Cardiology in the Young

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Occlusion of the os of the left coronary artery by dysplastic aortic valve tissue presenting as progressive mitral insufficiency and cardiac arrest

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Brief Report

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Abstract

Mitral regurgitation in the neonatal period is relatively rare. It can be secondary to a congenital malformation of the valve apparatus or mitral valve dysfunction and deformation secondary to myocardial dysfunction or volume load of the left ventricle. Less commonly, it can be due to coronary artery abnormalities leading to mitral valve papillary muscle ischaemia and subsequent dysfunction. Such coronary artery abnormalities include anomalous left coronary artery from pulmonary artery, left main coronary artery atresia, or a thromboembolic phenomenon. In this study, we describe a newborn with a dysplastic aortic valve causing obstruction of the os of the left coronary artery leading to progressive mitral insufficiency.

Case presentation

A full-term newborn with an uncomplicated delivery was noted to have a heart murmur shortly after birth. An echocardiogram at this time revealed a dysplastic aortic valve with thickening of the right and left cusps with possible fusion of these cusps and attachments to the sinotubular ridge (Fig 1). There was mild aortic stenosis and insufficiency. The coronary artery ostial positions and proximal courses appeared normal. At 6 weeks of age, an echocardiogram demonstrated new mild mitral regurgitation with mildly echobright papillary muscles. An electrocardiogram (EKG) at this time was normal with no evidence of ischaemia. At her visit one month later, her parents also noted increased fussiness. However, she continued to feed and grow appropriately. Her physical examination was notable for a new apical S1-coincident systolic murmur, and her echocardiogram showed progressive mitral valve regurgitation, now classified as moderate (Fig 2). Her aortic stenosis and insufficiency remained mild, and her EKG remained normal. Given the echocardiogram findings and concern for possible coronary artery anomaly, she was referred for a CT angiogram of the chest for further evaluation. During induction of anaesthesia for her CT, she developed ventricular fibrillation and subsequent cardiac arrest. Cardiopulmonary resuscitation (CPR) was initiated, and she was cannulated onto veno-arterial extracorporeal membrane oxygenation (ECMO). An echocardiogram at this time demonstrated antegrade colour Doppler flow in the left main and left anterior descending coronary arteries.

Following stabilisation, the patient was brought to the cardiac catheterisation lab for coronary artery angiography. Multiple aortic root injections demonstrated mild to moderate supravalvar narrowing and mild to moderate aortic valve insufficiency, which was felt to be enhanced by catheter location. There was also thick obstructive tissue that was contiguous with the aortic valve leaflet and resulted in obstruction of the os of the left coronary artery (Fig 3). The left coronary artery had a normal origin and course, but it was noted to have diminished and sluggish filling with contrast. The right coronary artery appeared normal.

The patient subsequently went to the operating room that evening for surgical repair. Inspection of the aortic valve revealed obstruction of the left coronary os by a flap of tissue from the left aortic cusp. The left coronary cusp was detached and used to form a bicuspid leaflet with the right coronary cusp in order to clear aortic valve tissue from the left coronary artery os. The ascending aorta was also enlarged with decellularised homograft patch. She was decannulated from ECMO at the end of the procedure. She was ultimately discharged from the hospital at 4 months of age. At the time of discharge, she was taking all feeds by mouth and was on room air. She is now 10 months old and has continued to do very well clinically at her outpatient cardiology visits.

Of note, genetic testing revealed three variants of uncertain clinical significance on her CHDs panel, none of which were felt to be causative of her clinical features. These variants of uncertain

