Clinical and experimental aspects of tracheal stenosis
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CHAPTER 3b
Right Aortic Arch, Right Ligamentum, and Absent Left Pulmonary Artery: A Rare Form of Vascular Ring

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Abstract

A 5-month-old infant presented with respiratory failure secondary to severe right bronchial compression. Diagnostic imaging revealed a right aortic arch and absent left pulmonary artery. Surgical relief was obtained via median sternotomy by dividing a right ligamentum and pexing the enlarged right pulmonary artery to the ascending aorta.
There are very few reports of vascular rings that can not be addressed through a left thoracotomy. We report the rare combination of a right aortic arch, right ligamentum arteriosum, and absent left pulmonary artery in a 5-month-old infant that presented with acute respiratory failure. Through a median sternotomy, a tight right ligamentum arteriosum (acting like a "bow-string") and an enlarged right pulmonary artery were found compressing and nearly occluding the right mainstem bronchus. Relief of the right bronchial compression was obtained by dividing the ligamentum and pexing the right pulmonary artery to the aorta.

**Case Report:**

A 5-month-old girl presented with a 2 day history of clear rhinitis, coughing, wheezing, and tachypnea without fever. Chest x-ray showed a hyperinflated right lung and right upper and lower lobe atelectasis. Progressive respiratory failure despite bronchodilators led to endotracheal intubation and mechanical ventilation. Progressive respiratory acidosis motivated a transfer to our institution. Initial arterial blood gas was (pH 7.17, pCO$_2$ of mmHg, pO$_2$ 84 mmHg) on an FiO$_2$ of 70%. Bronchoscopy demonstrated extrinsic anterior nonpulsatile compression of the right mainstem bronchus, causing nearly complete occlusion of this bronchus. The trachea and left main bronchus were normal. The bronchoscope did pass into the right main bronchus and there was purulent material suctioned from the distal bronchus. Computerized tomography scan of the chest revealed a right aortic arch, compression of the right main bronchus, and the left pulmonary artery could not be identified. An echocardiogram was performed to rule out pulmonary artery sling. This demonstrated normal intracardiac anatomy but again no evidence of a left pulmonary artery. Cardiac catheterization with simultaneous contrast tracheo-bronchography demonstrated a large right pulmonary artery with compression of the right mainstem bronchus between the pulmonary artery anteriorly and the right-sided descending thoracic aorta posteriorly (Figures 1,2,3). A retrograde injection of the left pulmonary veins revealed a tiny intraparenchymal left pulmonary artery which did not communicate with the main pulmonary artery.

Without a precise preoperative diagnosis, we elected to approach the right bronchial compression through a median sternotomy. The pericardium was opened and a right aortic arch noted. There was no evidence of a left sided ligamentum arteriosum or intrapericardial left pulmonary artery. A ligamentum arteriosum was identified coming off the postero-superior aspect of the right pulmonary artery and inserting into the right descending thoracic aorta.
Figure 1 Aortogram demonstrates a right aortic arch, mirror image branching of the brachiocephalic vessels, and a ductal “bump” which was consistent with the noted intraoperative right ligamentum arteriosum.

Figure 2 Contrast bronchogram with simultaneous levophase aortogram demonstrating a normal trachea and left main bronchus, with severe narrowing of the right mainstem bronchus at a site between the right descending aorta and right pulmonary artery.

Figure 3 Pulmonary angiogram illustrates a large right pulmonary artery, no left pulmonary artery, and no evidence for a pulmonary artery sling.

The right ligamentum was doubly ligated and divided. The right pulmonary artery was fully mobilized to rule out a pulmonary artery sling. Intraoperative bronchoscopy was performed and visualized a now improved opening of the right main bronchus. This opening was further improved (to near normal) by suspending the right pulmonary artery to the posterior aspect of the ascending aorta. Peak airway pressures required for adequate ventilation dropped
from 42 mmHg to 28 mmHg. The patient was extubated on the first postoperative day. Bronchoscopic follow-up 2 weeks, 3 months, and 10 months post-surgery revealed a slightly "flattened" but patent right mainstem bronchus and the child has continued to be symptom-free except for rare intermittent episodes of reactive airway disease.

**Discussion:**

We recently reported our experience with nearly 300 patients with vascular rings over a 50-year time span. In that series this is the first child with a right aortic arch, right ligamentum, and absent left pulmonary artery. Prior to this all of the patients with a vascular ring caused by a right aortic arch (79 patients) had the ring completed by a left ligamentum. In this patient, there was not a true "complete" ring, rather the right ligamentum acting like a "bow-string" along with the enlarged RPA. The RPA was enlarged because of the absent LPA, with the entire cardiac output passing through the RPA. This has been called a "hemodynamic vise." We have categorized patients such as this with those with "complete" vascular rings because of the similarities in clinical presentation, diagnostic techniques, and surgical intervention.

Robotin and colleagues from Paris reported an experience of more than 500 infants and children undergoing operation for symptomatic tracheo-bronchial compression. They described a single patient with a right aortic arch and right descending aorta that had compression of the right main bronchus. They alleviated the bronchial compression by mobilizing the descending aorta through a right thoracotomy. That child did have a left pulmonary artery and had a patent ductus arteriosus, presumably on the left side (this was not stated).

Döhlemann and colleagues reported 3 infants with hypoplasia or aplasia of the right lung, dextrocardia, and compression of the trachea by the ascending aorta as it traversed from right to left. In a sense, these are "mirror images" of our patient, the difference being that despite having an absent left pulmonary artery, our patient had a normal-sized left lung and levocardia, hence the great vessel orientation was essentially normal for a right arch.

Despite a thorough preoperative work-up, failure to precisely determine the nature of the compression was understandable given the unusual pathology. We therefore elected a median sternotomy approach. This allowed direct visualization of any type of vascular ring and also permits cardiopulmonary bypass, if necessary, for repair of pulmonary artery sling. Intraoperative bronchoscopy is an invaluable diagnostic tool that allows direct assessment of the extrinsic compression of the Airways, and documents the adequacy of repair. This case
Chapter 3b

illustrates how multiple diagnostic modalities may be necessary in the unusual patient with a vascular ring that does not fall into the "neat" diagnostic categories of double aortic arch, right aortic arch with left ligamentum, innominate artery compression syndrome, and pulmonary artery sling. Although the usual tendency is to obtain too many studies which simply continue to verify the diagnosis, when the diagnostic studies are not typical it is important to continue investigation as far as possible to insure appropriate surgical intervention. In this case, the final (surgical) diagnosis led to successful airway relief by combining division of the right ligamentum arteriosusm and pexing of the right pulmonary artery to the aorta.

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References
