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Brief Report

Severe stenosis of a long tracheal segment, with agenesis of the right lung and left pulmonary arterial sling

Hamish M. Munro,[1] Andrea M. C. Sorbello,[2] David G. Nykanen[2]

[1]Department of Cardiovascular Anesthesia, [2]Department of Cardiology, The Congenital Heart Institute, Arnold Palmer Hospital for Women and Children/Miami Children's Hospital, Orlando, Florida, United States of America

Abstract A baby presented at term with respiratory distress was managed with extracorporeal membrane oxygenation. Bronchoscopy revealed tracheal hypoplasia, complete tracheal rings, and agenesis of the right main bronchus. Echocardiography showed a left pulmonary arterial sling arising from the proximal part of the right pulmonary artery. Cardiac catheterization demonstrated abnormal pulmonary vasculature in the left lung which would have prevented survival, even after surgical repair. Diagnostic catheterization was important in delineating the anatomy, and aided in the decision not to proceed with surgical repair.

Keywords: Congenital heart disease; great vessel anomalies; lung

We describe an unusual anatomic variant of pulmonary arterial sling, with agenesis of the right lung, patency of the arterial duct, and complete tracheal rings. Angiography revealed hypoplasia of the left lung and diffuse tracheal stenosis, findings which influenced the decision not to proceed with surgical intervention.

Case report

A female infant weighing 2981 grams was delivered at term by Cesarean section, but rapidly developed respiratory distress and bradycardia. Orotracheal intubation was achieved with difficulty using a 2.5 millimetre endotracheal tube, and it was noted that the tube could not be advanced past eight centimetres. A chest radiograph demonstrated a left tension pneumothorax, and a chest tube was inserted. The right lung was displaced and compressed and the heart rotated, with mediastinal shift to the right. Fibreoptic bronchoscopy through the endotracheal tube revealed a long and hypoplastic tracheal segment, with complete tracheal rings and atresia of the right main stem bronchus (Fig. 1). Despite conventional ventilatory support, the patient remained cyanotic, acidotic, and hypercarbic. Because of this, we started veno-arterial extracorporeal membrane oxygenation. Transthoracic

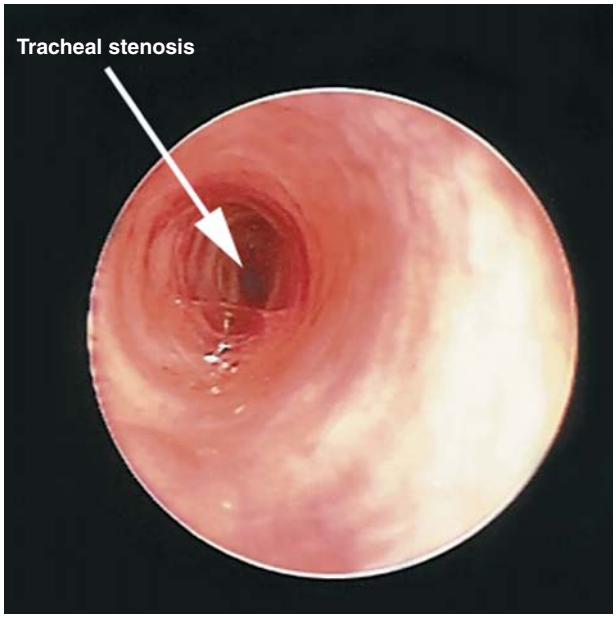

Figure 1.
Bronchoscopic image demonstrating stenosis associated with complete tracheal rings. The main stem of the right bronchus, not shown, was atretic.

Correspondence to: David G. Nykanen MD, Department of Cardiology, 50 West Sturtevant St, Orlando, FL 32806, USA. Tel: +1 407 649-6907; Fax: +1 407 481 2035; E-mail: david.nykanen@orhs.org

