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A case of congenital pulmonary stenosis

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Introduction

We present an unusual case of sequential right ventricular outflow tract obstruction (RVOTO) caused by pulmonary and subpulmonary stenosis, with formation of aortopulmonary collaterals.

Case presentation

We report a case of a 47 year old Senegalese man who presented with a three year history of shortness of breath on exertion which worsened over the past week. Cardiovascular examination revealed a parasternal heave, a systolic murmur loudest in the pulmonary area and no evidence of fluid overload.

A transthoracic echocardiogram revealed leftward septal deviation during systole due to right ventricular pressure overload, severe right ventricular hypertrophy (RV free wall end-diastolic thickness of 8mm) and normal systolic function (FAC 47%), a dilated right atrium and moderate tricuspid regurgitation with estimated maximum pressures of 112mmHg. On continuous-wave doppler of the right ventricular outflow tract (RVOT), there was a late peaking systolic flow with maximum velocity of 3.8m/s.

A cardiac computed tomography (CT) and cardiac magnetic resonance imaging (MRI) was performed which showed sequential stenoses of the RVOT; one at subvalvular level by an infundibular muscular ridge with an area of 0.7 cm² in mid-systole and one at the level of a dome-shaped pulmonary valve with planimetered valve area of 0.5cm², severe RV hypertrophy and normal RV ejection fraction. Aortopulmonary collaterals from proximal thoracic descending aorta were seen. The main pulmonary artery was shown to be dilated with no evidence of pulmonary artery branch stenosis. See image: In-plane phase contrast velocity flow mapping of the RVOT using a VENC of 80cm/s, showing aliasing at the level of the pulmonary valve (red arrow) and at the level of the infundibulum (white arrow).

Conclusion

Congenital pulmonary stenosis (PS) occurs in 8% of congenital heart defects. 80% of pulmonary valve stenosis is typically dome-shaped with commissural fusion whilst the remainder is of the dysplastic type. Congenital PS can be associated with RVOTO at the infundibular level secondary to reactive muscular hypertrophy. In our case, there is a discrete, circumferential muscular ridge at the level of the infundibulum resulting in sequential outflow tract obstruction.

PS with intact ventricular septum occurs in conjunction with varying degrees of right ventricular hypertrophy and elevated right ventricular systolic pressures. A dilated pulmonary artery is common in dome-shaped subtype of PS.

Survival into adulthood of severe PS is primarily dependent on the adequacy of pulmonary blood flow from systemic-to-pulmonary collateral arteries, which serve as an additive, or the only source of blood supply to the
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Survival into adulthood of severe PS is primarily dependent on the adequacy of pulmonary blood flow from systemic-to-pulmonary collateral arteries, which serve as an additive, or the only source of blood supply to the pulmonary arterial vasculature. These collaterals are usually seen in association with cyanotic congenital heart disease such as pulmonary atresia and tetralogy of fallot.