

Severe left ventricular dysfunction in an infant

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DESCRIPTION

A 5-month-old infant with poor weight gain was taken to the emergency department due to respiratory distress and feeding intolerance. On examination, she was tachypnoeic and was wheezing and was diagnosed with bronchiolitis. On heart auscultation, however, there was a grade II/VI holosystolic murmur on the left lower sternal border with irradiation to the axilla. Her liver was palpable 4 cm below the right costal margin and she had poor weight gain. Chest X-ray showed cardiomegaly (cardiothoracic index of 70%), which prompted referral to paediatric cardiology. ECG showed repolarisation anomalies in the left precordial leads, suggesting dilation of the left ventricle (LV); however, there were no signs of ischaemia. Echocardiogram showed LV dilation—diastolic diameter of 46 mm (Z-score +12), ejection fraction of 25% and moderate mitral regurgitation due to poor leaflet coaptation. The left coronary artery originated from the main pulmonary artery and there was retrograde flow in the left anterior descending artery (figure 1).

She was referred for cardiac surgery exclusively based on these investigations.^{1,2}

The surgical procedure was carried out through median sternotomy, under circulatory support, and myocardial protection was provided with hypothermia and administration of a cardioplegic solution and aortic cross-clamping. On inspection, the right coronary artery was markedly dilated (figure 2). The pulmonary trunk was transected above the level of the left coronary artery. The left coronary artery button was dissected and reimplemented in the aorta, creating a double flap. The pulmonary artery was anastomosed end-to-end with direct suture.^{3,4} Blood pressure was controlled

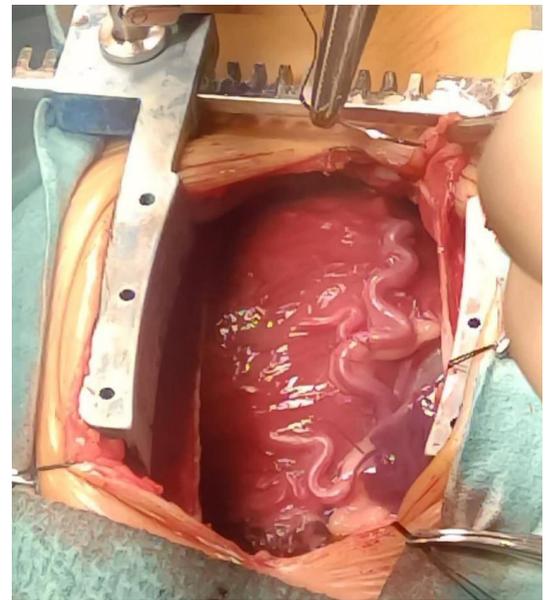


Figure 2 Intraoperative image: right coronary artery enlargement due to the development of extensive collateral coronary circulation.

intraoperatively and was normal. The chest was closed and the infant was transferred to the intensive care unit on mechanical ventilation and inotropic support. Postoperative echocardiogram showed LV dilation with severe dysfunction, but with improved segmental contractility, anterograde flow in the anterior descending artery, mild mitral regurgitation and right ventricle with good function. Despite being haemodynamically stable over the following hours, she went into cardiorespiratory arrest during aspiration of respiratory secretions and could not be resuscitated.

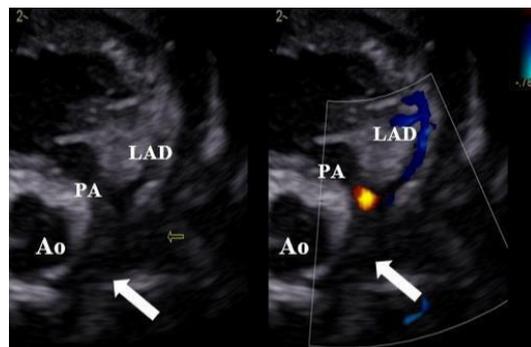


Figure 1 Transthoracic echocardiogram: parasternal short axis showing the left coronary artery originating from the main pulmonary artery (arrows); colour Doppler showing retrograde flow in the left anterior descending artery entering the PA in diastole. Ao, aorta; LAD, left anterior descending artery; PA, pulmonary artery.

Patient's perspective

We realised that our child's respiratory symptoms were related to the cardiac dysfunction. We understand the importance of early referral for Paediatric Cardiology of any child with poor weight gain and cardiomegaly, with or without murmur. We are grateful to the medical team for having made an accurate diagnosis before serious damages ensued.

Contributors AT has received the child and collected all data supporting the diagnosis and suggested treatment. MA performed the surgery. OM and NN wrote the case and gathered literature.

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Competing interests None declared.



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Learning points

- ▶ Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a very rare congenital anomaly that accounts for 0.25%–0.5% of all heart diseases, with a mortality rate of 90% during childhood when untreated, and the diagnosis of this entity requires a high index of suspicion.
- ▶ Failure to thrive in an infant should prompt thorough aetiological investigation as it may be a sign of a severe underlying condition.
- ▶ It is mandatory to exclude ALCAPA as a cause of dilation and left ventricle dysfunction in infants since at this stage it is amenable to surgical correction with good prognosis.

Patient consent for publication Parental/guardian consent obtained.

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