Aorta-to-Left Ventricle Tunnel Associated with Noncompaction Left Ventricle

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We present a case of aorta-to-left ventricle (LV) tunnel and noncompaction LV in a 26-year-old woman. She was referred to our echocardiography laboratory for evaluation of aortic regurgitation by echocardiographic examination, which led to the diagnosis of the congenital abnormality of aorta-to-LV tunnel and noncompaction LV. (J Am Soc Echocardiogr 2006;19: 1073.e1-e5.)

Aorta-to-left ventricle (LV) tunnel (ALVT) and noncompaction LV (NCLV) are congenital heart diseases that represent two distinct anatomic features. ALVT is a rare lesion in which a vascular connection between the aorta and the LV exists.[1] NCLV is an unclassified cardiomyopathy characterized anatomically by deep trabeculations in the ventricular wall, which define recesses communicating with the main ventricular chamber. We present a patient with ALVT and NCLV who presented to our echocardiography laboratory with a diagnosis of moderate to severe aortic regurgitation.

CASE REPORT

A 26-year-old woman was referred to our echocardiography laboratory for evaluation of aortic regurgitation. She had a history of functional class II heart failure and diastolic murmur. She had been followed up for aortic regurgitation for 7 years. She could complete her tasks, but had worsening of her symptoms for several months. She was referred to our center to evaluate the severity of her aortic regurgitation by transesophageal echocardiography (TEE). On physical examination she had a to-and-fro murmur at her left sternal border in auscultation. Her electrocardiogram and chest radiograph results revealed no abnormality.

Transthoracic echocardiography (TTE) showed moderate LV enlargement and an echo-free tunnel-shaped space from ascending aorta to LV in which blood flow traveled forward in systole and backward in diastole. TEE demonstrated an echo-free space (35 × 15 mm) immediately above the right and left coronary sinuses and above the level of the coronary arteries (Figure 1 and Movie 1). Aortic orifice diameter of the tunnel was 5 mm and LV orifice diameter was 7 mm (Figure 2). Aortic valve was tricuspid with trivial regurgitation (Figure 3 and Movie 2). Her ejection fraction was 50% to 55%. Pulmonary artery pressure was normal and LV study showed multiple trabeculations and recess formations that color went through with a myocardial noncompaction to compaction exceeding 1.8/1, suggestive of NCLV (Figures 4 to 6) (Movie 3 and 4).

DISCUSSION

ALVT is a congenital extracardiac channel that connects the ascending aorta above the sinotubular junction to the cavity of LV or (less commonly) right ventricle. It begins in anterior aspect of the ascending aorta above the level of right coronary origin, and bypasses the aortic valve, behind the right ventricular infundibulum and through the anterior upper part of the ventricular septum to enter the LV just inferior to the commissure between the right and left aortic cusps. It differs from a ruptured sinus of Valsalva aneurysm (sinus of Valsalva aneurysm fistula) in having its vascular orifice in the tubular aorta rather than a sinus of aortic valve.[2]

Echocardiography is a well-established diagnostic procedure for showing the anatomy in congenital heart diseases. Cardiac catheterization is required only in cases with inadequate information about coronary artery anatomy. Before surgical intervention for aorta-to-right ventricular tunnel is undertaken, every effort should be made to diagnose the coronary artery anatomy, because failure to do so in the case of aberrant origin of a coronary artery may prevent successful surgical correction.[3] NCLV results from intrauterine arrest in the normal process of myocardial compaction. A ratio of

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0894-7317/$32.00

doi:10.1016/j.echo.2006.03.020

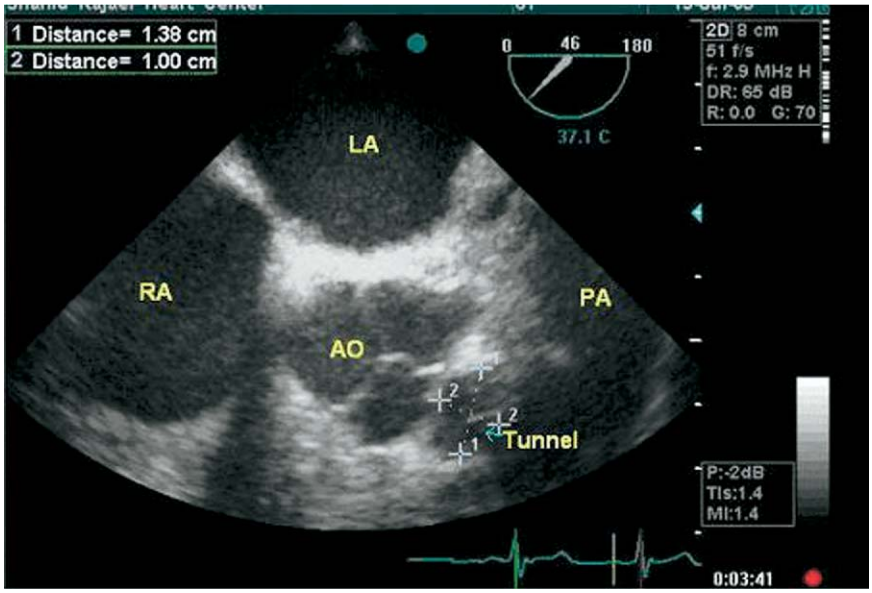

Figure 1 Transesophageal echocardiogram showing echo-free space at commissure between right and left aortic (*AO*) cusps. *LA*, Left atrium; *PA*, pulmonary artery; *RA*, right atrium.

