance of the thoracic inlet was 28 mm, likely due to a loss of

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**Fig. 1.** (A) 3-D reconstruction of chest computed tomography demonstrating severe trachael compression just distal to the endotracheal tube and of the left mainstem bronchus. (B) Pre-operative chest computed tomography showing the loss of normal thoracocervical kyphosis consistent with straight back syndrome and trachael compression by the aortic arch. The distance from the posterior manubrial plate to the anterior vertebral line is 28 mm.

normal thoracic kyphosis consistent with straight back syndrome (Fig. 1B). Consequently, a standard aortopexy was unlikely to relieve the tracheal compression.

The patient was taken to the operating room for median sternotomy, transposition of the innominate artery and aortopexy with intra-operative bronchoscopy [4]. Once the sternum was opened, the compression on the trachea was relieved as determined by intraoperative bronchoscopy. The relief was augmented by lifting the innominate artery and aorta anteriorly. However, the compression recurred when the sternal edges were reapproximated. Thus, the decision was made to perform a rib expanded sternoplasty instead of transposing the innominate artery. The 6th rib was harvested and divided into three 2 cm sections. The upper sternum was expanded using the 2 cm rib struts and stabilized with titanium plates (Fig. 2).

The aorta was secured to the posterior sternal plate with permanent thread prior to closure. The lower sternum was closed in standard fashion with wire. Pectoralis major advancement flaps were mobilized bilaterally to cover the defect. The tracheal compression was markedly improved upon completion of the reconstruction. He was able to be extubated on postoperative day 3 without recurrent airway symptoms. His hospital stay was extended due to post-cardiotomy syndrome treated with steroids. On follow-up 6 months later, he remains symptom free from airway obstruction. Repeat chest computed tomography demonstrates expansion of the thoracic inlet from 28 mm preoperative to 35 mm postoperatively (Fig. 3). Due to concern for worsening thoracic lordosis, an orthopedic consultation was obtained and continued observation was recommended.

Patient 2 was a 7-year-old girl with 15q partial trisomy and 9p partial monosomy syndrome with developmental delay and hypotonia. She had been followed for 18 months by the pulmonary allergy group for persistent bacterial bronchitis. Chronic antibiotics, reflux medications, and anti-inflammatory medications failed to relieve her symptoms and her cough worsened to such an extent that she was removed from her classroom. On exam, she had marked manubriosternal synostosis (pouter pigeon breast) with a manubriosternal angle of 135° by chest x-ray. Chest computed tomography displayed normal vasculature and confirmed the sternal deformity with tracheal compression (Fig. 4A). Diagnostic bronchoscopy found severe compression at the carina involving the left mainstem bronchus with an estimated 80% loss of circumference. Airway pressures of 35-40 mm Hg resulted in expansion to 50% of the normal circumference. This finding along with the CT scan suggested that aortopexy alone would not relieve the tracheal compression.

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