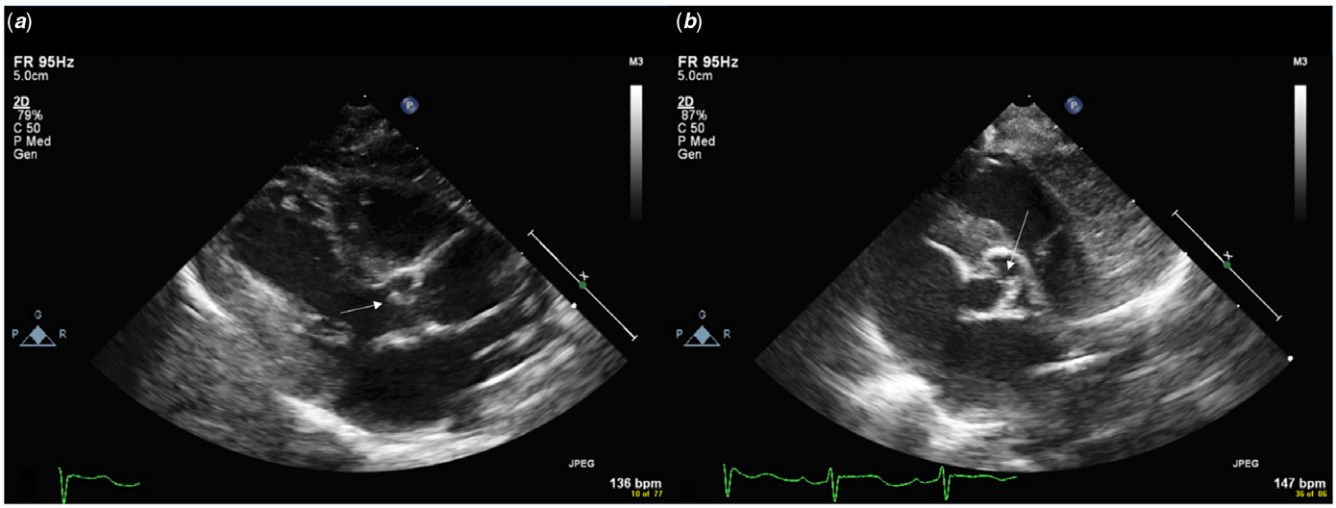

Figure 1. Parasternal long-axis view (***a***) and parasternal short axis view (***b***) demonstrating a dysplastic and thickened aortic valve (arrow) with attachments to the sinotubular ridge.

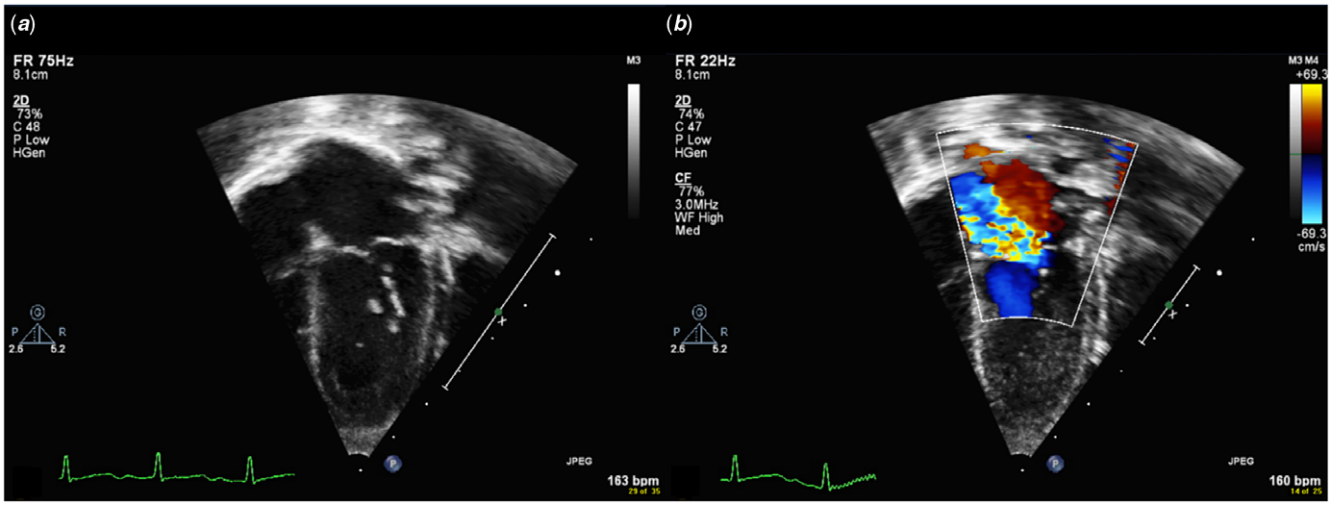

Figure 2. Apical four-chamber view demonstrating echobright papillary muscles (***a***) and moderate mitral valve regurgitation (***b***).

clinical significance were located in the ALMS1, DNAH5, and JAG1 genes. A single-nucleotide polymorphism microarray and karyotype were unremarkable, making Williams syndrome very unlikely.

