Title: Anomalous Origin of the Left Pulmonary Artery from the Ascending Aorta in a Patient with DiGeorge Syndrome and Right Aortic Arch

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Abstract:
Anomalous origin of one pulmonary artery from the ascending aorta (AOPA), also known as hemi-truncus, is an extremely rare congenital heart malformation first described in 1868.[1]This defect manifests with early pulmonary hypertension by two unique mechanisms: (1) one lung receiving an obligate entire cardiac output from the right ventricle and (2) systemic pressure driving systemic arterial blood into the lung with the anomalous origin of the branch pulmonary artery.[2] Early surgical repair with direct implantation of the anomalous artery into the main pulmonary artery has provided excellent results.[2] While anomalous aortic origin of either the right or left pulmonary artery from the ascending aorta can occur, the frequency and cardiac associations of each have led some to believe that they have separate embryologic considerations.[3] Anomalous aortic origin of the right pulmonary artery is approximately 4 to 8 times more common.[4]

There are only two reported patients with DiGeorge syndrome and anomalous aortic origin of the left pulmonary artery (AOLPA), both described by Dodo Et al in 1995[4]. Here we present a third case of AOLPA associated with Di-George deletion syndrome.

Case Report:
A 3.99 kg, 7-week old girl with recently diagnosed bronchiolitis and uncomplicated birth history presented for evaluation of heart murmur. ROS was positive for mild tachypnea, but otherwise negative. There was no family history of congenital heart disease.

Physical exam was pertinent for a peripheral oxygen saturation of 95%, a grade III/VI systolic ejection murmur best heard at the left upper sternal border that radiated to the bilateral axilla and back and clear lung fields. Echocardiogram demonstrated the left pulmonary artery (LPA) arising from the leftward aspect of the ascending aorta while the anatomy of the right pulmonary artery was normal. (Image 1) The right ventricle was mildly dilated and hypertrophied with a flattened interventricular septum. The aortic arch was not well imaged on initial imaging. The right ventricular pressures were estimated to be slightly sub-systemic.

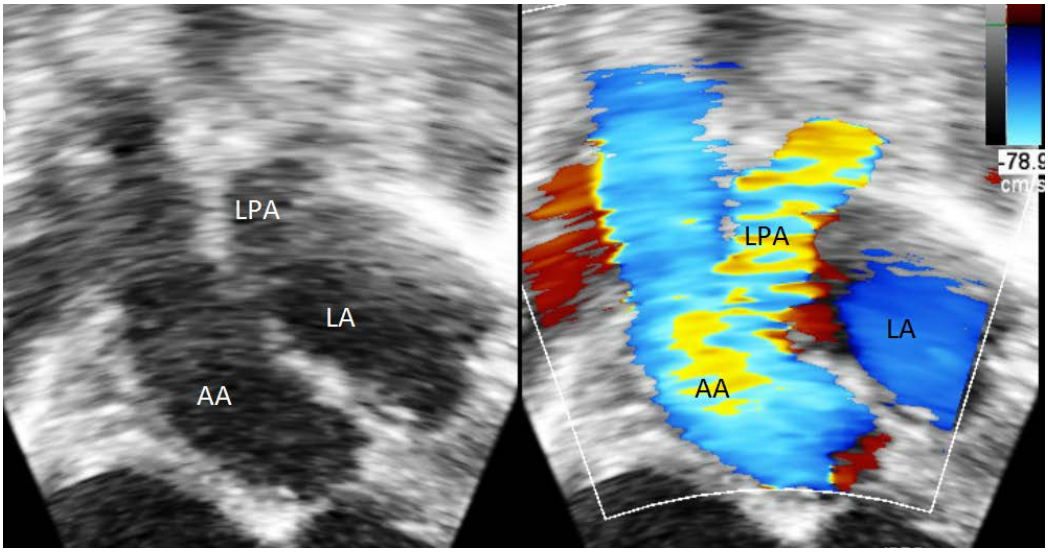

Image 1: Pre-surgical echocardiogram with color flow, apical view with anterior angulation, reveals ascending aorta giving rise to left pulmonary artery. AA- Ascending aorta, LPA- Left Pulmonary Artery, LA- Left Atrium.