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echocardiography demonstrated normal intracardiac anatomy, albeit with suprasystemic right ventricular pressures and severely depressed right ventricular function. Imaging of the pulmonary arteries showed a pulmonary trunk of normal size, but flow could not be visualized in the right pulmonary artery. The left pulmonary artery was thought to be hypoplastic with impaired flow (Fig. 2a). There was a small patent arterial duct, with bi-directional flow. The infant was weaned from extra-corporeal membrane oxygenation on the eighth day, and maintained on conventional ventilation. Acute deterioration 10 days later required high frequency oscillatory ventilation, and despite treatment with both helium and nitric oxide, hypercarbia persisted, needing the re-establishment of extra-corporeal membrane oxygenation. Cardiac catheterization was performed. Mixed venous saturations were 59 and 61% in the right atrium and pulmonary arteries, respectively. The descending aortic saturation was 79%, with a systemic saturation greater that 95%, indicating right-to-left shunting at the level of the arterial duct. The right ventricular pressure was suprasystemic, at 87 over 12 millimetres of mercury, with an aortic pressure of 58 over 38. A 0.035 inch soft tipped wire was introduced through the endotracheal tube into the main stem of the left bronchus to outline its intraluminal course. This facilitated angiographic confirmation of a left pulmonary arterial sling (Fig. 2b). The pulmonary trunk was of normal size, continued rightward as the proximal right pulmonary artery, and then gave rise to the left pulmonary artery, which then coursed posterior to the distal trachea. There was no evidence of proximal stenosis in the left pulmonary artery, albeit that the vessel was severely hypoplastic, measuring no more than 2.2 millimetres diameter throughout its course. Distal vascularity of the left lung was severely abnormal, with tortuosity, stenosis of the branches of the pulmonary artery, distal pruning, and perfusion defects. Pulmonary venous return and left ventricular function were normal.

Because the left lung was markedly abnormal, with severely hypoplastic pulmonary arterial vasculature in the setting of agenesis of the right lung, we decided that further surgical therapy would be futile. Life support was withdrawn, and the baby died shortly thereafter.

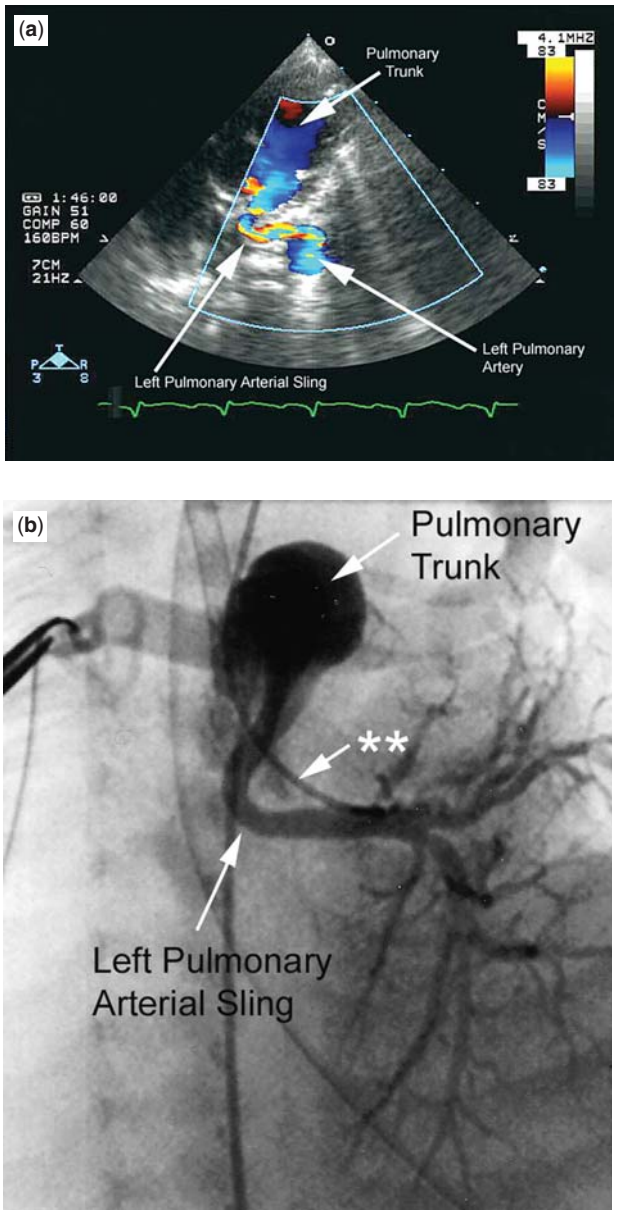

Figure 2.
*The parasternal short axis echocardiographic image (**a**) demonstrates the pulmonary trunk, with the left pulmonary artery arising as a sling from the proximal right pulmonary artery. Angiography (**b**) confirms the atresia of the distal right pulmonary artery. The left pulmonary artery courses posterior to the distal trachea, which has been "marked" by a wire inserted into the left mainstem bronchus (**). Note the hypoplasia of the left pulmonary artery, with multiple distal arterial stenoses.*

Discussion

Several case reports have discussed the management of patients with tracheal stenosis with complete cartilaginous rings, with or without right pulmonary agenesis, and left pulmonary arterial sling.[1–3] The pulmonary arterial sling is a congenital anomaly in which the left pulmonary artery originates from the right pulmonary artery, and then courses posteriorly above the main stem of the right bronchus before turning leftward behind the trachea and in front of the oesophagus. Half of afflicted patients also exhibit tracheal stenosis.[4] As the right pulmonary artery is usually absent when the right lung has failed to form, the combination of unilateral right pulmonary agenesis and left pulmonary arterial sling is extremely rare.

