Ductal Arteriosus Aneurysm, Right Aortic Arch, and Isolated Left Subclavian Artery in a Neonate

Claudeen K.F. Scott, MBBS,* David Meyer, MD,† Colin K.L. Phoon, MPhil, MD,* and Monvadi B. Srichai, MD‡

*Pediatric Cardiology Program and Department of Pediatrics, †Department of Cardiothoracic Surgery, ‡Cardiac Radiology Section, Department of Radiology, New York University School of Medicine, New York, NY, USA

ABSTRACT

Ductal arteriosus aneurysm (DAA) is a well-recognized condition, especially in infancy, and is usually asymptomatic. We report the first case of a newborn who presented with significant inspiratory stridor and, using multiple imaging investigations, was subsequently diagnosed with the rare constellation of a congenital DAA, a right aortic arch and an isolated left subclavian artery with normal intra-cardiac anatomy. The patient underwent surgical resection of the DAA with significant improvement in symptoms.

Key Words. Ductal Arteriosus Aneurysm; Subclavian Artery; Recurrent Laryngeal Nerve Palsy; Aortic Arch

Introduction

Ductal arteriosus aneurysm (DAA) may be congenital or acquired as a complication of surgical patent ductus arteriosus closure. A right-sided aortic arch is usually reported in association with a congenital cardiovascular defect, the most common being Tetralogy of Fallot.[1–3] Complete evaluation of the cardiovascular anatomy is important for medical and surgical management. We report the first case of a newborn who presented with significant inspiratory stridor and using multimodality imaging, was subsequently diagnosed with the rare constellation of a congenital DAA, a right aortic arch, and an isolated left subclavian artery (LSA) with normal intracardiac anatomy, and who underwent surgical treatment for amelioration of vocal cord paresis.

Case Report

A full-term male neonate was noted to be stridorous and cyanotic at birth. Despite continuous positive airway pressure ventilation, he continued to be symptomatic. Initial chest X-ray, neck X-ray, and ultrasound of neck were reported as normal. Arterial blood gas revealed hypoxemia with room air oxygen saturation of 91%. Laryngoscopy revealed bilateral laryngeal nerve paresis. Transthoracic echocardiography revealed a right aortic arch with a large right-sided DAA (8 mm in diameter), twice the size of the aorta (Figure 1). The remaining cardiovascular structures were normal. However, because the possibility of a vascular ring still remained, a computed tomography angiography scan of the chest was performed. Computed tomography angiography demonstrated a large ductal aneurysm with displacement and moderate compression of the right main and upper lobe bronchi. No vascular ring was identified. Incidentally noted was a proximal blind ending LSA (Figure 2). Cardiovascular magnetic resonance was performed to determine flow dynamics. This study confirmed an isolated LSA, which filled through retrograde flow in the left vertebral artery (Figure 3). On day 8 of life, the patient underwent resection of the DAA to relieve the airway obstruction and potentially to improve vocal cord movement by decreasing pressure on the right recurrent laryngeal nerve (Figure 4). Postoperatively, stridor and the right vocal cord movement improved. Recent laryngoscopy demonstrated normal right vocal cord movement.

Discussion

DAA is extremely rare in association with a right aortic arch and an isolated LSA particularly in a patient with no known chromosomal abnormalities or other intracardiac anomaly. It is thought to occur because of post-stenotic dilation of the

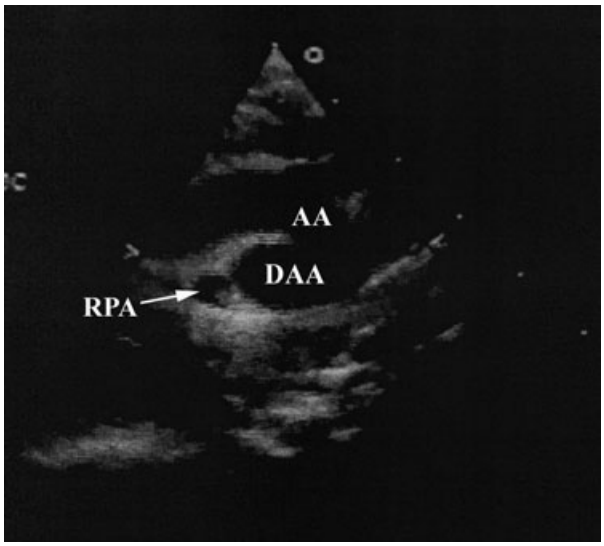

Figure 1. Transthoracic echocardiogram in the suprasternal view demonstrates the aortic arch with ductus arteriosus aneurysm. AA, aortic arch; DAA, ductal arteriosus aneurysm; RPA, right pulmonary artery.

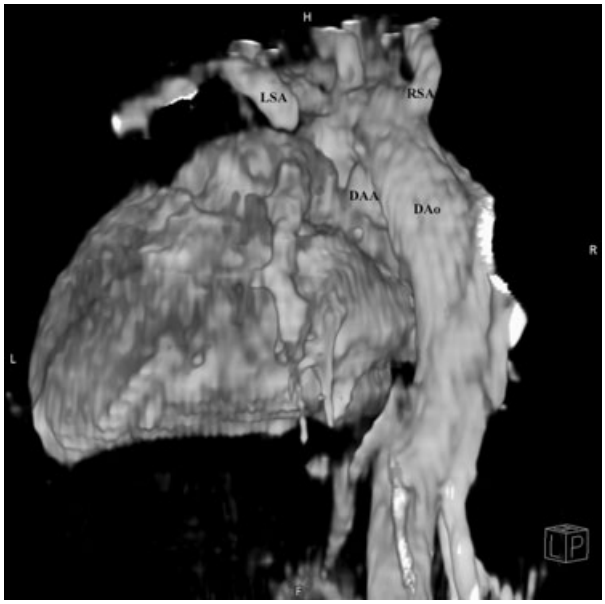

Figure 2. Three-dimensional computed tomography angiography volume rendered reconstruction visualized from the left posterior view demonstrates the ductal arteriosus aneurysm, the isolated left subclavian artery, and the right aortic arch. DAA, ductal arteriosus aneurysm; DAo, descending aorta; LSA, left subclavian artery; RSA, right subclavian artery.

