Subpulmonary Obstruction from Aneurysmal Ventricular Septum

in a Child with Dextrocardia and Congenitally Corrected Transposition of the Great Arteries

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A 3-year-old boy underwent evaluation for dextrocardia. Echocardiograms showed features of corrected transposition physiology, a perimembranous ventricular septal defect (VSD) (Fig. 1), and aneurysmal tissue beneath

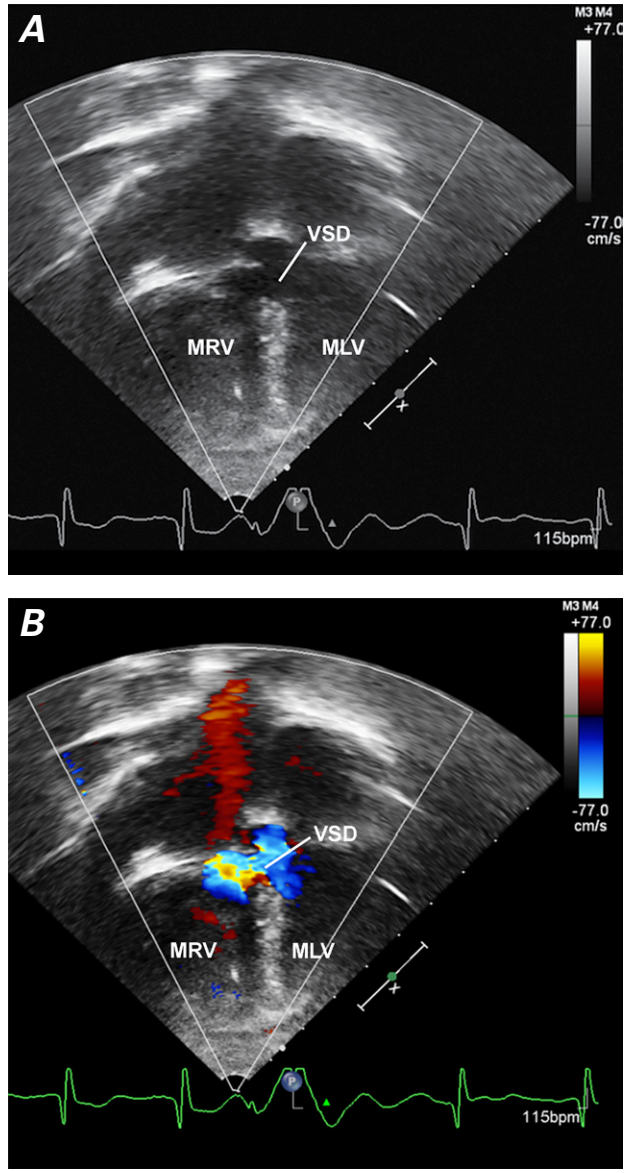

Fig. 1 *Two-dimensional echocardiograms **A**) in apical 4-chamber view from the right chest and **B**) in color-flow Doppler mode show a moderate-to-large ventricular septal defect (VSD).*

MLV = morphologic left ventricle; MRV = morphologic right ventricle

Supplemental motion image is available for Figure 1B.

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the pulmonary valve that caused severe subpulmonary stenosis (Figs. 2–4). Eighteen months after VSD closure and resection of the aneurysmal tissue, the patient was asymptomatic with only mild residual pulmonary outflow tract obstruction.

Comment

Congenitally corrected transposition of the great arteries (TGA) is usually associated with multiple cardiac defects. The hallmark finding is atrioventricular and ventriculoarterial discordance. Because of this double discordance, the circulatory physiology is normal: systemic venous return goes to the lungs, and pulmonary venous return goes to the body.[1,2] The usual anatomic arrangement is levocardia, visceroatrial situs solitus, L-loop ventricular inversion, and an anterior aorta on the left of the pulmonary artery {S,L,L}. Our patient's anatomy was rarer: dextrocardia with situs inversus, D-loop of the ventricles, and a rightward anterior aorta {I,D,D}.

