Cardiology in the Young

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Major aortopulmonary collateral arteries in association with atrioventricular septal defect with balanced ventricles and trisomy 21

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Brief Report

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Abstract

A major aortopulmonary collateral artery is a rare and easily missed diagnosis that is usually associated with Tetralogy of Fallot or pulmonary atresia. We present two cases of major aortopulmonary collaterals associated with trisomy 21 and atrioventricular septal defect with balanced ventricles in which the diagnosis went undetected until after initial cardiac repair.

Major aortopulmonary collateral arteries are anomalous arteries that arise either directly from the aorta or a major branch of the aorta and supply a segment of the lung. They can be the sole supply of pulmonary blood flow to a segment of lung or a duplicated supply to the pulmonary arterial flow. These systemic-to-pulmonary connections are remnants of the embryonic ventral splanchnic arteries which usually regress when pulmonary circulation develops in-utero.[1]

Major aortopulmonary collateral arteries are a rare defect predominantly associated with Tetralogy of Fallot or pulmonary atresia but have in rare instances been reported in association with other congenital cardiac defects including total anomalous pulmonary venous connection, double outlet right ventricle, tricuspid atresia, corrected transposition of the great arteries, and atrioventricular septal defects with unbalanced ventricular size.[2–3]

Case details

A 6.7-kg male with postnatal diagnosis of trisomy 21 and Rastelli type C atrioventricular septal defect with balanced ventricles[4] underwent scheduled repair of his cardiac defect at 4 months of age. Prior to scheduled operation, he had symptoms of pulmonary over-circulation including intermittent tachypnoea, tachycardia, and difficulty gaining weight, but was successfully managed with furosemide as an outpatient. During operative repair, the surgeon noted substantial left atrial return without evidence of a patent ductus arteriosus, and on attempt to close the atrial septal defect, left atrial pressure increased to 21 mmHg by direct measurement. These findings prompted concern for a major aortopulmonary collateral artery, and the infant was taken directly to the cardiac catheterization lab. Catheterization revealed an anomalous artery that arose from the thoracic aorta just below the left diaphragm and provided the sole source of pulmonary blood flow to multiple lower lobe segments of the left lung (Fig 1a). The patient was stabilized in the cardiac ICU prior to being taken back for anticipated unifocalization of the major aortopulmonary collateral artery. Intraoperatively, the segments of the lung supplied by major aortopulmonary collateral arteries were found to be infarcted. The decision was made to ligate the collateral without reimplantation to the left pulmonary artery. After a prolonged hospitalization, he was discharged and is stable off cardiac medications at 11 months.

A second 4.2-kg female infant with a prenatal diagnosis of Rastelli type A atrioventricular septal defect with balanced ventricles and trisomy 21 underwent scheduled cardiac repair at 6 months. Pre-operatively she had symptoms of pulmonary over-circulation requiring outpatient medical therapy with furosemide, and she had two separate hospitalizations with acute respiratory failure attributed to transient tachypnoea of the newborn and respiratory syncytial virus bronchiolitis, respectively. Her initial post-operative course was at a referring institution where full repair was performed and uncomplicated. However, on post-operative day 7, she acutely decompensated with respiratory distress that progressed to cardiorespiratory arrest requiring epinephrine and resuscitation efforts for 10 minutes. She was re-cannulated onto extracorporeal membrane oxygenation for low cardiac output. At this time, her right ventricle was noted to be severely dilated, concerning for supra-systemic right-sided heart pressures and pulmonary hypertension. She was taken to the cardiac catheterization lab, and a large major aortopulmonary collateral coming off the thoracic aorta and serving as the sole supply of blood flow to the

