

Isolated Coronary Artery Aneurysm in a 12-Year-Old Boy With Marfan Syndrome

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An asymptomatic 12-year-old boy with Marfan syndrome presented for routine follow-up and to establish cardiology care at our institution. His previous echocardiograms were reportedly normal. An electrocardiogram showed normal results. A routine echocardiogram revealed an ectatic proximal right coronary artery. Subsequent coronary computed tomographic angiograms showed a fusiform aneurysm beginning at the right coronary artery ostium (Fig 1). The aneurysm was 8 mm long and 5.2 cm in diameter (Z score, +3.8). The patient was prescribed low-dose aspirin.

Comment

Marfan syndrome is an autosomal dominant genetic disease caused by a mutation in the fibrillin *FBNI* gene (estimated prevalence, 1 in 3,000–10,000 people).¹ The gene is integral to elastic connective tissue, so patients with Marfan syndrome often have related abnormalities, especially cardiovascular, skeletal, and ocular.² Chief among the cardiovascular complications is aortic root dilation, which carries the risk of aortic dissection and may suggest a need for prophylactic aortic root replacement. In Marfan syndrome, coronary artery involvement is usually a consequence of aortic dissection,

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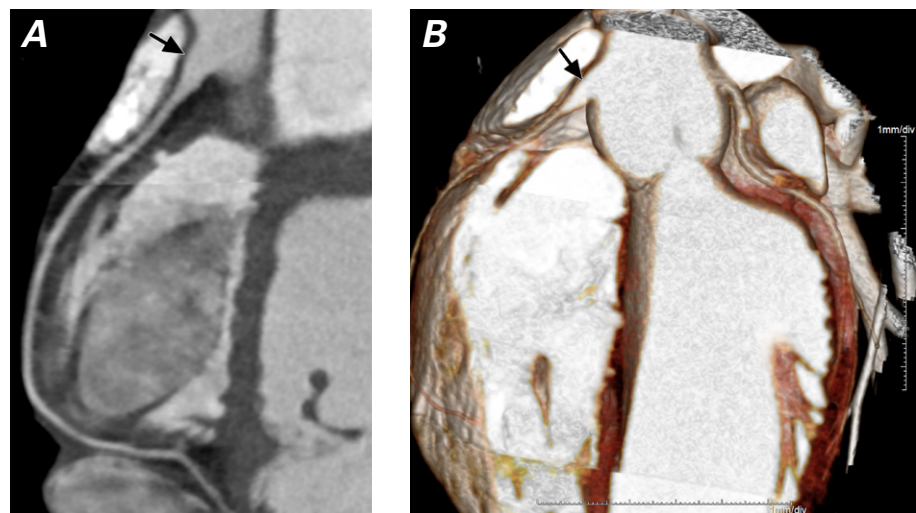


Fig. 1 Computed tomographic angiograms. **A)** Curved multiplanar reformatted image shows a mildly dilated fusiform aneurysm (arrow) beginning at the right coronary artery ostium.

B) Three-dimensional volume-rendered image shows the 8-mm-long, 5.2-mm-diameter aneurysm (arrow).

and isolated coronary artery aneurysms are rare³; to our knowledge, ours is only the second reported case in a pediatric patient.⁴

Routine coronary monitoring with the use of echocardiography and other imaging methods is important in patients with Marfan syndrome, including children, because patients who have coronary aneurysms are often asymptomatic.⁵ Detecting an aneurysm will enable early treatment and possibly improve the patient's overall prognosis.

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