



Aneurysmal Aorto-Left Ventricular Tunnel

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ABSTRACT

Aorto-left ventricular tunnel (ALVT) is a congenital anomaly of aortic root which is an extra cardiac connection between the aorta and the left ventricle. It is usually short and direct but this report describes an aneurysmal aorto-ventricular tunnel which due to large left ventricular and small aortic hole culminated in aneurysm. This type of ALVT might be misdiagnosed as other cardiac lesions.

► Implication for health policy/practice/research/medical education:

The case report is an unusual form of left ventricle to aortic tunnel which can be misdiagnosed.

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1. Introduction

Aorto-left ventricular tunnel (ALVT) is a rare congenital anomaly of aortic root that is an extra cardiac, vascular connection between the aorta and the left ventricle. It is usually short and direct but may be aneurysmal. Aorto-ventricular tunnel must be distinguished from other lesions which cause rapid run-off of blood from the aorta and lead to heart failure.

2. Case Report

An 18-month-old male infant with normal growth referred due to heart murmur and excessive sweating. The patient was visited by pediatric cardiologist and his parents were informed about him having a small inter-ventricular septal defect. On clinical examination, patient had Hyperdynamic precordium, normal S1 and S2, systolic click and to-and-fro murmur grade III/VI which was heard at the right upper sternal border radiating to supra sternal area.

Echocardiography showed small defect in sub-aortic area with bidirectional shunt, bicuspid aortic valve and trivial aortic regurgitation. (Fig. 1,2,3).

In angiography aneurysmal dilation was seen upper to right coronary cusp with normal right coronary artery, and bicuspid aortic valve without aortic regurgitation. (Fig. 4)

The same aneurysm and normal coronaries were shown in 64-Slice CT angiography (1).

Therefore, the diagnosis made was an aneurysmal dilation of aorto-left ventricular tunnel that was very rare.

It was thus decided to carry out surgical correction and closure of tunnel. In surgery aneurysm was found to have a large hole in left ventricle and a small hole in aorta, an anatomy which caused aneurysmal dilation of tunnel with pressure effect on inter-ventricular septum and bulging of septum into the right ventricle. Path closures of holes were done with no postoperative complication. (Fig. 6,7)

3. Discussion

Aorto-left ventricular defect is a rare anomaly of aortic root which accounts for 0.001% of all congenital heart diseases (2). In 90% of patients with aorto-ventricular

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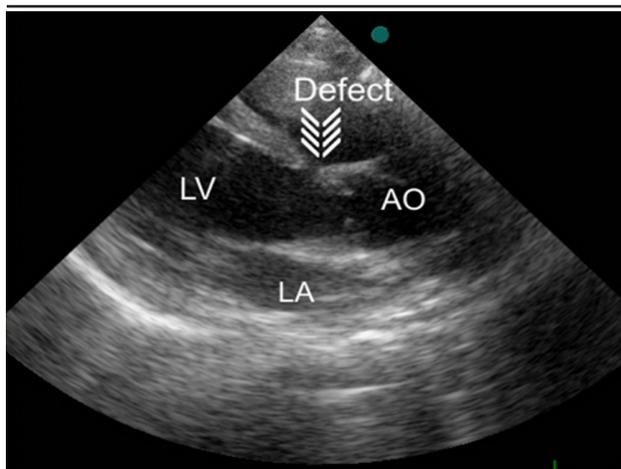


Figure 1. The defect which is like a small ventricular septal defect in pre-membranous part of septum in parasternal long axis view.

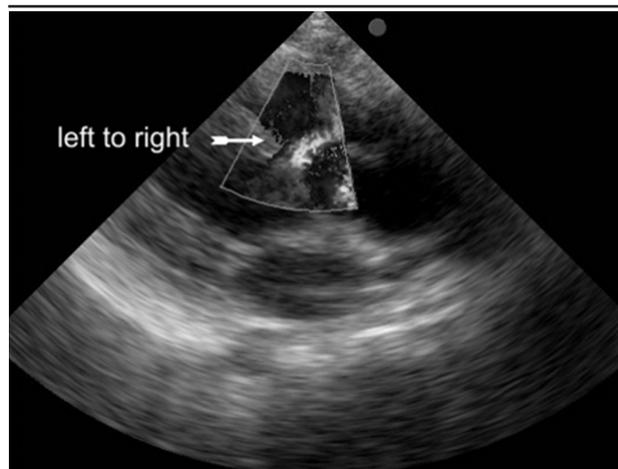


Figure 2. Left to right blood shunt through the defect in parasternal long axis view.

defect, the tunnel was connected to the left and in about 10% to the right ventricles (3).

