Diagnosing Left Coronary Ostial Atresia: Sometimes the Old Ways Are Best

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Abstract

Keywords

- coronary artery
- ostial atresia
- cardiac catheterization
- cardiac computed tomography

A 12-year-old boy with intermittent syncope associated with exercise. Exercise stress testing suggested myocardial ischemia and 2D echocardiography failed to confirm antegrade flow in the left main stem. Advanced imaging techniques including cardiac-gated computed tomography angiography and stress cardiac magnetic resonance imaging were falsely reassuring. Cardiac catheterization demonstrated left coronary artery ostial atresia with a good caliber left coronary system supplied by generous collaterals from the right coronary artery. The patient underwent successful coronary artery bypass grafting.

Noninvasive coronary artery imaging is an expanding subspecialty and has been suggested as an alternative to cardiac catheterization in the detection of coronary lesions.¹ We present a pediatric case in which cardiac computed tomography (CCT) and cardiac myocardial resonance (CMR) failed to demonstrate left main coronary artery (LMCA) ostial atresia. This was confirmed at cardiac catheterization.

Clinical Report

An otherwise well 12-year-old boy was referred for evaluation of “intermittent syncope.” Typical semiology included sudden loss of consciousness (less than 2 minutes), in the setting of exercise and preceded occasionally by presyncope or palpitations. There was no relevant family history. Physical examination, resting 12-lead electrocardiogram and Holter recordings were unremarkable. There were no inducible symptoms or arrhythmia on exercise stress testing (EST) but significant ST depression developed over the lateral chest leads at peak exercise.

Echocardiography demonstrated a structurally and functionally normal heart. 2D echocardiography suggested both coronary arteries appeared to arise from the appropriate sinuses. However, antegrade color flow in the left coronary artery was difficult to elicit. CCT reported a mild focal dilata-

tion of proximal left anterior descending artery with otherwise normal coronary anatomy (► **Fig. 1**). A CMR stress perfusion test identified no abnormality. In view of the history and EST result, despite reassurance from noninvasive modalities, cardiac catheterization was performed with selective coronary angiography. This identified LMCA ostial atresia with extensive collateral arterial supply from the right coronary arterial (RCA) system (► **Fig. 2 a–c**). The left coronary artery system is relatively well developed and there is

Fig. 1 Cardiac computed tomography reported as both coronary arteries arising from appropriate sinuses.

Fig. 2 Cardiac catheterization. (a) Aortogram demonstrating prominent right coronary arterial (RCA) and absence of left coronary arterial orifice; (b, c) selective RCA angiogram demonstrating extensive collaterals communicating with the left coronary system.

retrograde flow demonstrated back to the left main stem perhaps explaining why the CT angiogram was misleading (→Fig. 2 a–c). The patient underwent coronary artery bypass grafting of the left internal mammary to the left anterior descending artery with an uncomplicated recovery.

Discussion

Coronary artery abnormalities are a rare but recognized cause of syncope and sudden death in childhood. Left coronary ostial atresia is a very rare congenital abnormality characterized by proximal obstruction with full development of the distal left coronary system (due to collateral supply from RCA). Symptoms are related to the quantity of collateral coronary blood flow.^{2,3} In this case, the extensive collateral supply accounted for symptoms only with exercise.

Debate over the different modalities of coronary angiography continues. The concept of invasive angiography as the gold standard is progressively challenged by noninvasive technologies (CT and MRI). The case for any mode of imaging is governed by the following two parameters: “accuracy” and “patient risk.” Advanced noninvasive imaging includes CCT and CMR. These modalities produce wide ranging information on issues such as plaque composition, myocardial perfusion, and ejection fractions.⁴ This is particularly relevant in adult populations (and less so in pediatrics) given the high prevalence of coronary heart disease. In fact, cardiac computed tomography angiography has been recommended as an effective screening tool to exclude obstructive lesions in low- and medium-risk individuals.⁵

This case highlights the discrepancies between imaging modalities. Despite the attendant (low) risk of cardiac catheterization, invasive coronary angiography was necessary to confirm the diagnosis. This illustrates the strength of invasive angiography over noninvasive imaging and supports the case for invasive coronary angiography as the gold standard imaging modality.

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Conclusion

Clinical suspicion and evidence of ischemia on exercise testing should be comprehensively investigated. Despite advances in cardiac-gated CT coronary angiography, cardiac catheterization remains the gold standard for identifying coronary artery anomalies.

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