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Brief Report

Myocardial infarction in infancy caused by compression of an anomalous left coronary artery arising from the right coronary artery

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Abstract A 5-week-old child presented with a cardiac arrest secondary to myocardial ischaemia. Echocardiography demonstrated a single coronary artery arising from the right sinus of Valsalva. The coronary artery branched into left and right arteries, with the left artery then coursing anomalously in the tissue plane between the aortic root and the subpulmonary infundibulum. Compression of the left coronary artery caused severe myocardial ischaemia that resolved following construction of a bypass graft using the left internal thoracic artery. Stenosis at the anastomosis between the graft and the coronary artery was successfully treated by coronary angioplasty 2 years later.

Keywords: Internal thoracic artery bypass graft; coronary angioplasty; infant; coronary arterial surgery

A SINGLE CORONARY ARTERY IS RARELY FOUND AS an isolated abnormality. One large angiographic series documented an origin of a solitary artery from the right sinus of Valsalva, with aberrant origin of the main stem of the left coronary artery from the solitary coronary artery, in 0.015% of patients.[1] Such an isolated single coronary artery has been detected during childhood in only a few cases.[2] When the main stem of the left coronary artery arises from the right coronary artery it may follow a course to reach the anterior interventricular groove by running anterior to the right ventricular outflow tract, passing between the aorta and pulmonary trunk, coursing posterior to the ascending aorta or extending through the tissue plane between the aortic root and the subpulmonary infundibulum. A course between the aortic root and the subpulmonary infundibulum has been regarded as benign.[3] We describe a 5-week-old child with a single coronary artery, with the left coronary artery then arising from the right artery. The course of the left coronary artery between the aortic root and subpulmonary infundibulum caused severe myocardial ischaemia.

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Case report

A 5-week-old baby attending hospital for routine review of a head injury was found to have signs of low cardiac output and cardiac failure. He suffered a cardiac arrest during intravenous cannulation, and was successfully resuscitated. The electrocardiogram showed inversion of the T waves in leads I, II, aVL and V4–V6. Echocardiography demonstrated a moderately dilated and poorly contracting left ventricle, with a fractional shortening of 8%. Although cardiac structure was grossly normal, the left coronary artery was seen to arise from the right coronary artery, passing between the outflow tracts from the right and left ventricles to reach the left side of the heart (Fig. 1). Selective coronary angiography confirmed that a single coronary artery arose from the right sinus of Valsalva, giving rise to both left and right coronary arteries. The main stem of the left coronary artery followed a course behind the right ventricular outflow tract, in