Discussion

Mitral regurgitation in the neonatal period is relatively rare and can be a result of a congenital malformation or deformation secondary to volume load of the ventricle.[1,2] Less commonly, it can be due to coronary artery abnormalities.[3,4] The mitral valve has two papillary muscles that connect to the leaflets via the chordae tendineae. The anterolateral papillary muscle typically receives blood supply from the left circumflex and left anterior descending artery. The posteromedial papillary muscle is supplied by the right coronary artery in a right dominant system (most common) and by the left circumflex artery in a left-dominant system.[5] The papillary muscles play an important role in valve function and are the last portion of the heart to be perfused, resulting in a higher risk for ischaemia.[5] Ischemia of these papillary muscles results in mitral valve dysfunction and subsequent regurgitation. Although this is a well-established phenomenon, our case describes a particularly unusual mechanism for the development of mitral valve dysfunction: ischaemia occurring as a result of intermittent obstruction of the left coronary os by a dysplastic left aortic valve cusp. More commonly described mechanisms for this echocardiographic finding, albeit still very rare, include anomalous left coronary artery from pulmonary artery and left main coronary artery atresia.

This rare finding of left coronary artery occlusion by an aortic valve cusp has been described in the literature, primarily in older children and adults. To the best of our knowledge, there has been only one previously reported case of this finding in the neonatal period. However, ours appears to be the only case that presented primarily with progressive mitral valve insufficiency. Samuel et al. described a case of a 15 days old with a known dysplastic aortic valve with moderate aortic regurgitation who presented in cardiogenic shock with ischaemic changes on EKG. Both a cardiac MRI and cardiac catheterisation were performed to delineate coronary

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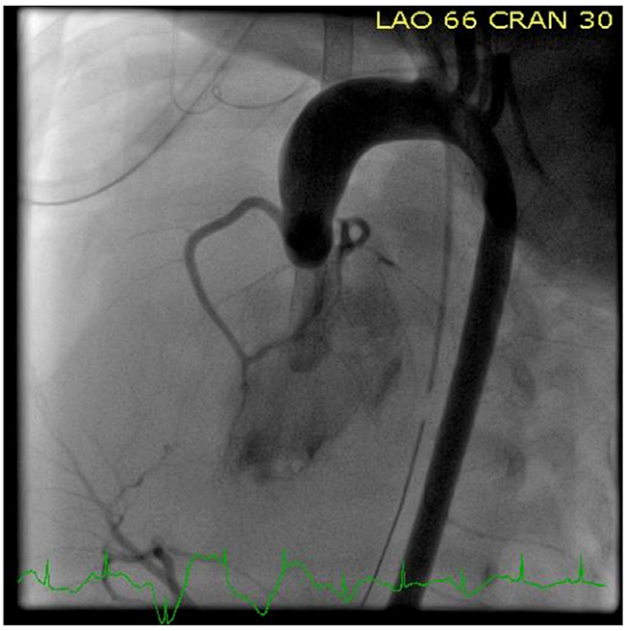

Figure 3. Angiogram demonstrating obstruction of the left coronary artery os.

artery anatomy, neither of which provided a clear aetiology for this presentation. In the operating room, direct surgical inspection revealed obstruction of the left coronary os by the left coronary leaflet.[6] The other cases found in the literature describe older children and adults, who presented with syncope, shortness of breath, angina, or sudden death. On echocardiogram, they commonly had aortic regurgitation or supravalvular aortic stenosis.[7–9]

The aortic valve is typically well-visualised by transthoracic echocardiography, allowing for delineation of the morphology of the commissures and fusion of leaflets. In addition, transthoracic echocardiography with colour Doppler is an important tool for evaluation of the coronary arteries and can reliably image the proximal coronary arteries.[10] In our case however, echocardiography did not serve as a reliable tool for diagnosis. On this patient's echo, the left coronary artery had a normal origin with antegrade flow and no stenosis of the vessel itself. Thus, although echocardiography is useful for diagnosis of coronary artery abnormalities, this unique abnormality of the aortic valve cusp with subsequent left coronary artery occlusion was not able to be visualised. This case demonstrates the utility of cardiac catheterisation in the functional assessment of both the aortic valve and the coronary arteries.

Conclusion

There are no known cases of progressive mitral valve regurgitation resulting from left coronary artery occlusion by dysplastic aortic valve tissue. It is important to consider this diagnosis in a patient with progressive mitral regurgitation, ventricular dysfunction, or cardiac arrest of unknown aetiology. As evidenced by our case, it is difficult to elucidate this diagnosis on transthoracic echo and CT angiography; cardiac catheterisation can therefore assist in recognising this abnormality. Any findings suggestive of myocardial ischaemia should be thoroughly investigated to evaluate for correctable coronary anomalies, and induction of anaesthesia must be slow and deliberate in these patients given their risk for life-threatening complications.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S104795112200292X

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Conflicts of interest. None.

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