Aorto-Left Ventricular Tunnel Arising From the Left Coronary Sinus of Valsalva

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ABSTRACT

Aortico-left ventricular tunnel is a rare congenital cardiac defect that bypasses the aortic valve via a para-valvular connection from the left ventricle to aorta. In most cases, the tunnel arises from the right aortic sinus. In this case report, we present a case of Aortico-left ventricular tunnel, in which the aortic orifice arose from the left aortic sinus, requiring special attention to avoid injuring the left coronary artery at the time of surgical repair.

Key words: Aortico-ventricular tunnel, Aortic regurgitation, Congenital cardiac defect.

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Introduction

Aortico-left ventricular tunnel (ALVT) is an extremely rare cardiac malformation characterized by a paraavalvular communication between the aorta and the left ventricle.\(^{(1)}\) Most commonly the tunnel\(^{(2)}\) is located above the right coronary sinus\(^{(3)}\) due to the thin left ventricular anterior wall where the right aortic sinus meets the membranous septum.\(^{(2)}\) This lesion represents about 0.1% of congenital cardiac malformations.\(^{(4,5,6)}\) The aortic orifice of the tunnel is usually located in the anterior wall of the aorta, above the right sinus of Valsalva (and the right coronary artery ostium). Occasionally, it can be found above the left coronary artery ostium. The ventricular orifice of the tunnel opens to the left ventricle just below the right and left aortic cusps. This malformation was first described by Levy and colleagues in 1963.\(^{(1)}\) We herein report a case of aortic-left ventricular tunnel, of which the aortic orifice arose from the left aortic sinus, requiring special attention to avoid injuring left coronary artery at the time of surgical repair.

Case Report

A 5-moth-old infant, who was a product of full term normal vaginal delivery, and uneventful perinatal history, was referred to Queen Alia Heart Institute in Amman-Jordan for cardiac evaluation and management. His cardiovascular examination revealed normal second heart sound, a grade III/VI to-and-fro murmur over the third left intercostal space with radiation toward the left axilla and bounding peripheral pulses. His chest x-ray revealed cardiomegaly with cardiothoracic ratio of 65% and increased pulmonary vascular markings. His EKG revealed a normal sinus rhythm with left ventricular hypertrophy. 2D-Echo showed aortic valve stenosis with maximum pressure gradient of 65 mmHg and trivial regurgitant flow, through both the aortic
valve and a tubular communication between the ascending aorta and the left ventricle (Fig. 1). The left ventricle was moderately dilated, but with good systolic ventricular function and a fraction shortening of 35%. Aortic angiogram revealed the tunnel which arose from above the left sinus of Valsalva and filled the left ventricle (Fig. 2). He underwent surgical intervention on mild hypothermia (32°C). After cross clamp the aorta was opened and the sinuses were inspected. The proximal opening of the tunnel arose from the left coronary sinus at the level of sinotubular junction (Fig. 3), and it was closed with a Gore-Tex patch using 5.0 prolene continuous sutures (Fig. 4). The distal tunnel was opened and the ventricular opening was visualized from inside the tunnel and closed with another Gore-Tex patch using 5.0 prolene continuous sutures. Finally, the tunnel was closed with 5.0 prolene. The aortic valve was tri-leaflet with fusion between the right and left coronary cusps for which an aortic commissurotomy was done. After closure of the aortic incision and declamping of the aorta, the patient was weaned from cardiopulmonary bypass without any difficulty. He had a smooth postoperative course.

The patient was discharged home on the seventh postoperative day. His last follow up at the outpatient clinic was 6 months after surgery. His 2D-Echo Doppler showed good systolic left ventricular function with mild aortic valve stenosis, a maximum instantaneous maximum pressure gradient over the aortic valve of 30mmHg and grade I aortic regurgitation.

**Discussion**

The incidence of ALVT is very low. Martins et al. reported 0.001% in patients with congenital heart disease at their institution's experience,(6) whereas Mckay et al. reported incidence of less than 0.1% in a clinico-pathological series.(7,8) More than 100 patients with ALVT have received surgical treatment. In most of the cases, the aortic orifice of the tunnel arose from or above the right coronary sinus, and the tunnel was located antero-laterally to the ascending aorta. However, our case demonstrated that the aortic orifice arose from the left coronary sinus and the tunnel lay postero-laterally to the ascending aorta. Reviewing the literature, we could find only 4 cases in which the aortic orifice opened from the left aortic sinus.(9-12) In this
situation, incision of the tunnel is difficult and the tunnel should be closed through aortic incision. As for the operative technique, Serino and colleagues report that closing the aortic defect by direct suture distorts the cusps by pulling them toward the weak aortic wall, which remains unsupported within the dilated aortic sinus. From this point of view, the patch technique is believed to reduce the risk. Careful follow up is needed in this anomaly after the surgical repair.

References