CONGENITAL AORTOPULMONARY WINDOW; AN UNUSUAL CAUSE OF BREATHLESSNESS

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Having recently moved to the UK, a 27 year old female with known uncorrected congenital heart disease was referred for cardiovascular magnetic resonance (CMR) imaging due to increased shortness of breath. CMR imaging revealed a large (40mm) aortopulmonary window (Figure 1) with complete mixing of the pulmonary and systemic circulations (SaO₂ 73%), moderate mixed aortic valve disease and mild-moderate pulmonary valve regurgitation. Left and right ventricular volumes and systolic function were normal.

An aortopulmonary window is a communication between the ascending aorta and the pulmonary trunk. It is distinct entity from truncus arteriosus due to the presence of two separate arterial valves rather than a single arterial valve and is one of the rarest congenital cardiac lesions, accounting for under 0.1% of all congenital heart defects. [1] Associated vascular lesions are common with anomalies of the aortic arch being the most prevalent including; interruption, coarctation, isthmal hypoplasia, double arch, and right aortic arch. Cardiac lesions are also commonplace including; ventricular septal defect, atrial septal defect and left and right ventricular outflow valve stenoses. [2]

In view of the fact that irreversible pulmonary hypertension and premature death are the natural history of uncorrected left-to-right shunt physiology in the setting of aortopulmonary window surgical correction, which is the gold standard of treatment, is usually offered to patients at the time of diagnosis before the development of lung injury and irreversible pulmonary hypertension.

Figure Legend

Figure 1 (A) A coronal view and (B) a transverse view of normal proximal aortic and pulmonary artery anatomy compared to (C) a coronal view and (D) a transverse view of the large congenital aortopulmonary connection marked by a * (RV – Right Ventricle, MPA - Main Pulmonary Artery).

Contribution

OR Scanned and analysed the data, MB Drafted the manuscript, SM reviewed the images and aided in manuscript revision

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Competing Interests

None

References

1. Jacobs JP, Quintessenza JA, Gaynor JW, Burke RP, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: aortopulmonary window. Ann Thorac Surg 2000;**69**(4 Suppl):S44-9
2. Bagtharia R, Trivedi KR, Burkhart HM, et al. Outcomes for patients with an aortopulmonary window, and the impact of associated cardiovascular lesions. Cardiology in the young 2004;**14**(5):473-80 doi: 10.1017/S1047951104005025[published Online First: Epub Date]].