The opening of tunnel in 40% of patients is above, in 25% below and in 16% at the level of right coronary artery (4).

Aortic regurgitation is the most common associated anomaly in LAVT and may be due to weakness of aortic sinus wall (5).

Many patients may present with congestive heart failure during the first year of life and also the patients may have signs resembling aortic incompetence including wide pulse pressure with low diastolic blood pressure, hyperactive dilated left ventricle, enlarged left atrium and load to-and fro murmur at the base of heart (1).

Male to female ratio is about 2:1. The etiology of ALVT is unknown but it seems that the tunnel may result from mal-development of the cushions that make the pulmonary and aortic roots, and abnormal separation of the pulmonary and aortic roots (3).

Chest radiography may reveal cardiomegaly, with dilation of ascending aorta found in all patients. The electrocardiogram shows varying degrees of left ventricular and atrial hypertrophy. Echocardiography is the gold standard for diagnosis of ALVT. It is possible

to reliably diagnose aorto-ventricular tunnel on fetal echocardiography. The differential diagnosis of this lesion includes rupture of sinus of valsalva, ventricular septal defect, aortic incompetence and coronary artery-left ventricular fistula. Echocardiography with Doppler color flow mapping and aortography can easily differentiate ALVT from above- mentioned lesions. Echocardiography with Doppler color flow mapping and aortography can distinguish this lesion from aortic insufficiency by the absence of retrograde flow through the aortic valve; from a coronary artery left ventricular fistula by normal right and left main coronary arteries; from an associated ventricular septal defect by the absence of a left-to-right shunt through the defect and no right ventricular overloading; and from a ruptured sinus of valsalva by the anterior position of the tunnel and the absence of a dilated sinus of valsalva.

In this case large left ventricular hole of the defect and small aortic hole made aneurysmal dilation of tunnel without any significant heart failure.

The treatment of ALVT is early surgical correction to prevent the development of aortic incompetence (1).

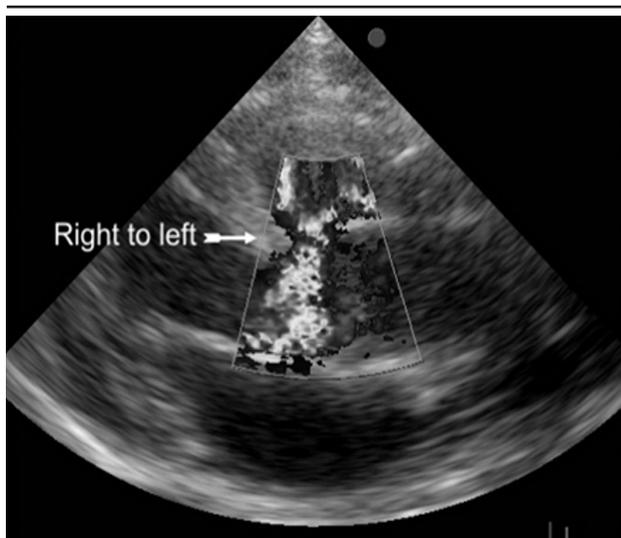


Figure 3. Right to left blood shunt through the defect in parasternal long axis view.

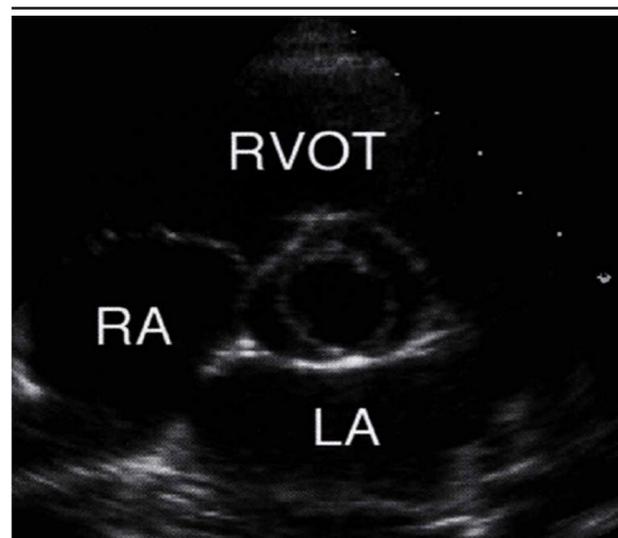


Figure 4. Bicuspid aortic valve in parasternal short axis view. RA: right atrium; LA: left atrium; RVOT, right ventricular outflow tract.