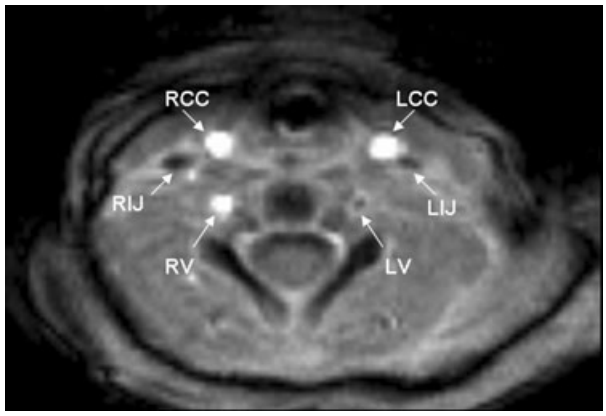

Figure 3. Magnetic resonance images using time of flight sequence at the level of the neck demonstrates directionality of blood flow in the neck vessels with bright areas (RCC, RV, LCC) within the arteries representing blood flow toward the head, and dark areas (RIJ, LIJ) within the veins representing blood flow toward the chest. The flow in the left vertebral artery is in the same direction as the veins representing retrograde flow within this artery. LCC, left common carotid artery; LIJ, left internal jugular vein; LV, left vertebral artery; RCC, right common carotid artery; RIJ, right internal jugular vein; RV, right vertebral artery.

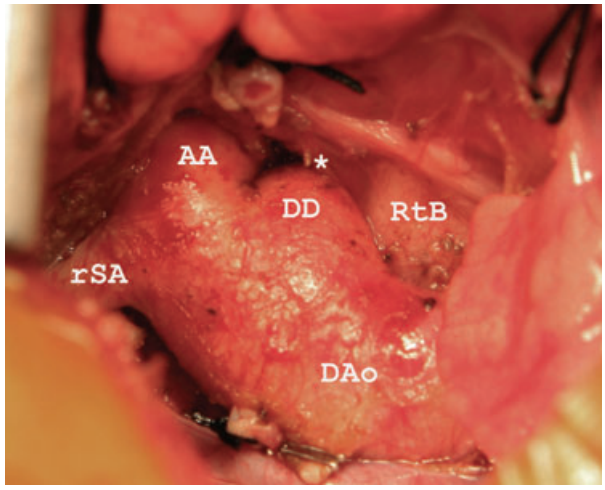

Figure 4. Operative photograph during a right thoracotomy demonstrating the right aortic arch and large right ductal arteriosus aneurysm compressing the right recurrent laryngeal nerve (*) and the right bronchus. AA, aortic arch; DAA, ductal arteriosus aneurysm; DAo, descending aorta; rSA, right subclavian artery; Rtb, right bronchus.

ductus from turbulent flow through a stenotic segment at its pulmonary artery end during fetal life or during perinatal asphyxia.[4] Only 0.8% of patients with a right aortic arch have a concomitant isolated LSA.[2] An isolated LSA is one in which the artery loses continuity with the aortic arch and instead is connected to the left pulmonary artery by a ductus, which may be patent or closed.[5] Although DAA often resolves spontaneously, complications such as rupture, vocal cord

paralysis, endocarditis, or mass effect on surrounding structures may develop,[4,6] leading to medical and/or surgical intervention.

This case demonstrates the value of multimodality imaging in defining the unusual branching patterns of the aortic arch. Although anatomic diagnosis can be made with computed tomography angiography, cardiovascular magnetic resonance can provide additional information on flow dynamics without radiation exposure. An anatomical reason for the stridor in this neonate (the DAA) had already been diagnosed, but further investigations for the presence of a vascular ring lead to the discovery of the isolated LSA. Our patient is currently asymptomatic, thus no surgical intervention is indicated at present. However, the patient is being monitored clinically for signs and symptoms of subclavian steal syndrome as an isolated LSA may cause vertebrobasilar insufficiency and limb ischemia in adulthood.

Surgical intervention for DAA is the treatment of choice when DAA persists beyond 4–6 weeks.[7] Early surgical resection is considered for patients who are symptomatic or who develop associated complications. Many surgeons believe that if DAA does not spontaneously resolve within a few days, then it should be resected because the rate of complications in infants less than 2 months has been reported as high as 31%.[3,8–10] Our patient had significant vocal cord paresis and airway compression which was successfully relieved with resection of the DAA.

This case emphasizes the need for meticulous attention to the aortic branching pattern in any patient with a right aortic arch, more so if a DAA is present. If vascular anatomy is not well defined by echocardiography, then additional diagnostic imaging should be performed. This is especially true for patients who present with stridor, respiratory distress, and/or laryngeal cord paralysis.

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Corresponding Author: Mondavi B. Srichai, MD, FACC, Department of Radiology, New York University School of Medicine, New York, NY 10016, USA. Tel: (+1) 212-263-0144; Fax: (+1) 212-263-8186; E-mail: srichm01@med.nyu.edu

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