A 25-year-old male presented to us with multiple episodes of syncope during the past 6 months. Physical examination was remarkable for bradycardia, an increased pulse pressure, and a systolic-diastolic murmur associated with thrill at the aortic area. ECG (Figure 1) showed complete heart block with a wide QRS escape. Two-dimensional transthoracic echocardiography with color Doppler (Figures 2 and 3 and Movies I–IV in the online-only Data Supplement) showed the presence of a calcified aneurysmal structure arising from the ascending aorta just above the sinutubular ridge, a left ventricular outflow tract (LVOT) aneurysm, a submitral left ventricle aneurysm, along with severe aortic regurgitation and infundibular pulmonic stenosis (PS). Multidetector computed tomography with volume rendering (Figures 4 and 5) clearly demonstrated the aortic and the left ventricular site of communication of the aneurysmal aorto-left ventricular tunnel (ALVT) as well as the unusual course of the ALVT burrowing into the proximal interventricular septum. The communication of the ALVT with the LVOT aneurysm was clearly demonstrated on multidetector computed tomography, as were the submitral aneurysm and infundibular PS. The PS was seen to be the result of a combination of infundibular hypertrophy and compression by the aneurysmal ALVT. The patient was implanted with a permanent pacemaker and referred to cardiothoracic surgery for definitive management.

ALVT is a rare congenital entity, with ≈130 cases having been described in literature so far. It is distinguished from a ruptured sinus of Valsalva aneurysm by having its aortic connection above the sinutubular ridge. Various hypotheses have been put forward regarding its cause. Some authorities believe it to be an abnormal coronary artery; others have ascribed it to early aortic dissection, and still others have attributed it to persistent embryonic rests of the fifth aortic arch or to malformation of the distal bulbus cordis. There is no specific genetic defect identified to date. However, cystic medial degeneration has been demonstrated in an aortic aneurysm from a patient with repaired ALVT, making connective tissue disease an important contender for the underlying defect. The presence of multiple cardiac aneurysms in this patient adds credence to this hypothesis. However, although the patient did have a marfanoid habitus with a reduced upper to lower segment ratio and a positive wrist and thumb sign (Figure 6), he did not meet the revised Ghent criteria for Marfan syndrome. There was no family history of Marfan syndrome and no ocular involvement. Genetic testing could not be performed because the facility was not available to us.

ALVT is commonly associated with other cardiac anomalies, the most frequent being coronary artery anomalies followed by aortic valve abnormalities. However, its association with other cardiac aneurysms has not been reported. On the right side, pulmonary valvular involvement occurs in ≤5% of cases. Significant right ventricular outflow tract (RVOT) obstruction in the context of ALVT has primarily been described postoperatively, when the ventricular end of the tunnel is left open while the aortic end is closed during repair. Nevertheless, RVOT obstruction attributable to compression by an aneurysmal ALVT is a possibility. However, the marked infundibular hypertrophy contributing to RVOT obstruction in this patient remains unexplained and may be unrelated.

Aortic regurgitation (AR), as seen in this patient, is a common accompaniment, especially in older patients. Although early surgery may be protective for future development of AR, progression of AR may occur even after successful surgical closure of the tunnel if the right coronary leaflet is not adequately supported.

The most common mode of presentation of ALVT is with heart failure in infancy, although the clinical course is highly variable, ranging from death in utero to presentation with heart failure in adulthood. Despite the anatomic proximity of ALVT with the cardiac conduction system, presentation with heart block is characteristically uncommon. This is explained by the fact that most ALVTs do not penetrate the septal musculature, having their ventricular opening in the fibrous triangle just below the left and right coronary cusps. The unusual course of the tunnel in this patient, burrowing through the proximal interventricular septum, is what possibly caused him to present with complete heart block, before symptomatic LV dysfunction could set in.
This patient’s presentation with multiple cardiac aneu-rysms, infundibular PS, and complete heart block not only adds to the spectrum of ALVT as we know it, but also makes a strong case for underlying connective tissue defect as a likely factor in the pathogenesis of ALVT.

Disclosures

None.

References


Figure 1. ECG showing complete heart block with right bundle-branch block escape morphology. aVF indicates augmented vector foot; aVL, augmented vector left; and aVR, augmented vector right.

Figure 2. Two-dimensional transthoracic echocardiography with color Doppler. Parasternal long-axis view clearly showing the aortic origin of the ALVT (A). On color Doppler, the jet of severe aortic regurgitation (AR) is noted simultaneously with the diastolic flow of blood from the aorta into the ALVT (B). The apical 5-chamber view demonstrates the calcified aneurysmal ALVT burrowing into the proximal interventricular septum; the submitral aneurysm is also noted (C). The apical 2-chamber view shows the submitral and LVOT aneurysms (D). Ao indicates aorta; ALVT, aorto-left ventricular tunnel; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; and TPI, temporary pace-maker implant.
Figure 3. Two-dimensional transthoracic echocardiography with color Doppler. Modified parasternal short-axis view at the basal level showing infundibular PS caused partly because of compression by the aneurysmal ALVT and partly because of RVOT hypertrophy (A). Color Doppler shows turbulent flow across the infundibular PS (B). Basal parasternal short-axis view demonstrates the calcified wall of ALVT (C) with jet seen entering the tunnel from aorta on color Doppler (D). ALVT indicates aorto-left ventricular tunnel; AV, aortic valve; LA, left atrium; PS, pulmonic stenosis; RA, right atrium; and RVOT, right ventricular outflow tract.

Figure 4. Axial imaging on multidetector computed tomography (MDCT) shows the aneurysmal ALVT and LVOT aneurysm (A) as well as the communication between them (B). The aortic and LV connections of the ALVT are clearly delineated in sagittal orientation after multiplanar reconstruction (C and D). ALVT indicates aorto-left ventricular tunnel; An, aneurysm; and LVOT, left ventricular outflow tract.
Figure 5. Multidetector computed tomography (MDCT) clearly shows the submitral and LVOT aneurysms in the coronal plane (A). The infundibular PS, resulting partly from compression by the ALVT and partly from infundibular muscle hypertrophy, is clearly delineated in the sagittal orientation after multiplanar reconstruction (B). Three-dimensional reconstruction of the MDCT images delineates the anatomic relationship between the 3 aneurysms, with the ALVT seen encircling the anterior aspect of the aortic root (C and D). Ao indicates aorta; ALVT, aorto-left ventricular tunnel; PS, pulmonic stenosis; and TPI, temporary pacemaker implant.

Figure 6. Photographs showing a positive wrist (A) and thumb (B) sign in this patient. However, he did not meet the revised Ghent criteria for Marfan syndrome.
Aorto-Left Ventricular Tunnel in a Multianeurysmal Heart
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