Aorto-Left Ventricular Tunnel – A Case Report

Abstract
Aorto-left ventricular tunnel (ALVT) is a rare congenital anomaly with extracardiac channel connecting the ascending aorta above the sinotubular junction to either left or right ventricular cavity.[1] The aortic opening of most tunnels lies above the right coronary sinus. It may be associated with aortic valve and/or coronary artery anomalies. Patients usually present in infancy with features of congestive heart failure due to nonvalvular aortic regurgitation.[1,2] Diagnosis is mainly done by echocardiography. In cases with inadequate information about coronary artery anatomy, cardiac catheterization is required.[3] Surgical closure is done at both ends of the tunnel. Transcatheter device closure can be done with favorable anatomy. Long-term outcomes are good with early intervention.[2]

Keywords: Aorto-left ventricular tunnel, congenital, morphology

Introduction
Aorto-left ventricular tunnel (ALVT) is a rare congenital extracardiac channel connecting the ascending aorta above the sinotubular junction to either left or right ventricular cavity.[1] The aortic opening of most tunnels lies above the right coronary sinus. It may be associated with aortic valve and/or coronary artery anomalies. Patients usually present in infancy with features of congestive heart failure due to nonvalvular aortic regurgitation.[1,2] Diagnosis is mainly done by echocardiography. In cases with inadequate information about coronary artery anatomy, cardiac catheterization is required.[3] Surgical closure is done at both ends of the tunnel. Transcatheter device closure can be done with favorable anatomy. Long-term outcomes are good with early intervention.[2]

Case Report
An 11-year-old male child presented with palpitations and dyspnea. To and fro murmur (5/6) was present in the left third intercostal space. On transthoracic echocardiography, it was diagnosed as coronary cameral fistula which was also confirmed by magnetic resonance imaging [Figure 1]. The child was taken to catheterization laboratory for device closure. On angiography, it was diagnosed to be a large aorta to left ventricular tunnel; the origin of left main coronary artery (LMCA) was not visualized for device closure of the tunnel [Video 1]. Hence, surgical closure was done. Intraoperatively, it was a large tunnel arising from the left coronary sinus giving origin to LMCA. The tunnel was traversing through the transverse sinus, posterior to the main pulmonary artery (MPA) and then downward to open in to the anteroseptal wall of left ventricle (LV). The MPA was transected, and the roof of the tunnel was seen and opened. The opening of the LMCA was seen in the tunnel. The tunnel was closed distal to the origin of LMCA with glutaraldehyde-treated pericardial patch. The ventricular opening was not accessible. The tunnel roof was closed and MPA anastamosed back. Aortic valve was tested which was competent and aorta closed. There was no difficulty in weaning patient from cardiopulmonary bypass. Postoperative recovery was uneventful.

Discussion
The aortic opening of the aorto-ventricular tunnel commonly lies above the right coronary sinus and rarely arises from left coronary sinus.[3,4] It usually communicates with LV in the fibrous triangle, beneath the left and right coronary commissure. When it opens into right ventricle, it is immediately above or below the subpulmonary infundibulum.[4]

In our case, the tunnel originated from the left coronary sinus. The LMCA was arising within the tunnel, making device closure unlikely. The aortic end could be closed as LMCA origin was proximal. Distal origins are difficult to close, and only the
ventricular end is closed in such cases to maintain coronary perfusion.\textsuperscript{[4]}

Tunnels lying above the left sinus or intercommisural area have different morphology and open into LV away from aortic valve. It is rarely seen to enter the LV cavity through myocardium which differentiates it from coronary cameral fistula. But in this patient, the ventricular end of the tunnel was opening in to the anteroseptal wall of the LV, quite distal to the aortic valve, partially through myocardium, making preoperative diagnosis as coronary cameral fistula. Its opening was not accessible through aorta or the left atrium and could not be closed. Hence, this child will require long-term follow-up for aneurysmal dilatation of LV due to the residual tunnel at ventricular end and also aortic valve incompetence.

Echocardiography is the single most important investigation for the diagnosis of ALVT. Parasternal views are used on transthoracic echocardiography in understanding the origin of the tunnel above the coronary ostium, its length, and opening into one of the ventricles.\textsuperscript{[1]} On Doppler, diastolic flow is seen from aorta to ventricle and systolic flow from ventricle to aorta. In tunnels arising from right sinus, the entire tunnel can be seen by clockwise rotation of the probe\textsuperscript{[4]} The size of the defect, both at entry and exit points, severity of the nonvalvular and valvular regurgitation, associated anomalies, LV dimensions, and functions are also assessed.

In our case, on transesophageal echocardiography, the origin of the tunnel was seen in mid-esophageal aortic valve short-axis view [Video 2]. It was arising from the left coronary sinus unlike the usual right sinus. The mid-esophageal long-axis views did not show the origin of the tunnel as it was arising posterior to the aorta. The ventricular opening of the tunnel was seen well in the basal transgastric short-axis view [Video 3]. It was not possible to trace the entire tunnel due to the unusual morphology, arising from left sinus posteriorly and opening into anteroseptal area [Video 4]. Doppler showed the diastolic flow from aorta to LV. The origin of left coronary could not be assessed as it originated within the tunnel. There was no involvement of the aortic valve. The LV was dilated and the functions were normal. There were no other anomalies like right ventricular outflow tract or pulmonary valve obstruction. Postoperatively, there was no residual flow across the aortic end [Video 5]. At the ventricular end, residual flow was seen as it was not closed [Video 6]. ALVT has to be differentiated from ruptured sinus of Valsalva and coronary cameral fistula. ALVT never passes through myocardium to reach the cavity of the ventricle, a feature that differentiates it from coronary–cameral fistula. A ruptured sinus of Valsalva aneurysm has its orifice in the sinus of the aortic valve.

**Conclusion**

We present a case of a rare congenital heart disease – the AVLT with unusual morphology. The tunnel originated from left coronary sinus and involved the LMCA which is also rare. Most importantly, the tunnel partially traversed the myocardium as in coronary cameral fistula which is very rare.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Kathare P, Subramanyam RG, Dash TK, Muthuswamy KS, Raghu K, Koneti NR. Diagnosis and management of aorto-left ventricular tunnel. Ann Pediatr Cardiol 2015;8:103-7
2. Martins JD, Sherwood MC, Mayer JE, Keane JF. Aortico-left ventricular tunnel: 35-year experience. J Am Coll Cardiol 2004;44:446-50.
3. Sadeghpour A, Peighambari M, Dalirrooyfard M, Esmailzadeh M, Maleki M, Nooli F, et al. Aorta-to-left ventricle tunnel associated with noncompaction left ventricle. J Am Soc Echocardiogr 2006;19:1073-e1.
4. McKay R. Aorto-ventricular tunnel. Orphanet J Rare Dis 2007;2:41.
5. Ono M, Goerler H, Boethig D, Breymann T. Surgical repair of aortico-left ventricular tunnel arising from the left aortic sinus. Interact Cardiovasc Thorac Surg 2008;7:510-1.