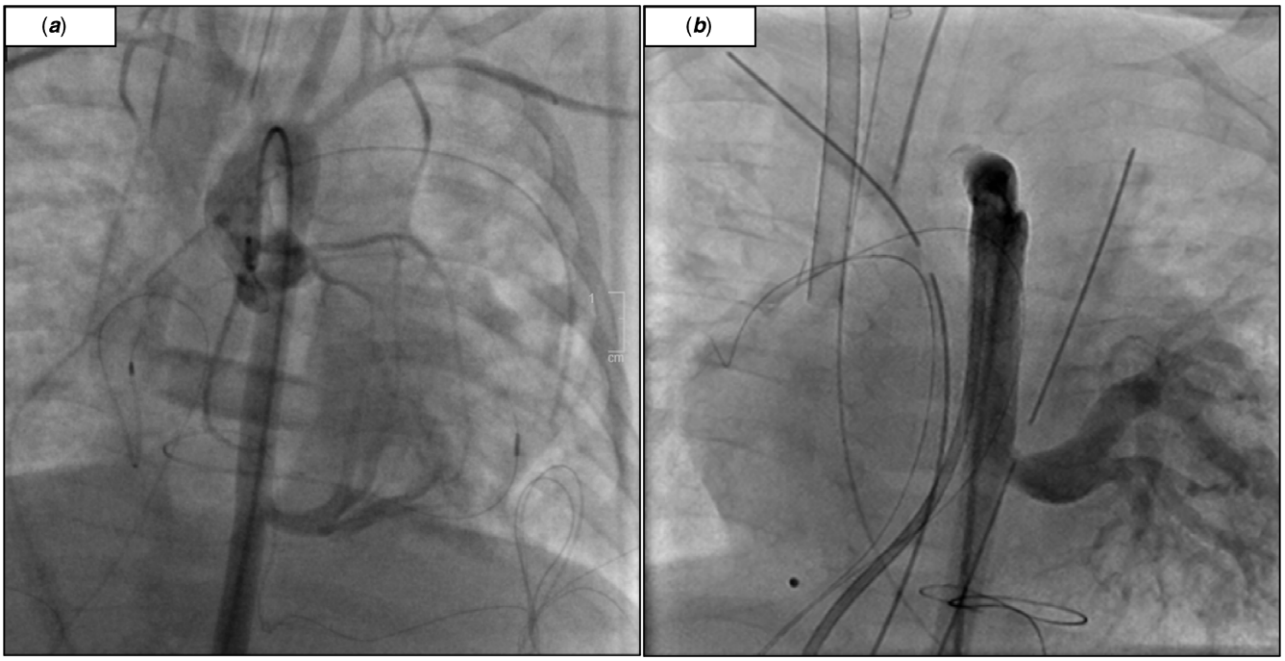

Figure 1. (*a*) Major aortopulmonary collateral originating from the abdominal aorta near the diaphragm. (*b*) Major aortopulmonary collateral artery coming off the descending thoracic aorta to supply the inferior segments of the left lung.

