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Original Article

Near-fatal neonatal coronary ischaemia associated with intermittent aortic regurgitation: successful surgical treatment

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Abstract An infant presented with features suggestive of an anomalous left coronary artery was found to have normal origins of both coronary arteries. Echocardiography during episodes of ischaemia showed marked aortic regurgitation with retrograde coronary flow. The left coronary leaflet was mildly hypoplastic. Surgical re-suspension of this leaflet prevented aortic regurgitation and the patient had no further symptoms and recovered cardiac function.

Keywords: Anomalous left coronary artery; ischaemic myopathy; life threatening events

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Background

Coronary ischaemia due to aortic regurgitation is a rare event usually associated with severe valvar regurgitation. We recently encountered a patient who presented at 2 weeks of age with symptoms mimicking an anomalous left coronary artery arising from the pulmonary artery and who also displayed concomitant echocardiographic evidence of aortic regurgitation. Surgical plication of the small left coronary aortic leaflet to the adjacent cusp relieved all the symptoms and signs.

A similar incident was recently published describing a 5-week-old infant with near-fatal episodes who was successfully treated by surgical placation of the left aortic valve cusps to each of the other two cusps with small pledgets and has been predicted with long-term asymptomatic survival.

Case report

The 2-week-old female infant we examined first presented with heart failure to an outside institution. Pregnancy and delivery of the baby had been unremarkable. At 2 weeks of age, she presented with tachypnoea and poor feeding. She was noted to have tachycardia with increased difficulty in breathing, and was transferred to the local emergency department where, on chest X-ray, she was found to have cardiomegaly, pulmonary vascular congestion, and an abnormal electrocardiogram. Her echocardiogram showed poor left ventricular function and suspicion of an anomalous left coronary artery from the pulmonary artery (Fig 1). She was transferred to our institution for further evaluation and potential surgery.

Initial echocardiographic imaging at our institution excluded anomalous left coronary artery from the pulmonary artery, as both the coronary arteries appeared to arise off the aortic root. We found moderate-to-severe aortic valve insufficiency without stenosis. The insufficiency was confined to the left coronary leaflet (Fig 2). The left coronary artery appeared to arise from the sinus of Valsalva, which was confirmed by three-dimensional echocardiography (Fig 3). This appropriate origin of the left coronary artery was also confirmed by cardiac catheterisation and angiography. The left ventricle showed aortic regurgitation with poor function, an ejection fraction of 30%, and no evidence of mitral valve insufficiency. Using several planes of examination, we noted retrograde flow in the left main coronary artery (Fig 4).

We attempted without success to treat the patient with anti-congestive medication and vasodilators;