Prolapsing aneurysms of a membranous ventricular septum rarely cause left ventricular (LV) outflow tract obstruction (pulmonary obstruction) in patients

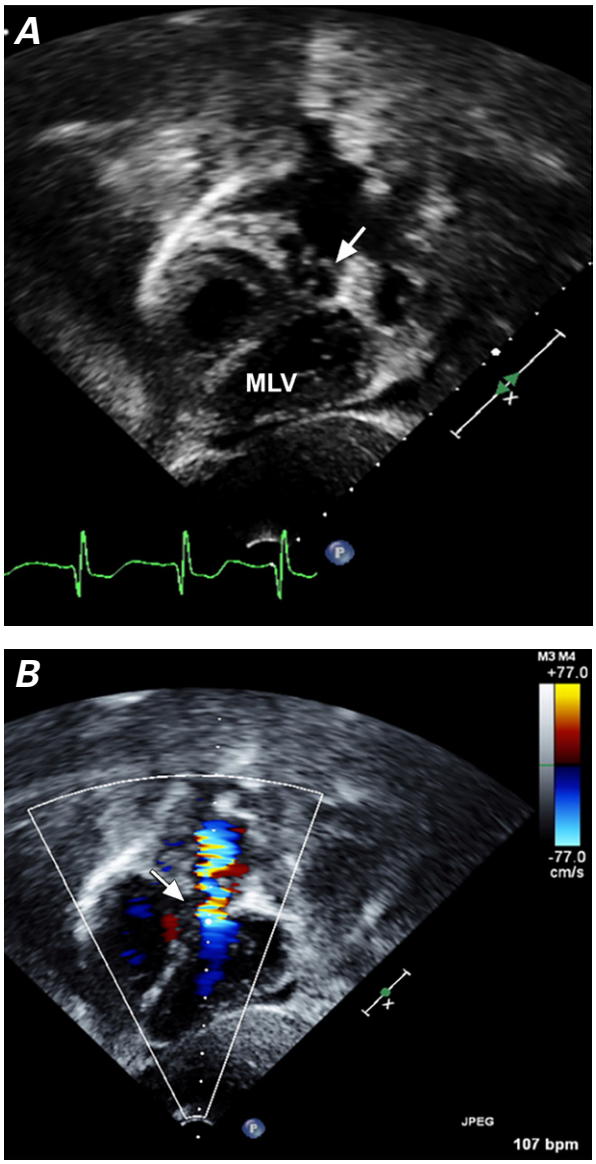

Fig. 2 *Echocardiograms (subcostal 4-chamber views).* ***A****) In the morphologic left ventricle (MLV), aneurysmal tissue (arrow) protrudes into the pulmonary outflow tract.* ***B****) Color-flow Doppler mode reveals turbulent flow (arrow) in the pulmonary outflow tract.*

Supplemental motion image is available for Figure 2A.

