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## Case Report

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# Under pressure: paediatric hypertensive dilated cardiomyopathy secondary to renal artery stenosis

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**Abstract**

Severe dilated cardiomyopathy in children may uncommonly be caused by abnormal loading conditions such as mid-aortic pathology and renal artery stenosis. Refractory hypertension and left ventricular dilatation with hypertrophy are important clues to reversible causes. We present a case of dilated cardiomyopathy in a child secondary to mid-aortic syndrome with renal artery stenosis.

## Case report

A 4-year-old female presented to a local emergency department with breathlessness, lethargy, and vomiting. She had a long-standing history of poor oral intake, worsening significantly in the two months before presentation. She was neurodevelopmentally normal, fully immunised, with no allergies.

Her weight at 11.2 kg was below the 1$^{st}$ centile for age, with a height on the 25$^{th}$ centile. She was tachypnoeic and hypertensive, with systolic blood pressures of 130–140 mmHg (> 99$^{th}$ centile for age and height). Her peripheries were cool, and she had lower body oedema palpable up to the knee, periorbital oedema, and hepatomegaly 5 cm below the costal margin. Her jugular venous pulsation was markedly elevated. There was no radial-femoral delay nor pulse differential. She had a 3/6 pansystolic murmur loudest at the displaced apex and radiating into the axilla. Her facial features were not suggestive of a syndromic cardiomyopathy.

Electrocardiogram (Figure 1a) demonstrated sinus tachycardia, voltage criteria for left ventricular hypertrophy, prominent mid-septal ST-segment elevation, and abnormal repolarisation with inferolateral T wave flattening. Chest radiograph demonstrated cardiomegaly with diffuse interstitial oedema. Lactate was 2.9 mmol/L, N-terminal pro-brain natriuretic peptide was 61,880 ng/L, and troponin T was 46 ng/L. She was urgently transferred to our referral centre for management of acute decompensated heart failure.

She was established on non-invasive ventilation and milrinone. Echocardiogram (Figure 1b,c) demonstrated severe dilatation of the left ventricle with an end diastolic diameter of 54 mm (Z + 9.5), severe impairment in left ventricular systolic function (ejection fraction: 20%), and evidence of elevated filling pressures with lateral E/E' of 15.8. There was severe mitral regurgitation related to annular dilatation. The right ventricle was moderately dilated with moderate systolic dysfunction. The interventricular septum and left ventricular posterior wall thickness were 8.4 mm (Z + 5.4) and 9.3 mm (Z + 6.7), respectively. The coronary artery origins were normal, and there was no left heart obstruction nor coarctation of the aorta. The proximal abdominal aortic Doppler profile was normal.

Her hypertension, initially attributed to her acute illness, was refractory to maximal doses of milrinone and the addition of sodium nitroprusside, prazosin, and clonidine. Renal Doppler ultrasound to investigate for secondary causes of hypertension (Figure 2a) could not demonstrate a main left renal artery arising from the aorta. Further investigation with CT abdominal angiography (Figure 2b, Supplementary Figure S1, Supplementary Video S1) demonstrated abdominal aortic stenosis (from 10 mm supra-renally to 5.5 mm at the level of the expected origin of the main left renal artery) and absent opacification of the ostial and proximal left renal artery.

A diagnosis of mid-aortic syndrome and renal artery stenosis with secondary severe hypertension and dilated cardiomyopathy was made. She proceeded to cardiac catheterisation and balloon angioplasty of the left renal artery (Figure 2c, d). There was dramatic improvement in hypertension, allowing weaning of her antihypertensive regimen, non-invasive ventilation and milrinone. She was established on oral medical therapy for heart failure (lisinopril, carvedilol, and spironolactone) and discharged.

Chromosomal microarray, connective tissue disease screening, and cardiomyopathy genetic panels were non-contributory. There was no evidence of systemic inflammation nor historical fevers suggestive of Takayasu arteritis.