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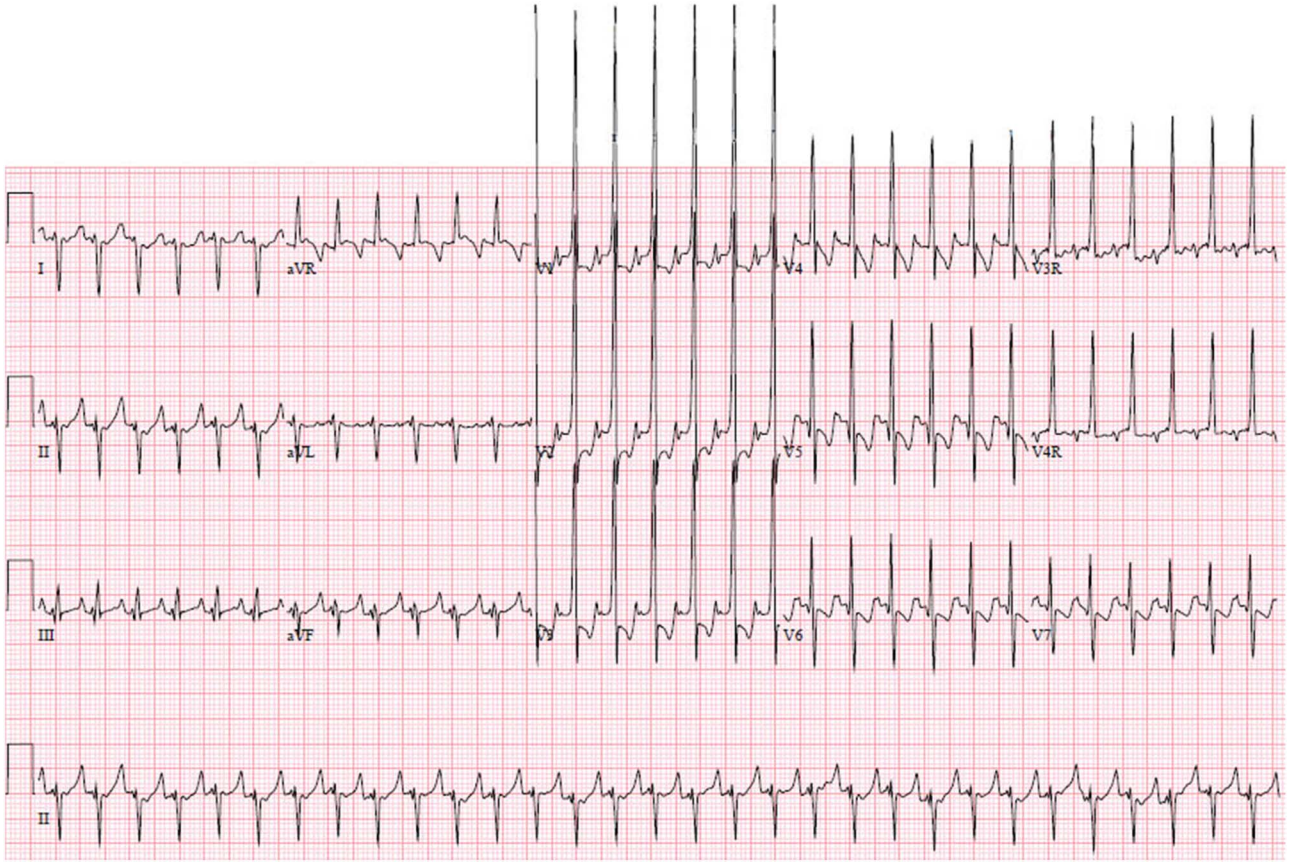

Figure 1.
Electrocardiogram of the patient shortly after admission showing tachycardia with a heart rate of 200. Right axis deviation, right ventricular hypertrophy, and marked left ventricular hypertrophy and strain.

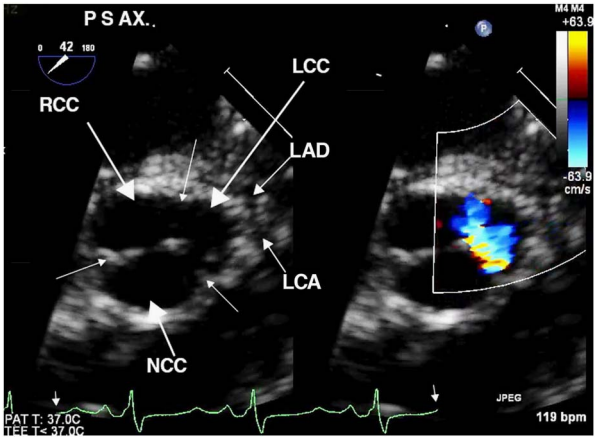

Figure 2.
This transoesophageal colour-compared Doppler image, taken in a view equivalent to the parasternal short-axis view (PSAX), shows the leaflet lines of coaptation (small arrows) and a small left coronary cusp (LCC). The frames are taken in diastole (arrow on electrocardiogram) with the aortic regurgitation confined exclusively to the LCC on the Doppler colour flow image. LAD = left anterior descending coronary artery; LCA = left (main) coronary artery; NCC = non-coronary cusp; RCC = right coronary cusp.

