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Case Report

Anatomic Repair of a Left Coronary Artery Main Stem Atresia

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ABSTRACT

Atresia of the main stem of the left coronary artery is the least observed congenital coronary anomaly; most patients tend to receive a coronary artery bypass graft, although some anatomical corrections have been described. A 17-year-old female patient with left coronary artery main stem atresia underwent a coronary trunk construction with an autologous pericardial patch in our department. At a 3-year follow-up, the patient was asymptomatic, with a normal cardiac stress test. The coronary computed tomography showed no stenosis between the aorta and coronary bifurcation. Long-term patency has yet to be determined.

RÉSUMÉ

L'atrésie de l'artère coronaire gauche principale est l'anomalie coronarienne congénitale la plus rarement observée; la plupart des patients subissent un pontage aorto-coronarien, bien que certaines corrections anatomiques aient été décrites. Une patiente de 17 ans présentant une atrésie de l'artère coronaire gauche principale a subi dans notre service, à une construction du tronc coronaire effectuée à l'aide d'un patch péricardique autologue. Au suivi après 3 ans, la patiente était asymptomatique et a obtenu des résultats normaux à l'épreuve d'effort cardiaque. La tomographie par ordinateur de l'artère coronaire ne révélait aucune sténose entre l'aorte et la bifurcation coronaire. La perméabilité à long terme reste à établir.

Atresia of the main stem of the left coronary artery is the least common congenital coronary anomaly, with an absent left main trunk from the aorta. As a consequence, both the left anterior descending and circumflex arteries receive a retrograde blood flow due to collateral circulation from the right coronary artery. Most patients are symptomatic, but symptoms slightly differ according to the age at diagnosis. Paediatric patients often develop a failure to thrive, syncope due to ventricular arrhythmias, and heart failure, whereas teenagers and adults may develop angina. Surprisingly, most reported cases have undergone a coronary artery bypass graft to treat this congenital anomaly.[1] We describe a case of left main stem atresia that underwent surgical left main stem construction with a pericardial patch.

Case Report

A 17-year-old girl, with no medical history, had 2 episodes of palpitations plus concomitant chest pain. An electrocardiogram showed fast junctional tachycardia and global myocardial ischemia associated with a significant rise in troponin. The echocardiogram showed no abnormalities. Coronary computed tomography and coronarography confirmed atresia of the left main stem of the coronary artery: no filling of the left main coronary artery was found from the aortic root, and massive collaterality from the right coronary artery (Fig. 1A). Cardiac magnetic resonance imaging revealed altered coronary reserves in the left anterior descending region as well the circumflex arteries, but with a lower impact.

The patient underwent surgical correction with a cardiopulmonary bypass and transection of both the pulmonary artery and ascending aorta. No ostium was found in the left coronary sinus of the aortic root; the proximal part of the left main stem was identified as a fibrotic chord (length 10 mm), connected distally to permeable bifurcation of the left anterior descending and circumflex arteries. We established a neotrunk joining the aortic root to the left anterior descending and circumflex bifurcation. Initially, left-facing ostium was created by a vertical incision of the aortic root wall. Then, the left main stem bifurcation was opened. Both aortic and coronary incisions were then joined together across the fibrotic segment by stitching an autologous fresh pericardial patch, which created continuity between this new ostium and the origin of the left anterior descending and circumflex arteries (Fig. 2).

The postoperative course was unremarkable and the patient was discharged 9 days after surgery with antiplatelet

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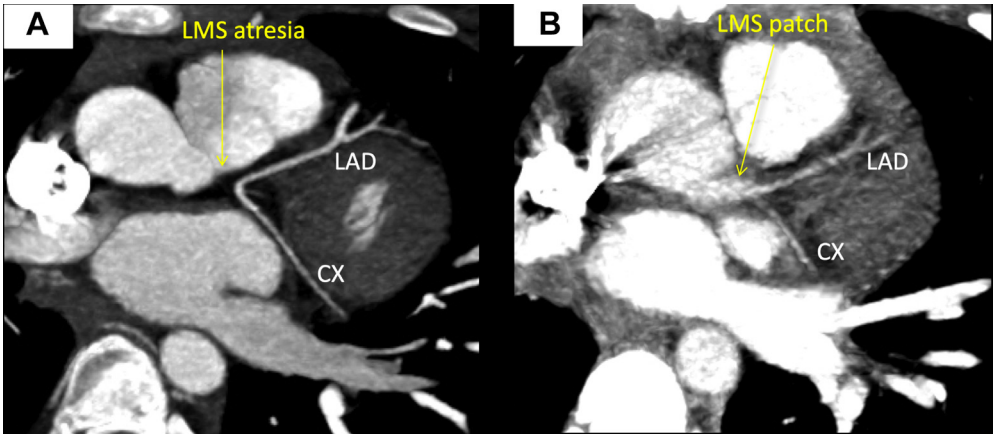

Figure 1. Coronary computed tomography comparing (**A**) preoperative and (**B**) 3-year postoperative. CX, circumflex artery; LAD, left anterior descending artery; LMS, left main stem.

therapy. The patient suffered no recurrence of episodes of junctional tachycardia and was free of chest pain. At 1 and 3 years after surgical correction, cardiac stress tests were negative and coronary computed tomography showed good permeability of the constructed left main trunk (Fig. 1B).