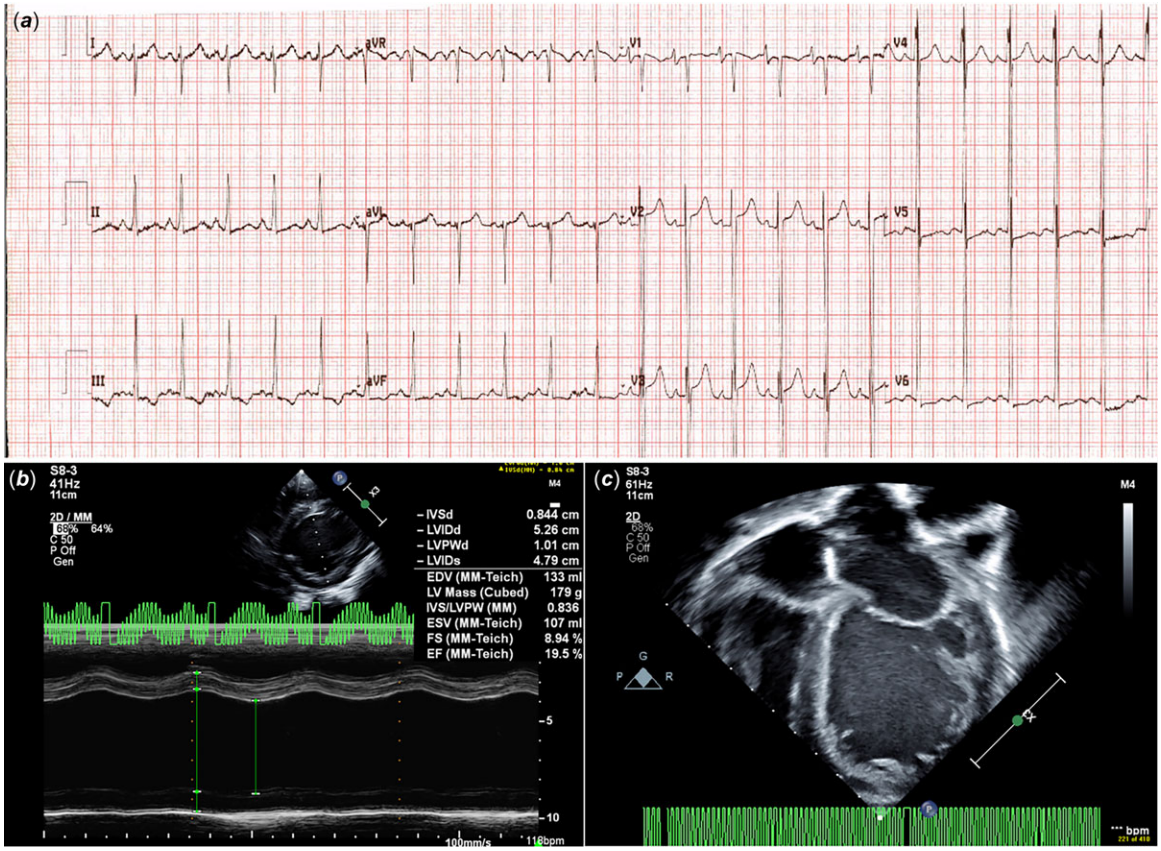


**Figure 1.** **(*a*)** ECG demonstrating sinus tachycardia with voltage criteria for left ventricular hypertrophy, mid-septal ST-segment elevation, and abnormal repolarisation with inferolateral T-wave flattening. **(*b*)** M-mode PSAX demonstrating markedly reduced left ventricular fractional shortening (9%) and IVS thickening (Z-score + 5.4). **(*c*)** Apical four-chamber view demonstrating severe dilatation of the left ventricle and left atrium with circumferential LVH.

At 3-month follow-up, renal artery Doppler demonstrated maintenance of pulsatile flow with normal left renal artery waveform and no recurrence of stenosis. At 6 months of follow-up, the patient remained clinically well, normotensive, and asymptomatic, tolerating oral heart failure therapy. Echocardiography continued to demonstrate severe dilatation of the left ventricle but improvement in left ventricular systolic function (ejection fraction: 47%).

## Discussion

The assessment of newly diagnosed dilated cardiomyopathy in childhood warrants investigation for reversible secondary causes, although its aetiology is idiopathic, familial, or genetic in the majority of cases.[1] An uncommon cause for dilated cardiomyopathy in children is sustained uncontrolled hypertension.[2] Renal artery stenosis and mid-aortic syndrome are rare treatable causes of severe and treatment-refractory hypertension in children.[3]

This case emphasises the importance of meticulous screening for secondary causes of cardiomyopathy. Early clues—such as unusually severe refractory hypertension and left ventricular hypertrophy (as opposed to the more commonly encountered "thinned out," hypokinetic left ventricle in chronic dilated cardiomyopathy)[1] on echocardiogram—suggested an atypical cardiomyopathy. A "normal" abdominal aortic Doppler profile initially excluded coarctation, but distal mid-aortic obstruction is inadequately screened for with standard subcostal echocardiographic imaging.[2]

In addition, the longstanding cardiomyopathy suggests a missed opportunity to identify hypertension and screen for secondary causes prior to the development of cardiomyopathy. Despite increasing community prevalence of primary hypertension in children, secondary causes remain more likely than in adults, especially in non-overweight children under 6 years of age.[4] Renal and renovascular causes are the most common secondary causes of hypertension.[5] Screening for hypertension is recommended but underperformed in children and is especially important in children with poor growth.[4] Lifetime exposure to hypertension from childhood increases risk of later-onset cardiovascular disease, even before the onset of end-organ complications, as in the present case.[6]

