Aneurysmal Aorto–Left Ventricular Tunnel and Bicuspid Aortic Valve with Severe Stenosis

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History
A 60-year-old male presented with increasing shortness of breath over 2 years.

Physical Examination
The heart rate was 80 bpm, with frequent premature beats. On auscultation, there were 3/6 systolic and diastolic murmurs at the left parasternal edge in the third intercostal space.

Transthoracic Echocardiogram
A transthoracic echocardiogram showed that the aortic valves were severely calcified (Figure 1-1*) with stenosis (peak velocity was 6 m/s) and mild regurgitation. An aorto–left-ventricular tunnel (ALVT) (T) with entry from the aorta (thick arrow) and exit (thin arrow) into the left ventricle (LV) was seen (Figure 1). Blood flows from the aorta into the ALVT (T), and then into the LV cavity which can be seen by the color Doppler (Figure 1-1B). Left ventricle hypertrophy with normal systolic function was noted (Video 1.1). Nonvalvular regurgitation (thick arrow) and a shunt from aorta into ALVT (thin arrow) was seen by color Doppler (Figure 1-1D, Video 1.2).

A computer angiogram showed that the ascending aorta was dilated, with an aneurysmal ALVT (T) arising from the aortic root and connected with the LV (*) (Figure 1-2A). The aortic annulus, aortic valves, and tunnel (T) were severely calcified (arrow), (Figure 1-2B). The connection (*) between the aortic root and ALVT (T) could be seen (Figure 1-2C). Three-dimensional reconstruction computed tomography imaging showed aneurysmal ALVT (arrow), left coronary artery (LCA) arising from aortic root (arrow) behind the aneurysmal ALVT, and significantly dilated ascending AO (Figure 1-2D).

Management
The patient underwent open heart surgery. Surgical inspection revealed a 3 cm aneurysmal tunnel with smooth surface, it was (white arrow) arising from the left Valsalva sinus, located at below the LCA orifice (Figure 1-3). The aortic valve was bicuspid and severely...
The aortic and LV orifices of the ALVT were repaired, and the aortic valve was replaced. The patient recovered well after the operation. A follow-up echocardiogram showed a well-seated and functioning prosthetic aortic valve.

**Discussion**

An aorto–left–ventricular tunnel (ALVT) is a congenital extracardiac channel connecting the ascending aorta to either left or right ventricular cavity. It is extremely rare with incidence as low as 0.001% of all congenital heart diseases [1]. Hovaguimian and colleagues [2] classified the ALVT into four types (I, II, III, and IV) that have a bearing on the appropriate surgical techniques of repair: A slitlike opening at the aortic end with no valve distortion in type I (prevalence, 24%), a large extracardiac aneurysm in type II (44%), an intracardiac aneurysm of the septal portion of the tunnel with or without right
ventricular outflow tract obstruction in type III (24%), and a combination of types II and III in type IV (8%). In most cases, the aortic orifice of the tunnel arises from the right coronary sinus and is on the anterolateral side of the ascending aorta [2]. The ALVT rarely arises from the left coronary sinus, and there are only a few reports of this presentation [3, 4].

In our patient the aortic orifice of the aneurysmal tunnel arises from the left coronary sinus below the orifice of the left coronary artery. The exit of the tunnel into the left ventricle appears narrow and has severe calcification. This patient had congenital ALVT associated with a bicuspid aortic valve with severe calcification and stenosis. To the best of our knowledge, this is very rare in the literature.

**Associated Anomalies**

Aortic valve abnormalities like dysplastic or bicuspid valve with stenosis are frequent [5], but aortic atresia has rarely been reported [6]. Stenosis of the pulmonary valve [7] and subvalvular pulmonary obstruction due to a tunnel have been reported [8]. Proximal coronary anomalies like coronary ostium lying within the tunnel or atresia of coronary ostium have been documented [9].

**Clinical Presentation**

The clinical presentation of ALVT varies depending upon the compression of the coronary arteries, the presence of right or left ventricular outflow obstruction, and the diameter of the tunnel [10–12]. Congestive heart failure frequently develops during the first year of life [10]. The onset, severity, and progression of heart failure vary and range from *in utero* fetal death [13] to asymptomatic adulthood [14], and this depends on the cross-sectional area of tunnel and the amount of aortic regurgitation [15]. Chronic preload due to regurgitation leading to LV dilatation is seen in asymptomatic grown-up patients. Early diagnosis and surgical correction are essential to prevent irreversible myocardial dysfunction and heart failure. Untreated cases may progress to the development of...
native aortic valve regurgitation. The development of symptoms may be delayed if the tunnel terminates in the right ventricle and has a significant right ventricular outflow tract obstruction, thereby limiting the magnitude of the shunt [16].

In our case, the tunnel terminus was narrow, which limited the regurgitation from the aorta and resulted in the ALVT becoming aneurysmal. Due to a small amount of regurgitation, there was no left ventricular dilatation and heart failure until the patient was 60 years old. Shortness of breath is caused by bicuspid aortic valve stenosis.

**Diagnosis**

Echocardiography is the most important test for the diagnosis of ALVT; cardiac catheterization is required only in those cases with inadequate information about coronary artery anatomy [17]. Parasternal views are particularly useful in understanding the origin of the tunnel and its relation with the coronary ostium, its length and opening into one of the ventricles. An ALVT never passes through myocardium to reach the cavity of the ventricle, a feature that differentiates it from a coronary-cameral fistula. Fistulous connections of the coronary arteries, however, always pass through the myocardium to reach the lumen of a cardiac chamber, and do not involve the hingepoint of an aortic valvar leaflet. As we will see, these features do not always serve to distinguish a fistula from a tunnel extending to open within the right ventricle but they do contrast with most tunnels that open within the left ventricle [18]. Another close differential diagnosis is a ruptured sinus of Valsalva aneurysm, which has its orifice in the sinus of the aortic valve.

The computer tomogram has the advantage of providing three-dimensional information, which helps in understanding the close relation around ALVT. In our case, the CT images clearly indicate the anatomic arrangement underscoring the malformations.

**Treatment**

Without surgical treatment, most patients die early in life due to congestive heart failure. Therefore, medical management should only be to prepare the patient for surgery. Transcatheter closure. Although, aorto-left ventricular tunnel could be treated by transcatheter closure using amplatzer duct occluder in literatures [19, 20], it is not the established method of tunnel closure due to the complex anatomy of the tunnel, such as its proximity to the aortic cusp and right coronary ostium, or the coronary ostium inside the tunnel wall, or already distorted cusp anatomy with valvular regurgitation. The tunnel itself is a significantly distensible structure. Hence, careful selection is needed for transcatheter closure.

**Surgical Management**

Volume overload due to severe regurgitation is a feature of ALVT; hence, it warrants correction as soon as the diagnosis is made. Survival following surgical repair has improved from around 20% to nearly 100% [21].

Even if the patients are treated, they should be followed up in case the tunnel persists, and also for aortic aneurysm because of the occurrence of aortic insufficiency in the long term and worsening heart failure. The incidence of aortic insufficiency in patients with ALVT following surgery ranges from 16–60%, and the requirement for aortic valve replacement ranges from 0–50% [22, 23].
Key Points
1. Aorto-left ventricular tunnel is a rare congenital extracardiac channel with progressive left ventricular dilatation, which needs early correction.
2. After the treatment, all patients should be followed up for tunnel recurrence, aortic valve incompetence, left ventricular dysfunction, and aortic aneurysm throughout their lives.
3. In ALVT, coronary artery abnormalities and other associated abnormalities should be observed.

References