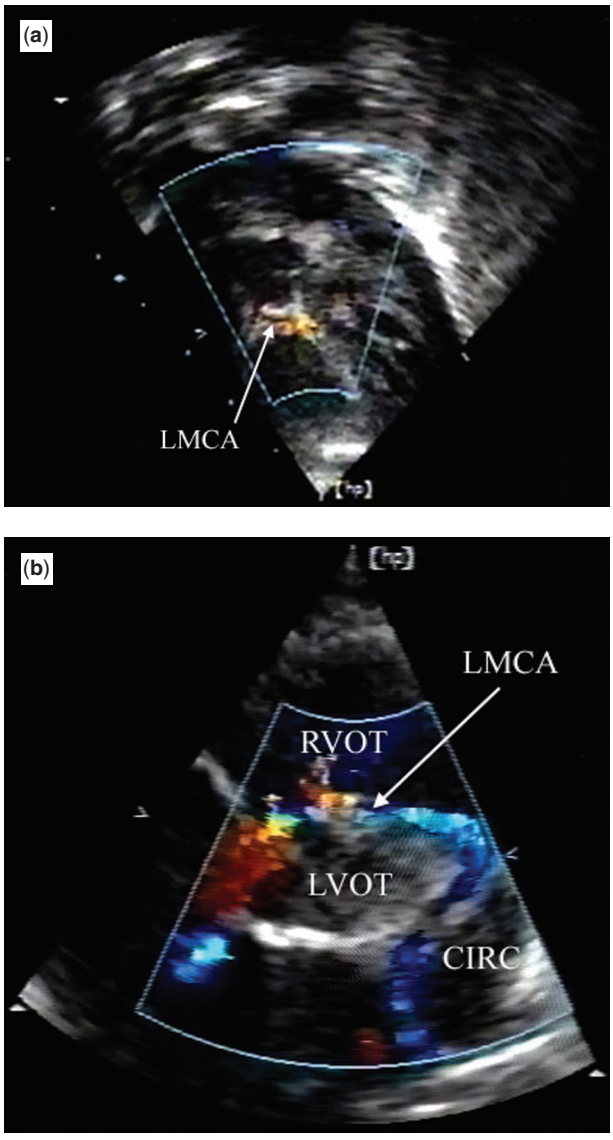

Figure 1.
*Transthoracic echocardiogram showing (**a**) foreshortened apical view of the single coronary artery, demonstrating the main stem of the left coronary artery arising from the right coronary artery and passing between the aortic root and the subpulmonary infundibulum towards the anterior interventricular groove. (**b**) Short axis view of the main stem passing between the left and right ventricular outflow tracts. Colour Doppler demonstrates flow acceleration where the artery tunnels between the aortic root and subpulmonary infundibulum, suggesting compression. The circumflex artery turns posteriorly towards its normal position in the atrioventricular groove. CIRC: circumflex coronary artery; LMCA: left main coronary artery; LVOT: left ventricular outflow tract; RVOT: right ventricular outflow tract.*

the tissue plane between the aortic root and subpulmonary infundibulum, before bifurcating to give rise to left anterior descending and circumflex branches. The segment of the main stem running between the outflow tracts was narrow throughout the cardiac cycle, with evidence of systolic compression (Fig. 2).

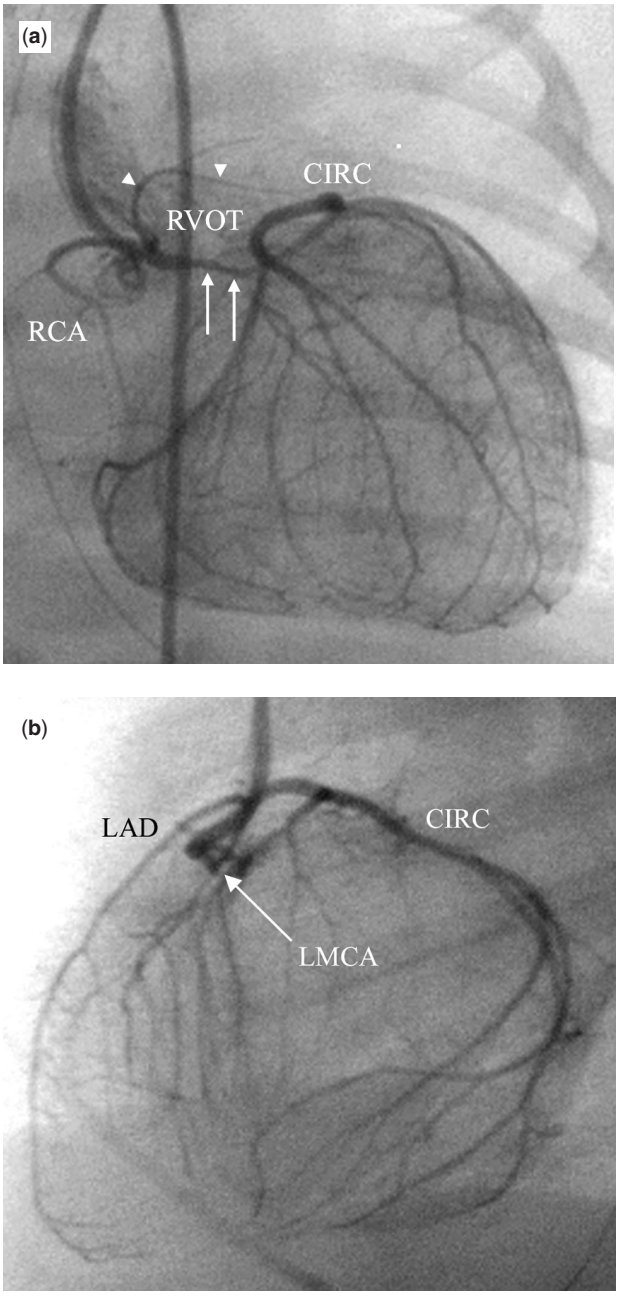

Figure 2.
*Coronary angiogram showing (**a**) right anterior oblique view of the right and left coronary arteries arising from a single orifice in the right sinus of Valsalva. The left main stem loops caudally, anteriorly and to the left, indicating a course between the aortic root and subpulmonary infundibulum (marked by the arrows). A conal branch of the right coronary artery (marked by arrowheads) defines the position of the anterior right ventricular outflow tract. The section of the left coronary artery that lies between the outflow tracts is narrower than the circumflex artery, which turns posteriorly to reach its normal position in the atrioventricular groove. (**b**) Left anterior oblique view showing the bifurcation of the left coronary artery and the small diameter of the left anterior descending coronary artery. CIRC: circumflex coronary artery; LAD: left anterior descending coronary artery; LMCA: left main coronary artery; RCA: right coronary artery; RVOT: right ventricular outflow tract.*

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The patient developed marked ST depression and inversion of the T waves during agitation or handling, reflecting the tenuous nature of the myocardial blood supply. He was deeply sedated, ventilation was maintained with a high concentration of inspired oxygen, myocardial function was supported with intravenous enoximone, catecholamines were avoided, and intransal glyceryltrinitrate was used to break through any ischaemic episodes. Two days after presentation, fractional shortening had improved to 20%. Surgery was performed 2 weeks after presentation, as there were initially concerns that an intraventricular haemorrhage (initially classified as grade II) would extend during cardiopulmonary bypass. The left internal thoracic artery was anastomosed to the left anterior descending coronary artery with interrupted 8/0 prolene sutures.