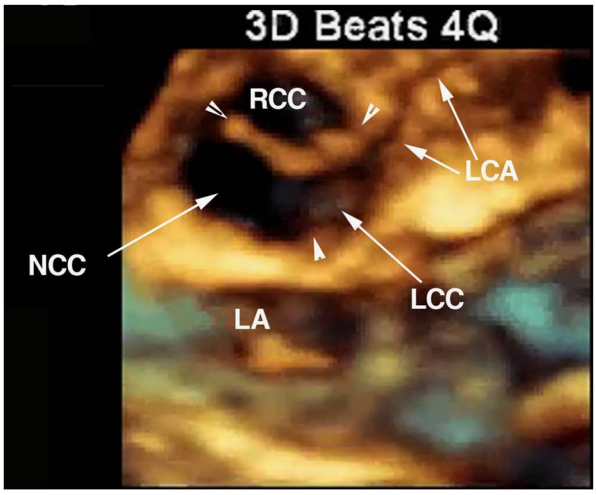

Figure 3.
This three-dimensional (3D) synthesis from a 3D zoom clip, viewed form a conventional echocardiographic view, confirms the small left coronary cusp (LCC) on the synthesised image with the left coronary artery (LCA) arising from it. The small LCC), the non-coronary cusp (NCC), and right coronary cusp (RCC) are labelled and arrowheads mark their commissures. LA = left atrial.

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however, the patient continued for some period to have episodes of irritability associated with electrocardiographic changes, suggestive of ischaemia.

We, therefore, elected to repair the lesion as described by Martin et al.[1]

Upon initiation of cardiopulmonary bypass, the patient was noted to have severe aortic insufficiency, which originated through the region of inadequate left coronary leaflet apposition. The left coronary leaflet appeared to be foreshortened and hypoplastic relative to the other leaflets. The abnormal leaflet occupied <20% of the aortic annular area. Inspection of the aortic valve demonstrated a trileaflet structure with a pliable leaflet; however, despite this, the leaflet exhibited appropriate mobility and suppleness. Pericardial pledgets were created. Leaflet commissuroplasties were performed using 6-0 Prolene sutures through the pericardial pledgets to attach the left coronary leaflet to the non-coronary leaflet and the left coronary leaflet to the right coronary leaflet (Fig 5). At the end of the procedure, there was minimal aortic regurgitation.

Since the procedure was completed, there have been no further episodes, whereas there has been a substantial improvement in ventricular function. The patient is now 2 years old. At our last examination, she appeared to be well with no further episodes of ischaemia, with an echocardiographic ejection fraction of 58%.