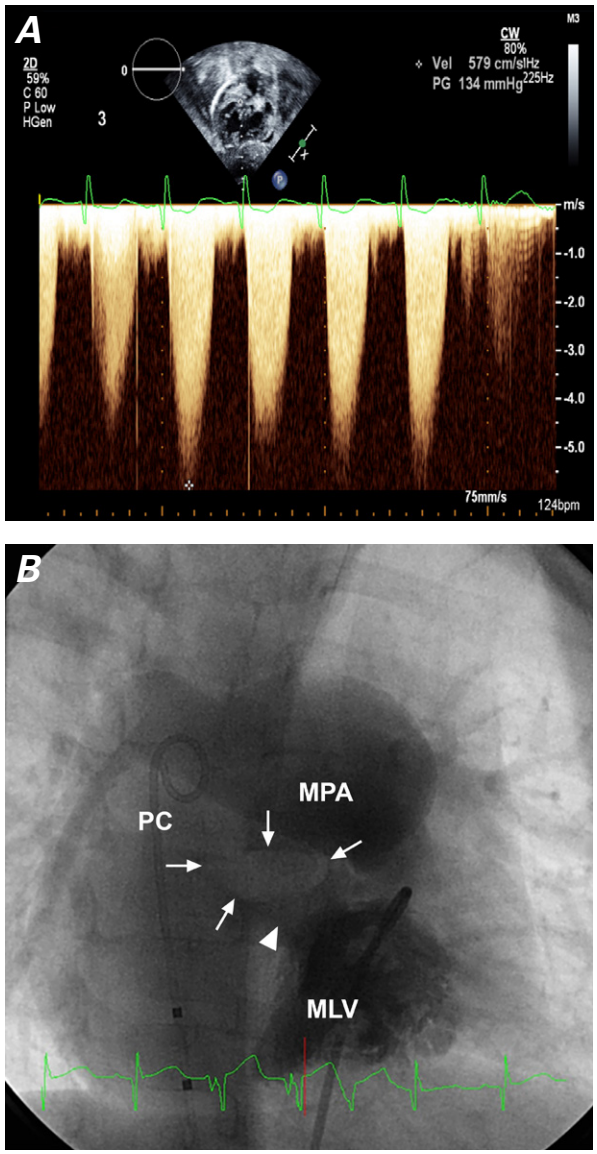

Fig. 3 ***A****) Continuous-wave Doppler echocardiographic recording across the pulmonary outflow tract shows a peak velocity >5 m/s, suggesting severe obstruction.* ***B****) Angiogram (60° left anterior oblique view) from the morphologic left ventricle (MLV) shows pulmonary outflow tract obstruction from aneurysmal tissue (arrows), the ventricular septal defect (arrowhead), and poststenotic dilation of the main pulmonary artery (MPA). A pigtail catheter (PC) is in the descending aorta.*

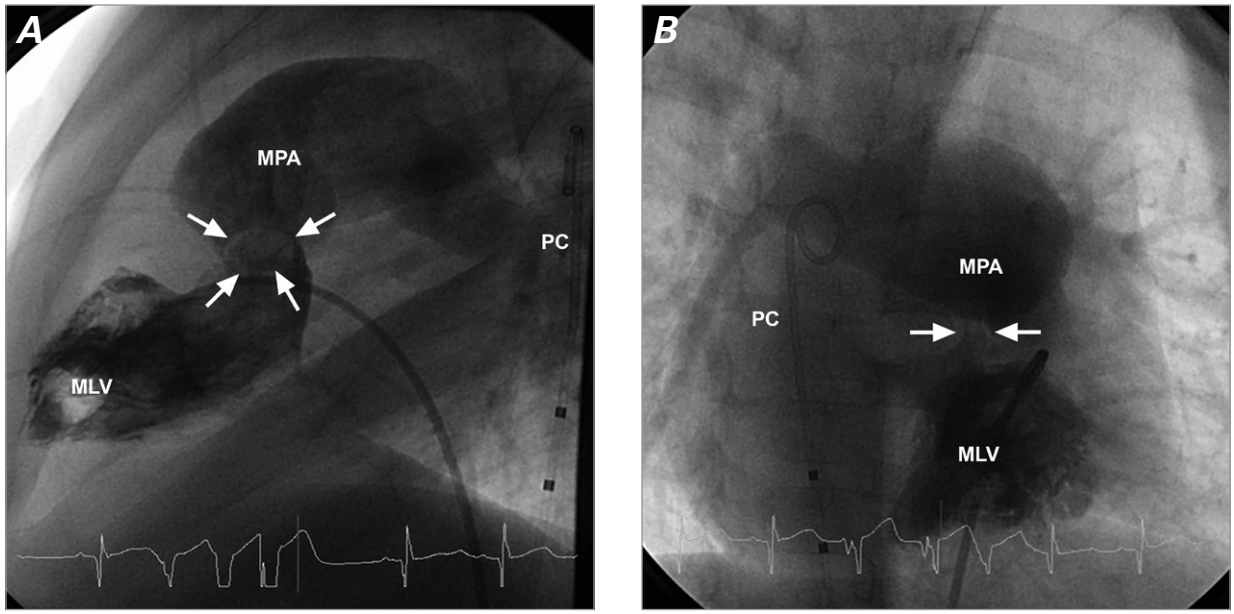

Fig. 4 *Cineangiographic image from the **A**) left lateral view reveals the morphologic left ventricle (MLV) and the pulmonary outflow tract obstruction from aneurysmal tissue (arrows). **B**) The 60° left anterior oblique view reveals a severely narrowed right ventricular outflow tract (arrows). Both frames show poststenotic dilation of the main pulmonary artery (MPA) and a pigtail catheter (PC) in the descending aorta.*

Supplemental motion image is available for Figure 4B.

who have normally related great vessels.[2-5] However, in patients with TGA who have higher right ventricular pressure, such an aneurysm can protrude into the LV outflow tract and cause pulmonary outflow tract obstruction.[6] Similarly, in patients with corrected TGA (who lack a conal septum and crista supraventricularis in the morphologic LV), even the proximity of a small aneurysm to the pulmonary valve can cause pulmonary outflow tract obstruction.

Doppler echocardiography and cardiac catheterization with selective cineangiography help to define the lesions and are the diagnostic tests of choice. Surgical aneurysm resection and VSD patch closure—with care to avoid injuring the vulnerable conduction system—is recommended.[2-5]

Acknowledgment

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