Anomalous aortic origin of a coronary artery with an intraseptal course: novel techniques in haemodynamic assessment

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DESCRIPTION
A 7-year-old boy presented to the emergency room after an episode of syncope while at home. Although, it was non-exertional, it lasted 2 min and was preceded by angina-like chest pain. Vitals, physical examination, chest radiograph and ECG were normal. Subsequent cardiology evaluation led to an echocardiogram that showed anomalous origin of the left main coronary artery from the opposite sinus of Valsalva with adjacent ostia which was confirmed on CT angiography. In addition, it showed a long 2.5–3 cm deep intraseptal course of the left anterior descending (LAD) and short intraseptal course of the left circumflex (LCX) artery. Stress nuclear perfusion scan and Holter monitoring were negative, but stress cardiac MRI showed an inducible perfusion defect along the anterior interventricular septal wall most prominent at the mid-ventricular and basilar levels. Coronary angiography (figure 1 and video 1) and intravascular ultrasound demonstrated significant systolic compression of the intraseptal segment of the LAD and LCX (figure 1 and video 2). Fractional flow reserve (FFR), which determines ratio of flow in the coronary artery to aortic pressure, at rest and at maximal hyperaemia with 3 min of intravenous adenosine infusion at 140 mcg/kg/min, was also performed. FFR in the LAD and LCX declined from 0.89 to 0.79 and 0.88 to 0.74, respectively, demonstrating significant

Video 1 Selective coronary angiogram showing long intraseptal course and significant systolic compression of the intraseptal segment of left anterior descending coronary artery.

Video 2 Intravascular ultrasound showing significant systolic compression of the intraseptal segment of left anterior descending coronary artery.

Figure 1 Selective coronary angiogram of the left coronary artery, showing the intraseptal course (blue line) of the left anterior descending coronary artery in diastole (A) and systole (B). Intravascular ultrasound images (with chromaflow) obtained in the intraseptal segment of the left anterior descending coronary artery during diastole (C) and systole (D).
compromise in flow (figure 2). Surgical treatment likely would have required an infundibulotomy and intracavitary unroofing of the intraseptal coronary into the right ventricular outflow tract with unclear long-term results. Given the degree of invasiveness of such procedure and possible long-term morbidity, surgical therapy was not pursued at this time. He was exercise restricted and started on beta-blocker therapy.1–3

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REFERENCES

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