Further electrocardiogram changes following surgery indicated ongoing inferior and lateral ischaemia. He was initially treated with intravenous heparin and glyceryltrinitrate, and later with aspirin, transdermal glyceryltrinitrate, and oral propranolol. One week after surgery the electrocardiogram had normalised, with no signs of myocardial ischaemia when crying. He remained well after discharge, and was able to stop the propranolol and glyceryltrinitrate after 5 months. Left ventricular function remained mildly impaired, with a fractional shortening of 21%.

Eight months after surgery, his mother described an acute life-threatening event at home. Repeat cardiac catheterisation showed good flow into the right coronary artery, but little antegrade flow into the left coronary artery, albeit that there was excellent flow down the left internal thoracic arterial graft, which showed mild stenosis at its point of anastomosis with the left anterior descending coronary artery. By 15 months of age, there were again concerns about coronary ischaemia, as he repeatedly woke at night screaming, bringing his arms to his chest, apparently in pain. A cardiac myoview study with adenosine showed a fixed defect at the apex, in keeping with previous myocardial infarction, but did not show ischaemia after stress. Coronary angioplasty was performed at 2 years of age. A moderate stenosis at the distal end of the left internal mammary artery graft was successfully dilated with Stormer balloons having the dimensions of 2 by 10 millimetres and 2.5 by 15 millimetres (Medtronic AVE, Santa Rosa, CA, USA). There was no significant residual stenosis. After over a year free from symptoms, he developed chest pain again, and a reversible defect was found on perfusion scanning. Angiography at almost 4 years of age revealed moderate recurrent narrowing at the anastamotic site. Dilation was performed with a 3-millimetre balloon. While deflating the balloon, he developed ventricular fibrillation. During resuscitation, the guide catheter dissected the proximal part of the left internal thoracic artery. Successful defibrillation was followed by loss of cardiac output. Despite implantation of 3 stents in the artery, restoring a good lumen, myocardial function did not recover, and he died in the catheterisation laboratory.

Discussion

Although the majority of patients with an anomalous course of the main stem of the left coronary artery between the aortic root and subpulmonary infundibulum do not develop symptoms, myocardial ischaemia, ventricular tachycardia and even sudden death have been reported.[3–6] Our patient clearly had a significant reduction in coronary arterial flow, resulting in myocardial ischaemia, infarction and cardiovascular collapse, confirming that this aberrant course of the left coronary artery is not always benign.

Compression of the artery between the left and right ventricular outflow tracts was the most likely cause of the ischaemia. Such compression may cause arterial pressure to exceed aortic pressure in diastole.[7] Parallels can be drawn with myocardial bridging, where there is systolic compression and a persistent diastolic reduction in coronary arterial diameter. These changes are exacerbated at faster heart rates, and are associated with reduced distal coronary flow reserve.[8,9] The clamping effect leads to significant ischaemia at faster heart rates, when the diastolic time period is reduced and myocardial consumption of oxygen is increased.

The aberrant artery can be identified prospectively by transthoracic echocardiography. It is easier to distinguish a course between the aortic root and subpulmonary infundibulum from a course between the two arterial trunks using a foreshortened apical view, as the exact level of the sinutubular junctions is unclear in short axis. Colour Doppler flow mapping is of value in distinguishing linear artefacts from the true coronary arterial lumen, determining the direction of flow and identifying areas of increased flow velocity.[4]

To the best of our knowledge, symptoms caused by an anomalous origin of the main stem of the left coronary from either the right coronary artery, or directly from the right sinus of Valsalva, have not been reported before at such a young age. Older patients with this anomaly have been successfully treated using the left internal thoracic artery as a graft to the left anterior descending branch, in combination with a saphenous vein graft to the circumflex artery.[10] Our case suggests that a graft to the circumflex artery may be unnecessary, as the patient remained asymptomatic whilst the left internal thoracic arterial graft was patent. Ischaemia in the early postoperative period was probably caused by oedema

at the site of anastomosis. The long-term effect of competitive flow on graft patency in such a case is uncertain. As far as we know, ours is also the youngest patient to undergo angioplasty of the left internal thoracic artery, although ultimately the procedure proved fatal.

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