Anomaly of the Origin of the Left Coronary Artery in Children: Presentation as Mitral Valve Prolapse with Mitral Insufficiency and Normal Left Ventricular Function

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Key words: mitral valve prolapse, mitral insufficiency, anomalous origin of left coronary artery

IMAJ 2003;5:127–129

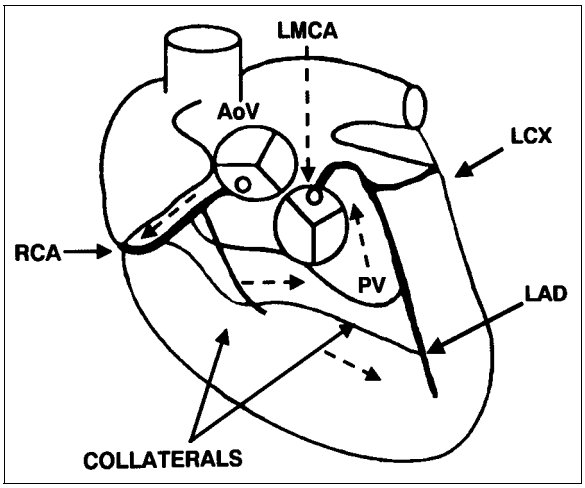
Anomalous origin of the left coronary artery from the pulmonary artery is a rare condition, accounting for 0.25–0.5% of all congenital heart malformations [1]. It causes the blood to flow away from the myocardium, with retrograde flow through the left coronary artery system into the pulmonary artery. The most common clinical presentation is severe congestive heart failure in early infancy, due to a diffusely dilated and poorly contracting left ventricle [2–4]. Occasionally, it may present later in childhood and sometimes in adulthood as angina, syncope or even sudden death [5]. In the latter cases, left ventricle contractility may be normal.

We describe four children who presented with mitral valve prolapse and mitral insufficiency relatively early in childhood, without impairment of left ventricular contractility. A comprehensive diagnostic evaluation demonstrated an abnormality of the origin of the left coronary artery in all four cases [Figure].

Patient Descriptions

Patient 1

A 10 year old boy had been referred for evaluation of a cardiac murmur at the age of 5 months. The clinical diagnosis of mitral valve prolapse and moderate mitral insufficiency was confirmed by two-dimensional echocardiography. The left atrium and left ventricle were mildly dilated but ventricular function was normal. Repeat two-dimensional echocardiographic studies demonstrated the left coronary artery



Anomalous origin of the main coronary artery from the pulmonary artery. AoV = aortic valve, PV = pulmonic valve, LMCA = left main coronary artery, LCX = left circumflex coronary artery, LAD = left anterior descending coronary artery, RCA = right coronary artery.

arising from the aorta. The electrocardiogram showed a persistently flat T wave in L1 and an inverted T wave in aVL.

The child developed normally and was asymptomatic. Over the course of follow-up, clinical signs of mitral regurgitation diminished. A color Doppler examination, which became available during the course of the follow-up, demonstrated some turbulent flow of unclear significance in the pulmonary artery. Cardiac catheterization was performed. An aortic root injection showed a dilated right coronary artery and retrograde filling of the left coronary artery, which emptied into the main pulmonary artery. The child underwent successful surgical re-implantation of the left coronary artery into the aorta. Follow-up color Doppler two-dimensional echocardiography demonstrated normal left ventricular function with negligible mitral insufficiency.