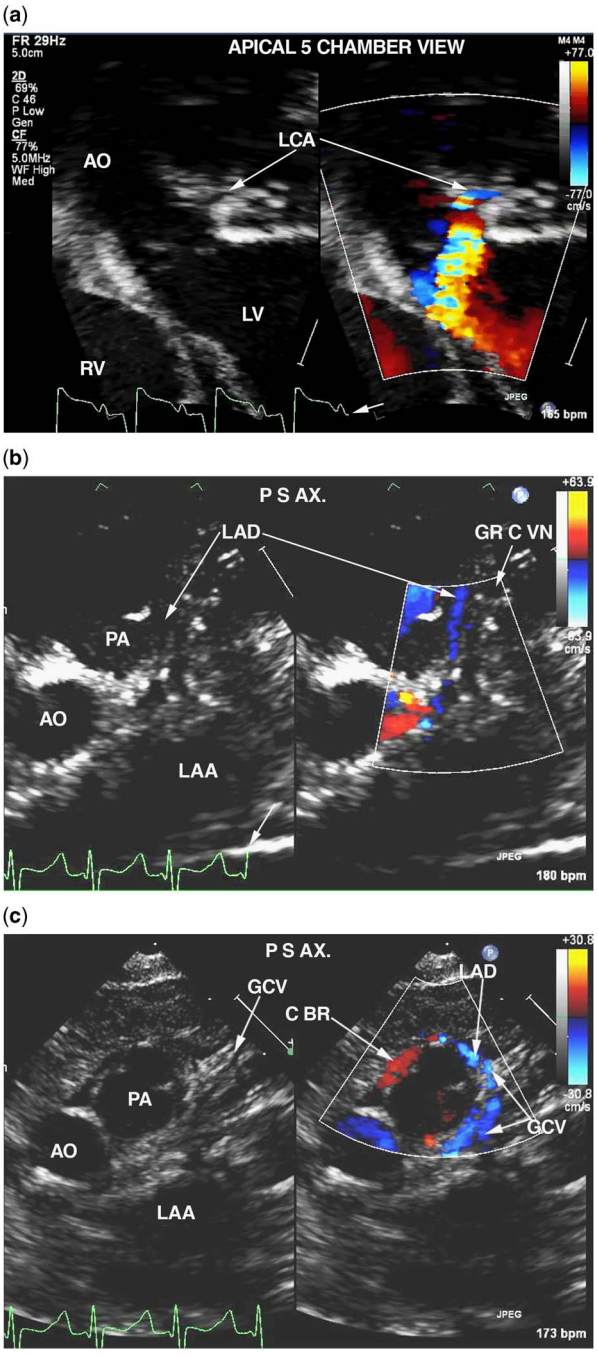

Figure 4.
*These frames show colour-compared Doppler with conventional imaging in simultaneous frames. (**a**) The conventional black and white image (left) shows a magnified apical five-chamber view of the aorta (AO) arising from the left ventricle (LV). The left coronary artery (LCA) is seen arising from the middle of the left sinus of Valsalva. In the right-hand frame, taken in diastole, as indicated by the arrow on the electrocardiogram, the diastolic flow is identified arising from within the right coronary artery entering the AO, then entering the left ventricular outflow tract, and striking the ventricular septum before running down towards the cardiac apex. (**b**) These colour-compared images are taken before the aortic valve opens in parasternal short-axis view (PSAX) and demonstrates retrograde flow into the LCA with flow extending out of the artery into the left sinus of Valsalva or the AO in the Doppler colour flow image. The great cardiac vein (Gr C Vn) runs alongside the left anterior descending (LAD) coronary artery. (**c**) In this PSAX view taken before aortic valve opening, the coronary flow can be identified in the Doppler colour flow image in the right. A conus branch (C BR) enters the LAD artery in a retrograde manner and in this frame can be differentiated from the great cardiac vein, although the flow in both vessels is directed away from the transducer. The coronary flow surrounds the anterior pulmonary artery (PA), which lies in front of the AO to its right and the left atrial appendage (LAA) behind it. RV = right ventricle; GCV= great cardiac vein.*

Discussion/comment

Previously, one of the authors had reported the occurrence of aortic regurgitation associated with a deficient left coronary cusp with ischaemic symptoms.[1]

The combination of a diminutive left coronary cusp and aortic insufficiency interferes substantially with the function of the sinus of Valsavla, as described by Bellhouse and Talbot[2] and Yoganathan et al.[3] Ischaemic symptoms may take the form of a near-fatal episode or, as in this case, may mimic the finding of an anomalous left coronary artery. In both instances, the diagnosis was established through a combination

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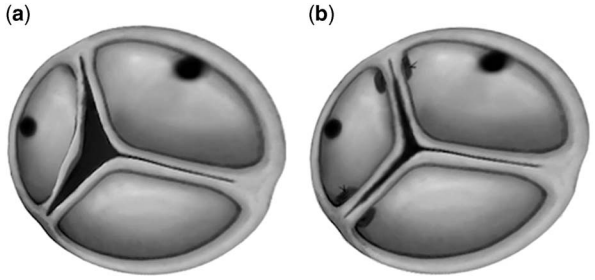

Figure 5.
*This diagram depicts the aortic valve from the surgeon's view from above. (**a**) The left coronary leaflet of the aortic valve is small, distorted, and does not coapt with its fellows. (**b**) The operative manoeuvre is shown, with plication of the left coronary cusp to the other two leaflets with two plegeted sutures that bring the deformed cusp into apposition with the other leaflets. See text for details.*

of echocardiography, angiography, and detection of elevated troponin levels.