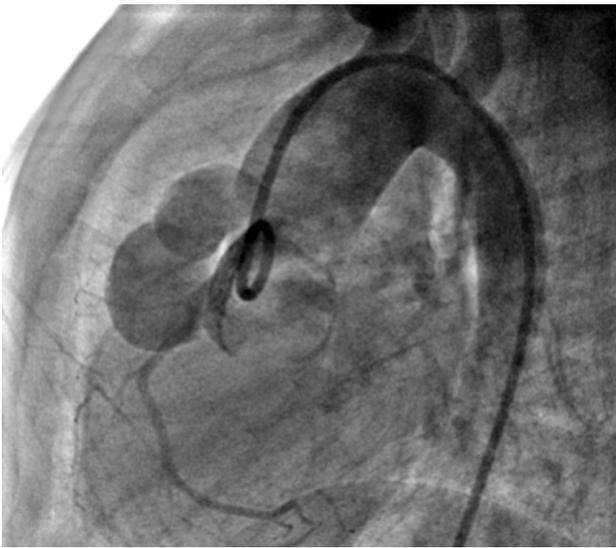


Figure 5. Aortogram in lateral view shows bicuspid aortic valve and aneurysmal dilation superior to right aortic cusp.

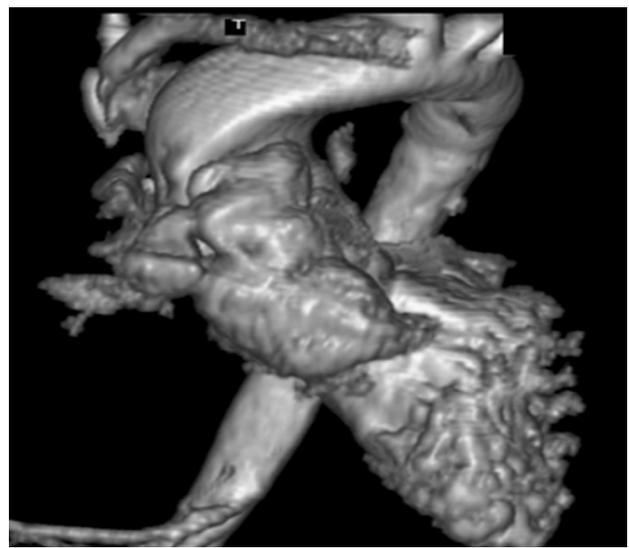


Figure 6. 64-slice computerized angiography shows aneurysmal dilation superior to right aortic cusp.

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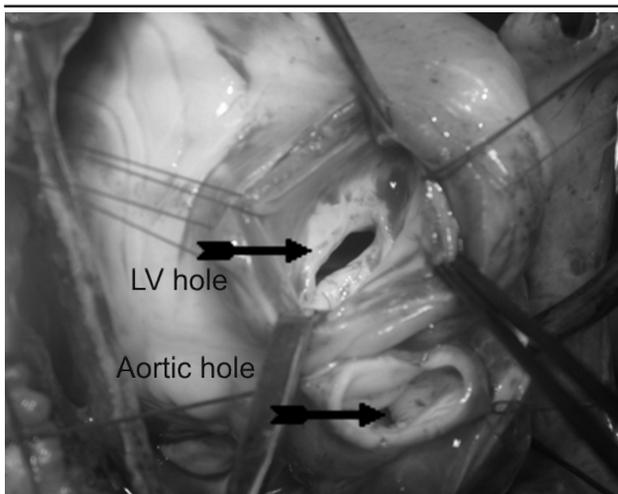


Figure 7. Large left ventricular (LV) hole and small aortic hole.

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