Comment

Congenital atresia of the ostium of the left main coronary artery carries an unfavourable prognosis; almost all patients will eventually develop myocardial ischemia and the high risk of sudden cardiac death is well established in this setting.[1] Tanawuttiwat et al.[2] reviewed 53 cases of left main coronary artery atresia in 2013. Roughly half of the cases did not receive surgical revascularization and their outcomes have been poor.[2] Four surgical reconstructions of the left main coronary artery were performed, whereas 23 coronary artery bypass graftings were performed.[2] However, paediatric coronary artery bypass grafting has medium outcomes in the Kawasaki population: 20-year graft patency was 87% for the internal thoracic artery, but was only 42% for the saphenous vein graft, and event-free survival was 60% in the 25-year follow-up.[3]

From the 2000s onwards, some authors have described different types of left main trunk construction that allow antegrade blood flow to the anterior interventricular and circumflex arteries, which preserves the possibility of a coronary bypass several years later. We decided to build a neo-trunk of the left main coronary artery with a fresh autologous pericardial patch, because of its availability and pliability that allowed us to create a funnel shape with the patch; at 3 years, the pericardial patch was not dilated or calcified. Others have used it instead of saphenous vein patch for surgical reconstruction of left main coronary artery stenosis.[4] This surgical strategy was effective for mid- to

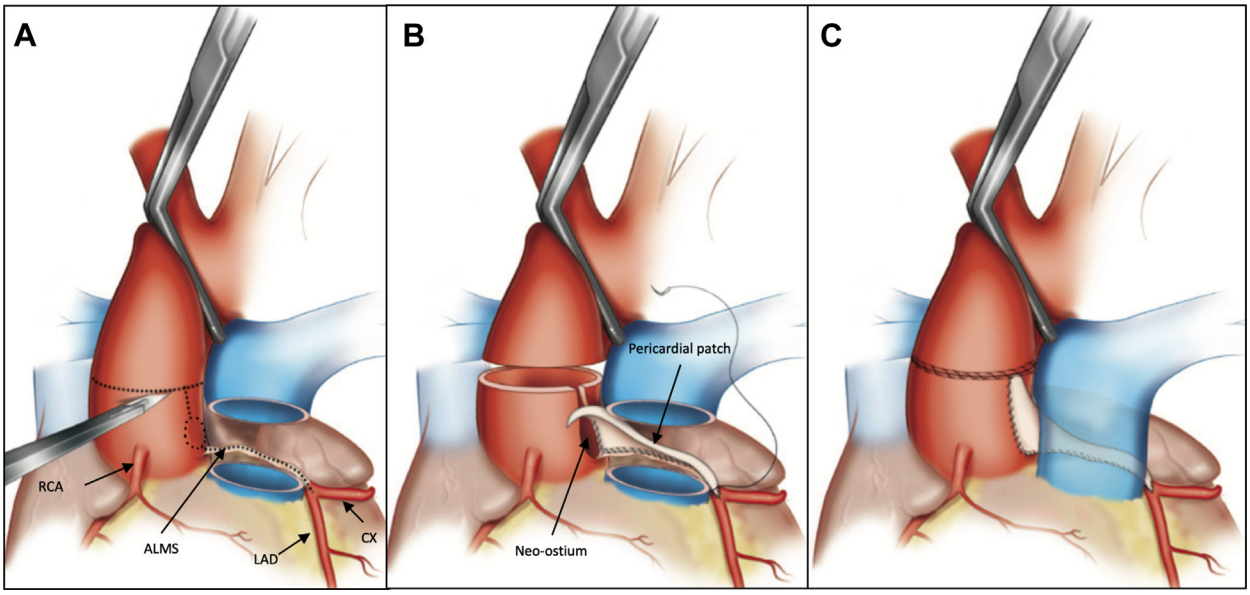

Figure 2. Operative illustrations of left main coronary artery atresia as (**A**) a fibrotic cord and (**B**, **C**) neo-ostium formation using a pericardial patch. ALMS, atresia of the left main stem; CX, circumflex artery; LAD, left anterior descending artery; RCA, right coronary artery.

long-term follow-up.[4] Kaczorowski et al.[5] used a pulmonary homograft patch to construct the left main stem in 2 reconstructions. Having a left main trunk surgical angioplasty avoids and spares the bypass graft material in patients, and an endovascular procedure may still be performed. Various patch materials have been used without a clear superiority.

In conclusion, atresia of the left main stem is an uncommon congenital coronary anomaly, and surgical autologous pericardial patch angioplasty is safe with good 3-year outcomes. This anatomical repair should be first considered over coronary artery bypass grafting and could offer greater opportunities for the long-term outcomes of children and young adults.

Disclosures

The authors have no conflicts of interest to disclose.

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