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Coronary Artery Stenosis in Asymptomatic Child after Arterial Switch Operation: Detection by Transthoracic Colour-Flow Doppler Echocardiography

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Running title: Coronary stenosis after arterial switch operation

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Abstract
Arterial switch operation has become the definitive anatomical correction for transposition of great arteries. Left coronary artery ostial stenosis was detected by transthoracic Doppler echocardiography as a flame like colour flow diastolic signal and coronary flow reserve was low; 1.3. It was treated successfully by a drug eluted stent. These findings could be diagnostic for coronary ostial stenosis.

Conclusion: Coronary artery stenosis can be detected with colour Doppler echocardiography. Assessment of coronary flow reserve provides information of the physiological significance of the coronary stenosis.

Key words: Arterial switch operation, Coronary flow reserve, Coronary ostial stenosis

Introduction
Coronary artery stenosis is a rare, but potentially life-threatening condition. It occurs after the arterial switch operation (ASO) for transposition of great arteries (TGA) in symptom-free subjects in 3-7.8% of cases (1-2). Selective coronary angiography is the gold standard in detecting coronary stenosis, but it provides limited insight into its physiological significance (2). It is invasive, includes risk of radiation and intra-procedure ischemia by occluding the ostium with the catheter tip. Transthoracic Doppler echocardiography (TTDE) is non-invasive, cheap, and repeatable. The coronary flow reserve (CFR) is the ratio of the maximal hyperaemic coronary flow velocity to resting flow velocity (3). It can be used to evaluate the physiological significance of coronary stenosis (4).
**Case Report**

Arterial switch operation was performed on one week old boy with d-TGA and normal coronary artery pattern (Yacoub type-A coronary tree) (5). The postoperative course was uncomplicated. At 5 years of age, he had normal physical activity level, ECG, exercise ECG and echocardiography. Routine coronary angiography showed left coronary ostial stenosis with retrograde filling of the left coronary artery from the right coronary artery and was treated by balloon dilatation with instant recoil. The procedure was complicated by intimal dissection along the origin of the left coronary artery but further operative treatment was postponed at this stage. Surgery is of high risk to be performed on asymptomatic child of 5 years of age.

At 9 years of age he continued to be asymptomatic and was playing ice-hockey. The method for recording of coronary flow by TTDE is described in detail elsewhere (6). A flame-like, colour-flow diastolic signal at the left coronary ostium indicated a high flow velocity due to stenosis (Figure. 1). The peak flow velocity (PFV) in diastole over the stenotic ostium was 200 cm/second (normal 30–40 cm/second); the average flow velocity (AFV) was 55 cm/second (normal, 25 cm/second) and the velocity time integral (VTI) in diastole was 62 (normal 16 cm). To assess the CFR adenosine 140 mcg/kg/minute was infused for 4 minutes. The CFR measured distal to the stenosis was reduced to 1.3 (normal >3.5 in young adults) indicating a hemodynamically significant stenosis (7). There was no ischemia at rest but, stress-induced ischemia was demonstrated by adenosine infusion on both magnetic resonance imaging and myocardial scintigraphy. The ischemia was localised in the anterolateral wall and in the apex of the left ventricle (Figure. 2 A and B).

Selective coronary angiography revealed significant 90% ostial stenosis of the left main coronary artery. The right coronary artery was dominant with collateral flow through to the left ventricle. A modified double guide wire technique was used and standard balloon
dilatation (Maverick 2 x 15 mm, 20 atmospheres) performed. There was instant coronary artery recoil after dilatation. Drug-eluting stent (Taxus Express 2.5–12 mm, 14 atmospheres) was deployed at the ostium and the proximal part of the left main coronary artery. A high-pressure balloon (Maverick 2.0 × 15 mm balloon) was dilated with a 20 atmospheres pressure up to 2 mm in diameter. The patient was successfully treated with no residual stenosis judged by angiography immediately after the stent implantation (Figure. 3). A drug eluted stent was chosen over bare metal because of the proximal location of the stenosis. The placement of the stent in this patient predated the recent concerns regarding drug eluted stents causing increased mortality, probably due to late stent thrombosis (8). Coronary flow studies by TTDE one day after the procedure showed normalised flow velocities: PFV was 70 cm/second, AFV 34 cm/second and VTI 25 cm. The flame-like, colour-flow diastolic signal at the left coronary ostium has disappeared. He was started on once-a-day acetylsalicylic acid (3–5 mg/kg/day) and clopidogrel bisulphate (3–5 mg/kg/day). These medications will be continued for 2 years and then acetylsalicylic acid alone for life to decrease the risk of late thrombosis. After 6 months follow up, selective left coronary angiography revealed widely open left coronary artery ostium with no restenosis. Repeat magnetic resonance and myocardial scintigraphy studies after 6 months demonstrated total regression of ischemia. No further surgery will eventually be needed, but there will be a further balloon dilatation of the drug eluting stent.

Discussion

Laminar flow signals by TTDE in the main left coronary artery and its two branches (anterior descending and circumflex artery) can be seen in over 90% of healthy children (9). The measurement of CFR distal to stenosis precisely defines the hemodynamic significance of stenosis (4). Ostial stenosis may be detected using colour flow Doppler as a flame-like signal.
This signal persists after increasing the Nyqvist limit to a level beyond that of the velocity in the affected coronary artery. It represents a vortex created by the stenotic ostium.

A diastolic colour-flow signal indicated high maximal spectral velocity with PFV of 200 cm/second, over the ostial stenosis. The flame colour sign is relatively easy to register, which makes the method an important diagnostic tool in the detection of coronary stenosis.

Adenosine infusion is well tolerated in children and can be done without anaesthesia or sedation. A low CFR value, 1.3 in our patient, supports the diagnosis of coronary ostial stenosis. A value of CFR < 2 indicates haemodynamically significant coronary stenosis (7). However, in our institution, intracoronary Doppler guide wire study after ASO found that the median CFR in the left anterior descending coronary artery was 3.7 (3.0–4.8) (10). TTDE with demonstration of flame-like colour flow diastolic signal at the coronary ostium and decreased CFR under adenosine infusion could be diagnostic of coronary ostial stenosis.

References


Legends to the figures

Figure 1. A flame-like colour-flow signal indicates left coronary artery (LCA) ostial stenosis.

Figure 2
Pannel A. A perfusion defect is observed by magnetic resonance imaging at the apex and at the mid-ventricular antero-lateral wall of the left ventricle during adenosine infusion (arrows) but not at rest. This indicated ischemia in this region.

Pannel B. A perfusion defect is observed by single photon emission computed tomography at the apex and the anterolateral wall of the left ventricle during adenosine infusion (arrows) but not at rest. This indicated ischemia in this region.

Figure 3.
A. Left coronary artery angiogram showing the ostial stenosis
B. Deployment of the stent
C. Left coronary artery angiogram after stenting
During Adenosine Infusion

During Rest
During Adenosine Infusion

During Rest