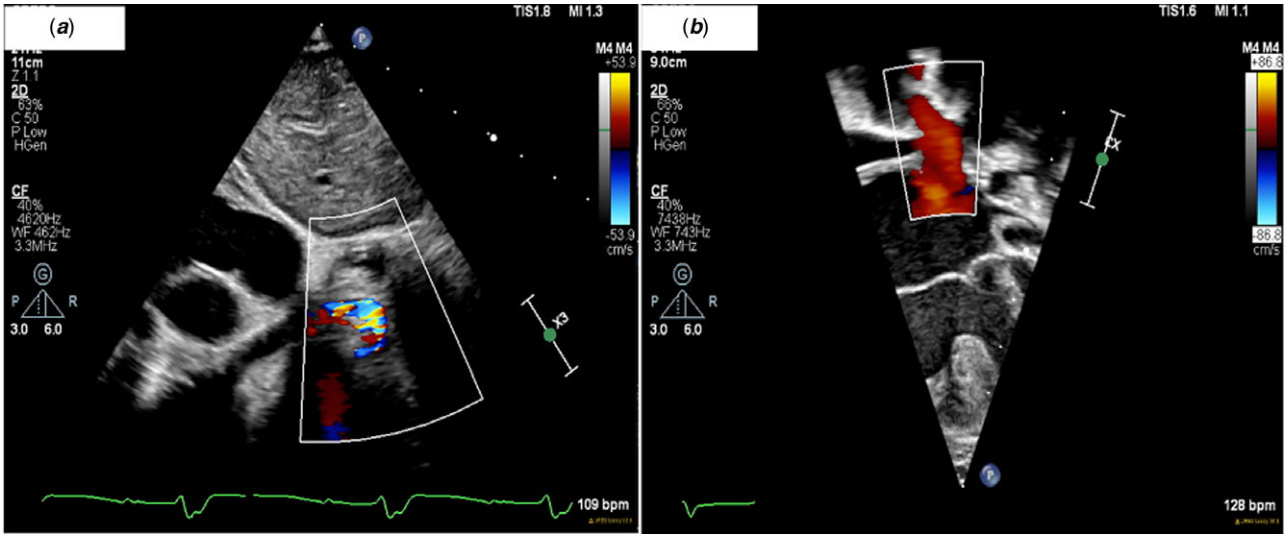

Figure 2. (*a*) Aortopulmonary collateral from the abdominal aorta. (*b*) Increased pulmonary venous return as demonstrated by the high Nyquist limit.

lower lobe of the left lung was identified (Fig 1b). Two days later, she underwent successful unifocalization of the major aortopulmonary collateral artery to her left pulmonary artery. This was done as a primary connection, and a triangular patch of autologous pericardium was used to augment the connection. Fifteen months post-operatively, she remains in good condition off supplemental oxygen.

Discussion

Diagnosis of major aortopulmonary collateral arteries, when not in association with Tetralogy of Fallot, can be challenging, as they are unanticipated, and clinical presentation can be confounded by symptoms of associated cardiac defects. If the collateral artery originates in the descending thoracic aorta below the diaphragm, it can be easily missed by standard echocardiographic evaluation.[3] Evaluation for major aortopulmonary collateral arteries, particularly color evaluation of the abdominal aorta, should be included in patients with atrioventricular septal defects.

Though major aortopulmonary collateral arteries may be difficult to visualize by echocardiography (Fig 2a), down-stream effects of the increased pulmonary blood flow, such as right ventricular hypertrophy or dilation, left atrial dilation, and increased pulmonary venous return (Fig 2b) can be seen on echocardiogram. Both the patients presented had right ventricular hypertrophy on echocardiogram done prior to their cardiac repairs. This, however, was confounded by their intracardiac left to right shunt.

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Aortopulmonary collaterals can be diagnosed by visualization in the cardiac catheterization lab or by computed tomography angiography. Historically cardiac catheterization had been considered the gold standard for diagnosis. With advances in CT scanner technology, studies have demonstrated that CT angiography is equivalent to catheterization in detection and delineation of major aortopulmonary collateral arteries.[5]

In this series, we described a rare association of atrioventricular septal defect with balanced ventricles and major aortopulmonary collaterals in two individuals with trisomy 21. Both patients had multiple echocardiograms performed in which the major aortopulmonary collateral arteries were not detected by the routine imaging protocol. In the first case, the substantial left atrial return noted at time of repair and the rising left atrial pressures with repair of the atrial septal defect are what led to concern for a major aortopulmonary collateral. In the second case, the patient went to the catheterization lab for haemodynamic evaluation because of suspicion for pulmonary hypertension. She was found to have the aortopulmonary collateral. In both cases, the diagnosis was confirmed with visualization on cardiac catheterization.

Conclusion

Though major aortopulmonary collateral arteries generally occur in association with pulmonary atresia or tetralogy of Fallot, they can occur in conjunction with other cardiac defects as well. Signs and symptoms may become more apparent once other cardiac defects are repaired. The diagnosis should be suspected when clinical symptoms seem out of proportion for the intracardiac physiology, or when echocardiographic findings cannot be explained from the known cardiac defect. Diagnosis can be confirmed using either cardiac catheterization or CT angiography.

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Conflicts of interest. None.

Ethical standards. This case report does not involve human and/or animal experimentation.

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