

Images in Congenital Cardiac Disease

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# Severe coronary artery ectasia in a paediatric patient with Noonan syndrome presenting for transcatheter pulmonary valve placement

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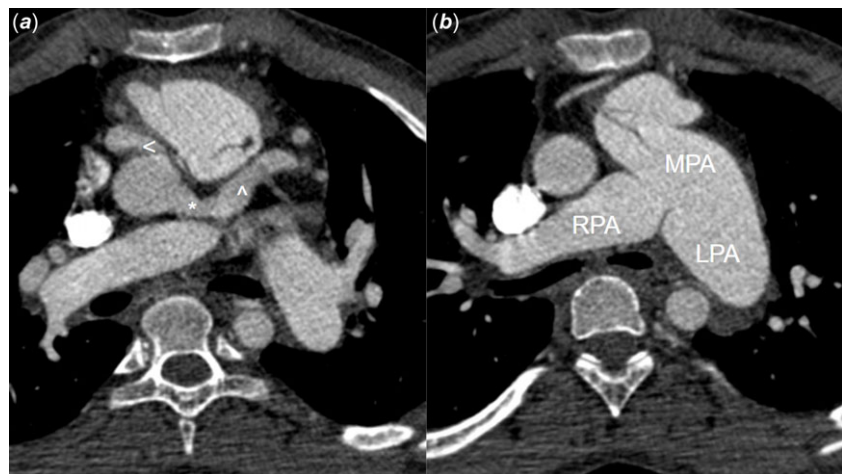
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**Abstract**

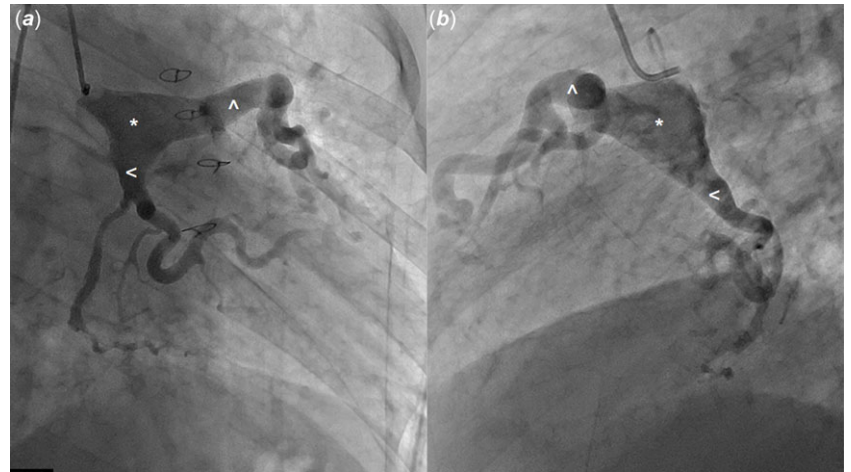
Coronary ectasia is a very rare phenomenon seen in Noonan syndrome with only a few documented case reports. We describe a 14-year-old with Noonan syndrome and tetralogy of Fallot with described coronary artery ectasia since infancy who presented for possible transcatheter pulmonary valve placement and was found to have severe ectasia of bilateral coronary arteries.