In our previously reported case, the precipitation event for ischaemia occurred before the passing of a pigtail catheter across the aortic valve in the catheterisation laboratory. The small left coronary leaflet adhering to the aortic wall had created acute aortic insufficiency with ischaemic symptoms. Echocardiographic monitoring during the catheterisation procedure showed that the insufficiency was terminated by an aortic angiogram in the left coronary cusp, with documented disappearance of the aortic regurgitation and absence of ischaemia on electrocardiogram. In this child, other near-fatal episodes were terminated either spontaneously or by cardiac massage, and the patient always reverted to normal function after these episodes.

In our current patient, the episodes were similarly intermittent, but they appeared to be more chronic, with persistent loss of cardiac function. Even when the insufficiency was absent or minimal, the episodes of ischaemia persisted long enough to cause loss of global cardiac function. Owing to the insufficiency, the entire sinus of Valsalva flow in the left coronary system was compromised, due to a demonstrable runoff of blood in diastole from the left coronary artery, similar to that occurring in an anomalous left coronary artery from the pulmonary artery (Fig 4).

The reason as to why coronary ischaemia occurs so rarely in patients with aortic insufficiency might relate to a functional deficiency between the left aortic leaflet morphology and the left sinus of Valsalva.

Recently Ciotti et al measured the aortic valve leaflets and the area of the sinuses of Valsalva in normal infant and children hearts. They found that, on echocardiogram, the largest aortic leaflet with respect to the leaflet circumference was the non-coronary leaflet ($38 \pm 5\%$), followed by the right leaflet ($32 \pm 3\%$), and, finally, the left sinus and cusp were the smallest, contributing to approximately $28 \pm 3\%$ of the subtended area of the opposed aortic leaflets.[4] Thus, the leaflet most likely to have a particular sinus fail due to aortic insufficiency is likely to arise from the left sinus.

Much of the experimental work regarding the valvar leaflet and sinus of Valsalva function was carried out by Bellhouse and Talbot.[2] He noted that the valvar closure was dependent upon vortices that form in the sinuses of Valsalva. Yoganathan et al.[3] showed that the flow enters near the middle part of the crescent of the leaflet and exits at the sides of the leaflets adjacent to the aortic wall. During deceleration of the blood in late systole, the intra-sinus of Valsalva pressure exceeds the pressure in both the linear flow area and the open valve, as they begin to close. If this phenomenon does not occur, as in our patient with a hypoplastic left coronary valvar leaflet, then the vortex pressure within the sinus of Valsalva fails to occur, and the leaflet can collapse against the aortic wall, causing aortic regurgitation to follow.[5] In our current patient, we cannot invoke this mechanism entirely, as the valve leaflet was also deficient, and the whole mechanism and function within this leaflet and its sinus, as well as the coronary artery that lies within its wall, appeared to be defective, with compromise of forward flow in diastole into the left coronary system (see Fig 4).

There are similarities and differences in the presentation of disease in these two patients. Electrocardiograms showed similar ischaemia in both patients, with aortic regurgitation as the central link. In both the patients, the left coronary cusp was distorted and smaller than normal. Although without surgical treatment they both might have died in infancy, both of them achieved an excellent result after undergoing an identical operation.

The differences were in the patients' clinical presentations. In the first case, the patient presented with near-fatal episodes but returned to normal function and electrocardiogram after the episodes, whereas the second patient presented with features suggesting an anomalous left coronary artery from the pulmonary artery with persisting features of ischaemic myopathy.

An unanswered conundrum is why aortic regurgitation can lead to such severe coronary flow changes. It could have been due to intermittent blocking of the coronary artery by the aortic leaflet in the former case or due to total failure of pro-grade coronary flow in the left sinus of Valsalva in the latter. This symptom appears very rarely for morphological and physiological phenomena to be investigated further. Our report focusses on the range of findings related to coronary leaflet closure, and perhaps with an increased awareness of this phenomenon more events

will be uncovered and this entity may be better understood.

Ethical Standards

The author assert that all procedures contributing to this study comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008.

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