Childhood renal artery stenosis and mid-aortic syndrome frequently present with severe hypertension, and in 44% of cases are refractory to medical treatment.[2] However, impairment of systolic function and dilated cardiomyopathy results infrequently from mid-aortic syndrome when compared to adult case-series.[2,7] Endovascular intervention in renal artery stenosis and mid-aortic syndrome in adults has shown evidence of reversal of hypertension, left ventricular hypertrophy, and heart failure symptoms.[8] Concerns persist regarding the longevity of therapeutic response to balloon angioplasty and the potential need for reintervention.[2] However, this case avoided the risk of technical failure (reported in 28% of cases) and had a reassuring therapeutic response in the elimination of hypertension following percutaneous balloon angioplasty, both positive prognostic indicators consistent with potential for reverse remodelling.[2,9] At a median follow-up of 4

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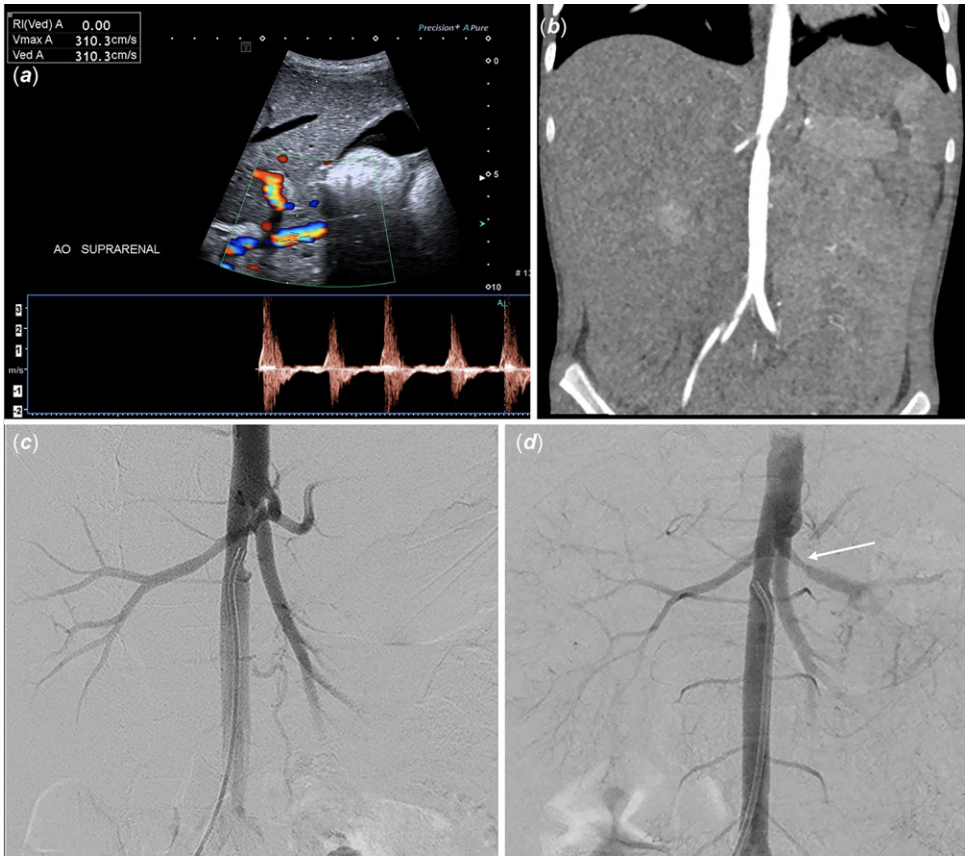


**Figure 2.** (***a***) Doppler ultrasound of kidneys could not demonstrate a main left renal artery, with evidence of increased supra-renal aortic velocity. (***b***) CT angiogram demonstrating mid-aortic narrowing with absent opacification of the ostial and proximal left main renal artery. (***c***) Pre-balloon angioplasty angiography demonstrating mid-aortic narrowing distal to the origin of the superior mesenteric artery to the level of the expected origin of the main left renal artery. No evidence of a main left renal artery is present, instead showing collateral supply to the left kidney. (***d***) Post-balloon angioplasty angiogram demonstrating newly patent left renal artery with residual stenosis (arrow) and increased calibre of the mid-aorta.

years, more than two-thirds of those who underwent isolated balloon angioplasty achieved adequate blood pressure control, although the majority require ongoing antihypertensive therapy.[7] Lifelong cardiology, renal, and local medical provider review will be important to identify recurrence of hypertension.

## Conclusion

This report highlights the critical importance of investigating secondary causes of dilated cardiomyopathy and hypertension in children to identify treatable causes of heart failure.

**Supplementary material.** The supplementary material for this article can be found at https://doi.org/10.1017/S1047951125109748.

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**Competing interests.** None.

**Ethical standard.** The authors assert that all procedures contributing to this work comply with the ethical standards of Australia's national guidelines on human experimentation—the National Statement on Ethical Conduct in Human Research—and with the revised Helsinki Declaration in 2008, and have been approved by the Sydney Children's Hospitals Ethics Committee.

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