**Introduction**

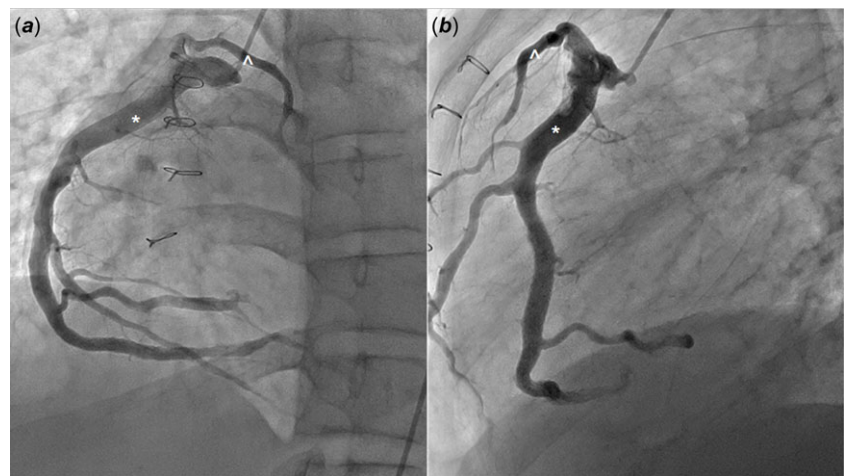
Coronary ectasia is a rare finding in Noonan syndrome with only a few documented case reports.<sup>1–5</sup> A 14-year-old male with Noonan syndrome presented to our office for continued follow up of his CHD consisting of pulmonary valve stenosis that was surgically augmented at 2 months of age using a transannular patch in a foreign country, hypertrophic cardiomyopathy, and severely dilated bilateral coronary arteries. His surgical report from infancy noted prominent bilateral coronaries at the time of surgery. He had significant right ventricular dilation due to his free pulmonary insufficiency and, based on CT scan, was a possible self-expanding transcatheter pulmonary valve candidate, so he presented for cardiac catheterisation (Fig 1). Selective coronary angiography depicted a severely dilated left main coronary artery with ectasia of the left anterior descending and left circumflex coronary arteries (Fig 2). The right coronary artery was ectatic throughout its entire length (Fig 3). No discrete aneurysms were seen. The right and left pulmonary arteries were found to be severely dilated along with the main pulmonary artery (Fig 4). A 40 mm sizing balloon was placed across the right ventricular outflow tract with continued flow around the balloon so the patient was deemed too high risk for possible transcatheter pulmonary valve implantation with currently available devices. The patient was taken to surgery for a pulmonary valve replacement.



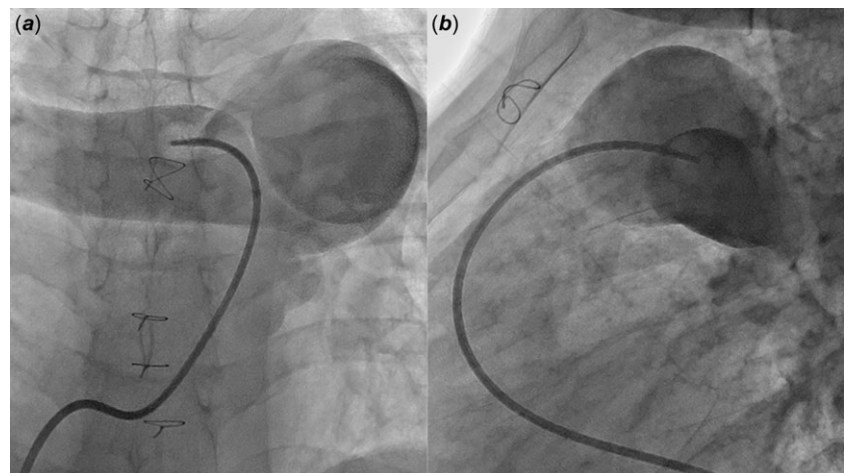
**Figure 1.** (a) Axial plane of CT angiogram depicting a severely dilated left main coronary artery (\*), left anterior descending artery (^), and right coronary artery (<). (b) Axial plane of CT angiogram depicting a severely dilated main pulmonary artery as well as a right and left pulmonary artery.



**Figure 2.** (a) Right anterior oblique (RAO)/Cranial angulation and (b) lateral angulation with selective left main coronary angiography. There is a severely dilated left main coronary artery (measuring 7 mm at the ostium and 20 mm just prior to the bifurcation)(\*) with significant ectasia of the left anterior descending artery (^)(measuring 9.9 mm) and left circumflex artery (<)(measuring 7 mm).



**Figure 3.** (a) Anterior posterior angulation and (b) lateral angulation with selective right coronary angiography. There is a severely dilated right coronary artery (\*) (6.4 mm proximal) with significant ectasia of a conal branch (^).



**Figure 4.** (a) RAO/cranial angulation and (b) lateral angulation with angiography in the main and branch pulmonary arteries. There is severe dilation of the right, left, and main pulmonary arteries.

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**Competing interests.** None.

**Compliance with ethical standard.** All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent was obtained from all individual participants included in this case report.

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