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Weber et al.[5] reported five patients, with ages ranging from 2 to 6 months, and with congenital tracheal stenosis and unilateral pulmonary agenesis. Their initial investigations consisted of a chest radiograph, computed tomographic scan, and bronchoscopy. Each child deteriorated acutely after the bronchoscopy. Endotracheal intubation was performed in two, followed by repair within 12 hours. The other three had more severe tracheal stenosis, with an abrupt deterioration that needed emergency intervention. An emergency tracheostomy was carried out on one, an urgent thoracotomy in another, and the third required extra-corporeal membrane oxygenation prior to repair 48 hours later. Operative repair consisted of segmental resection and anastomosis in one patient, while the other four patients had rib-cartilage tracheoplasty. Of the five patients, two died, one secondary to pneumonia, and the other due to tracheitis and aorto-tracheal fistula.

Unilateral pulmonary agenesis is associated with other congenital defects, such as hemivertebras and hemifacial microsomia, cardiac malformations which are primarily septal defects, and occasionally with tracheal abnormalities.[4] Mardini et al.[6] reported 4 patients with isolated pulmonary agenesis. All had parental consanguinity, suggesting an autosomal recessive mode of inheritance. The combination of agenesis of the right lung, left pulmonary arterial sling, and hypoplasia of a long segment of the trachea, as seen in our patient, is rare. To our knowledge, only six cases have been reported in the English literature.[4] Of those 6 cases, three had funnel shaped tracheas with complete tracheal rings, while in four patients, external compression by the left pulmonary artery and aorta was noted. Some form of obstruction is invariably present in the airways, either due to intrinsic tracheal stenosis or external compression by the aorta anteriorly or the vascular sling posteriorly. Although two of the patients reported by Pu et al.[4] underwent cardiac catheterization, the authors concluded that "accurate diagnosis can be achieved noninvasively by echocardiography and by computed tomography or magnetic resonance imaging, avoiding the need for cardiac catheterization". Interestingly, one of their reported cases underwent surgical repair of the tracheal stenosis and mobilization of the left pulmonary artery. Suprasystemic pulmonary arterial pressures encountered postoperatively, however, prevented separation from cardiopulmonary bypass, and the patient died.

In our patient, the aetiology of the abnormal pulmonary vasculature is unknown. We can speculate that the hypoplasia of the left pulmonary artery may have resulted in inadequate flow to the "normal" right lung during fetal life, or it may have resulted from multiple pulmonary embolisation from two periods of extra-corporeal membrane oxygenation. Although we were able correctly to diagnose the cardiac and vascular anatomy using echocardiography, cardiac catheterization was essential more clearly to identify the pathology. The findings on cardiac catheterization precluded further surgical intervention to repair the tracheal stenosis and left pulmonary arterial sling. In the light of our experience, we recommend catheterization and pulmonary angiography prior to surgical treatment of this rare anomaly.

References

1. Kwan Kim H, Tae Kim Y, Whan Sung S, et al. Management of congenital tracheal stenosis. Eur J Cardiothorac Surg 2004; 25: 1065–1071.
2. Grillo HC, Wright CD, Vlahakes GJ, MacGillivray TE. Management of congenital tracheal stenosis by means of slide tracheoplasty or resection and reconstruction, with long-term follow-up of growth after slide tracheoplasty. J Thorac Cardiovasc Surg 2002; 123: 145–152.
3. Lin JH, Chen SJ, Wu MH, Wang JK, Li YW, Lue HC. Right lung agenesis with left pulmonary artery sling. Pediatr Pulmonol 2000; 29: 239–241.
4. Pu WT, Chung T, Hoffer FA, Jonas RA, Geva T. Diagnosis and management of agenesis of the right lung and left pulmonary artery sling. Am J Cardiol 1996; 78: 723–727.
5. Weber TR, Connors RH, Tracy TF Jr. Congenital tracheal stenosis with unilateral pulmonary agenesis. Ann Surg 1991; 213: 70–74.
6. Mardini MK, Nyhan WL. Agenesis of the lung. Report of four patients with unusual anomalies. Chest 1985; 87: 522–527.