The patient was referred for urgent surgical correction. She underwent surgical implantation of the LPA to the main pulmonary artery (MPA) with pericardial patch augmentation of the connection. (Image 2) Inspection of the great arteries revealed a right sided aortic arch with possible aberrant left subclavian artery and dilated MPA and RPA. No thymic tissue was visualized. After implantation of the LPA into the MPA, right ventricle and aortic systolic pressures were directly measured to be 25 and 75 mmHg, respectively. Approximately two and a half weeks after the procedure she developed post-pericardiotomy syndrome with pericardial effusion requiring a short course of steroids.

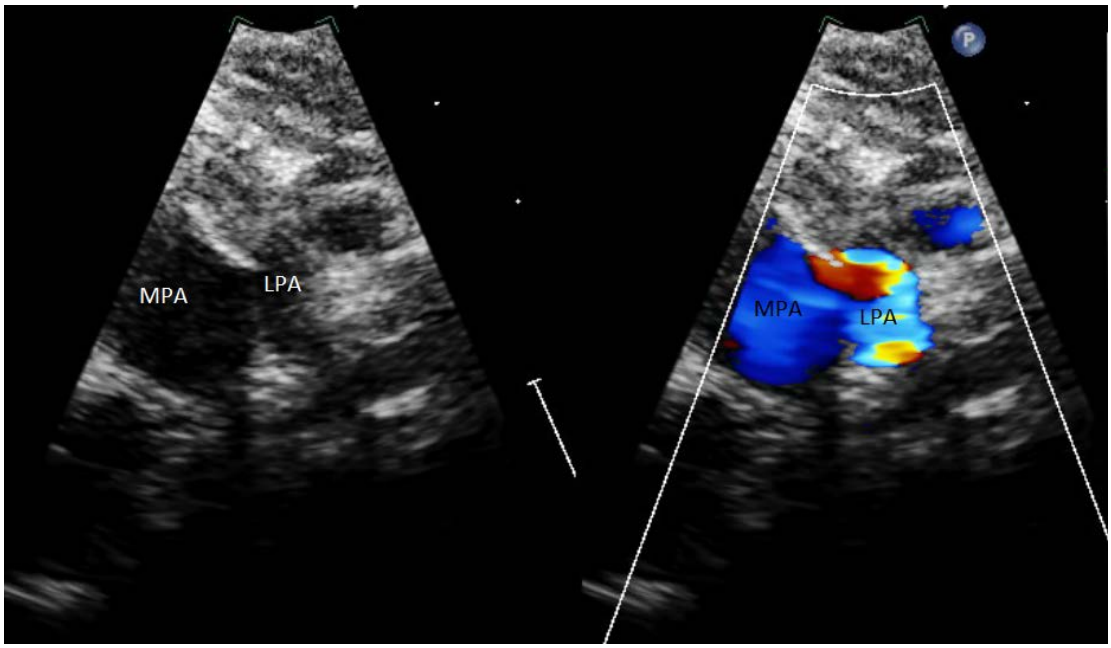

Image 2: Parasternal short axis color flow echocardiogram performed after intervention showing reimplantation of the left pulmonary artery to the main pulmonary artery. MPA-Main Pulmonary Artery, LPA-Left Pulmonary Artery

Genetic workup revealed 22q11.2 deletion syndrome. She continues to do well from a cardiopulmonary standpoint.

Discussion:

This is the third documented incidence of AOLPA in a patient with DiGeorge syndrome. As with the other two cases described by Dodo, our patient presented in the first two months of life with respiratory symptoms and had a right aortic arch. In each case there was good surgical outcome with the direct implantation of the anomalous left pulmonary artery to the main pulmonary artery. DiGeorge syndrome is associated with conotruncal abnormalities including tetralogy of Fallot, interrupted aortic arch and truncus arteriosus, among other rare associations. We now encourage clinicians to consider AOLPA in this small group of rare associations.[6] It is important that clinicians recognize this pattern when AOLPA is suspected or confirmed, especially in combination with right aortic arch. This may allow for earlier diagnosis of 22q11.2 deletion syndrome, leading to earlier multidisciplinary care. Furthermore, hypocalcemia and

hypomagnesemia are known complications of 22q11.2 deletion syndrome especially during times of biological stress and should be monitored closely in the perioperative period. Finally 22q11 fluorescence in situ hybridization (FISH) testing may be warranted for patients with AOLPA.

References:

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