Patient 2

A 10.5 year old girl had been followed since the age of 18 months. At that time, a cardiac murmur was detected and mitral valve prolapse with mild to moderate mitral insufficiency was diagnosed by two-dimensional echocardiography. The patient was asymptomatic throughout follow-up, and the murmur decreased in intensity over time. At age 10 years she experienced a syncopal episode that was attributed to a vasovagal episode. However, an exercise stress test showed ST depression of 3 mm in lead aVF. Repeat echocardiography demonstrated mitral valve prolapse with mild mitral regurgitation. The origin of the left coronary artery was not well visualized. The right coronary artery was mildly dilated, but no pathologic flow was detected in the pulmonary artery. Cardiac catheterization was performed. A left ventriculogram revealed good ventricular function and mild mitral insufficiency with two narrow jets. An aortic root injection showed filling of an enlarged right coronary artery. A right coronary arteriogram demonstrated retrograde filling of a small left coronary artery, which ended blindly without connecting to the aorta or the pulmonary artery. This finding was consistent with a diagnosis of an atretic origin of the left main coronary artery. The child underwent successful coronary artery bypass surgery in which the left internal mammary artery was connected to the left anterior descending coronary artery. Follow-up color Doppler echocardiogram demonstrated minimal mitral insufficiency.

Patient 3

An 8 month old girl was referred for cardiac evaluation because of a heart murmur. Her medical history was unremarkable and she was asymptomatic and thriving. Physical examination showed a mildly active precordium, and on auscultation a pansystolic murmur of mitral regurgitation was heard. An electrocardiogram showed left atrial enlargement and inverted T waves in leads L1 and aVL. Echocardiography showed prolapse of both mitral valve leaflets with moderate mitral regurgitation and normal left ventricular function. The right coronary artery was enlarged and the left coronary artery was connected to the main pulmonary artery. Retrograde flow into the main pulmonary artery was seen by color Doppler. Cardiac catheterization confirmed the diagnosis and the patient underwent successful re-implantation of the left coronary artery into the aorta. A follow-up color Doppler echocardiogram 1 year after the operation demonstrated normal left ventricular function, but moderate mitral insufficiency was still evident.

Patient 4

A 5 year old girl underwent cardiac catheterization at the age of 3.5 years for evaluation of cyanosis. She was diagnosed as suffering from tetralogy of Fallot. In addition, there was mild to moderate mitral regurgitation and an enlarged left atrium. At age 5 she was referred for complete repair. Preoperative cardiac catheterization showed an anomalous left coronary artery arising from the pulmonary artery. The tetralogy was repaired through the right atrium, and the left coronary artery was ligated at its origin from the pulmonary artery. Cardiac evaluation by both two-dimensional echocardiography and cardiac catheterization several years after her operation revealed only mild mitral insufficiency and good left ventricular function.

Comment

The diagnosis of an anomalous origin of the left coronary artery from the pulmonary artery is usually considered in an infant presenting with a clinical picture of congestive cardiomyopathy. The patients described here presented with mitral insufficiency, echocardiographic evidence of mitral valve prolapse, and good left ventricular function. It is noteworthy that the mitral insufficiency decreased in severity over the course of preoperative follow-up in these infants, who were not correctly diagnosed initially. The fourth child had the unusual association of tetralogy of Fallot and anomalous origin of the left coronary artery, which caused mitral regurgitation.

Mitral valve prolapse with mitral regurgitation is uncommon in infancy, unless associated with the Marfan syndrome. The present report should prompt clinicians to conduct a careful assessment of coronary artery anatomy in similar cases. The diagnosis of an anomalous origin of the left coronary artery may be difficult to reach by two-dimen-

sional echocardiography, and the artery may appear to be attached normally to the aorta [2]. Color Doppler may help to establish the diagnosis. On the other hand, an unusual malformation, such as atresia of the origin of the left main coronary artery, may remain undiagnosed by color Doppler. Thus, it would be advisable to proceed with an aortogram or right coronary angiography in suspect cases. Surgical repair of a left coronary artery of anomalous origin is indicated even in asymptomatic patients. It is interesting that direct re-implantation of the left coronary artery into the aorta was feasible in cases 1 and 3 in this series. This contrasts with the case of infants presenting with severe heart failure in whom the left coronary artery connects to the left side of the pulmonary artery and may be difficult to re-implant directly [3–5].

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