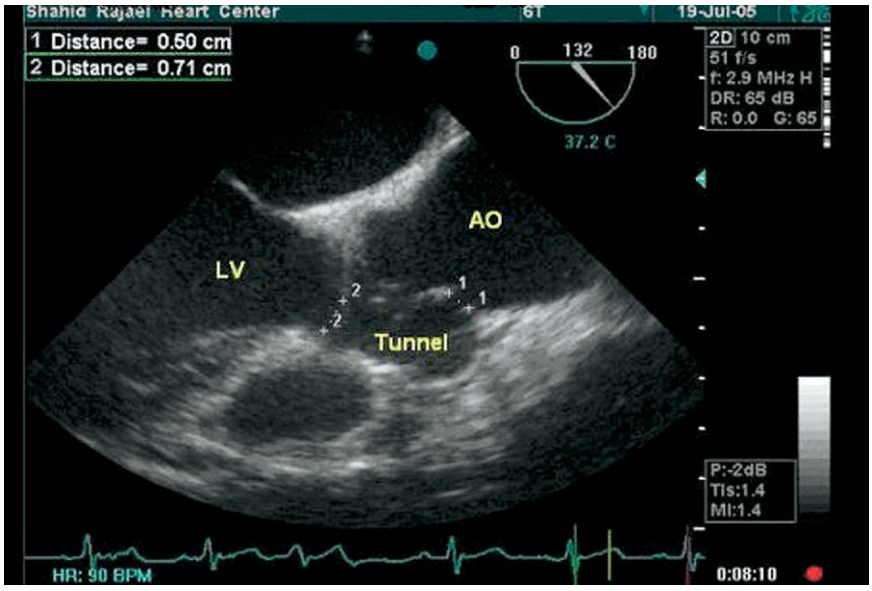

Figure 2 Transesophageal long-axis view showing aortic (*AO*) and left ventricular (*LV*) orifices.

noncompacted to compacted myocardium greater than 2 is diagnostic for noncompaction cardiomyopathy.[4] A significant number of patients have transient recovery of function followed by later deterioration, which may account for many patients presenting as adults, some manifesting an "undulating" phenotype.[5]

We diagnosed the disease with TTE and complete establishment of the details was done by TEE. TTE windows are usually adequate, as in our patient. TEE may provide an alternate approach in patients with poor TTE windows. Angiography is performed to rule out other coronary artery anomalies.

Operation is the state-of-the-art treatment. Because ALVT is associated with congestive heart failure in infancy, surgical intervention is indicated in most cases in early childhood. Early operation is recommended to prevent distortion of the aortic valve, dilatation of the LV, and distortion of the aortic annulus. However, there is a high incidence of

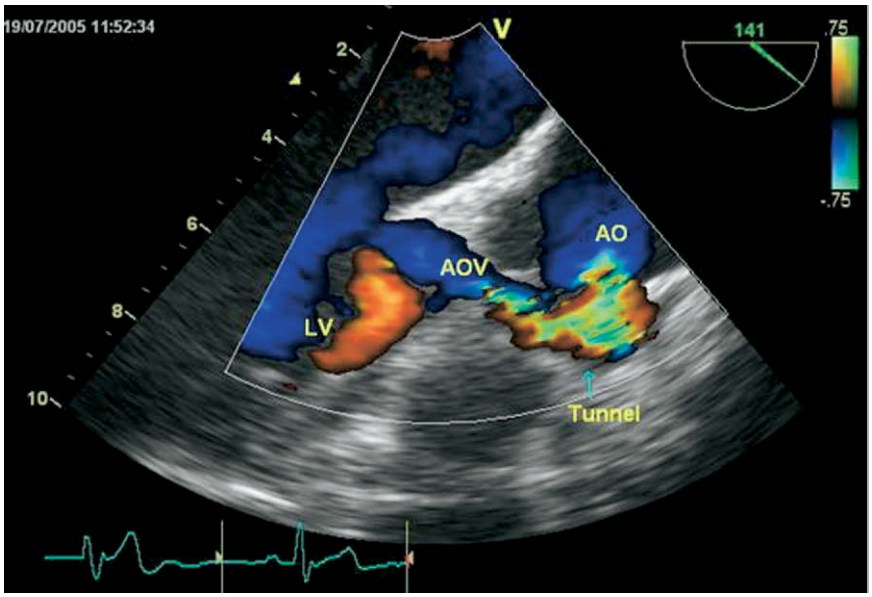

Figure 3 Transesophageal long-axis view showing aorta (*AO*)-to-left ventricular (*LV*) tunnel. Flow goes through it. *AOV*, Aortic valve.

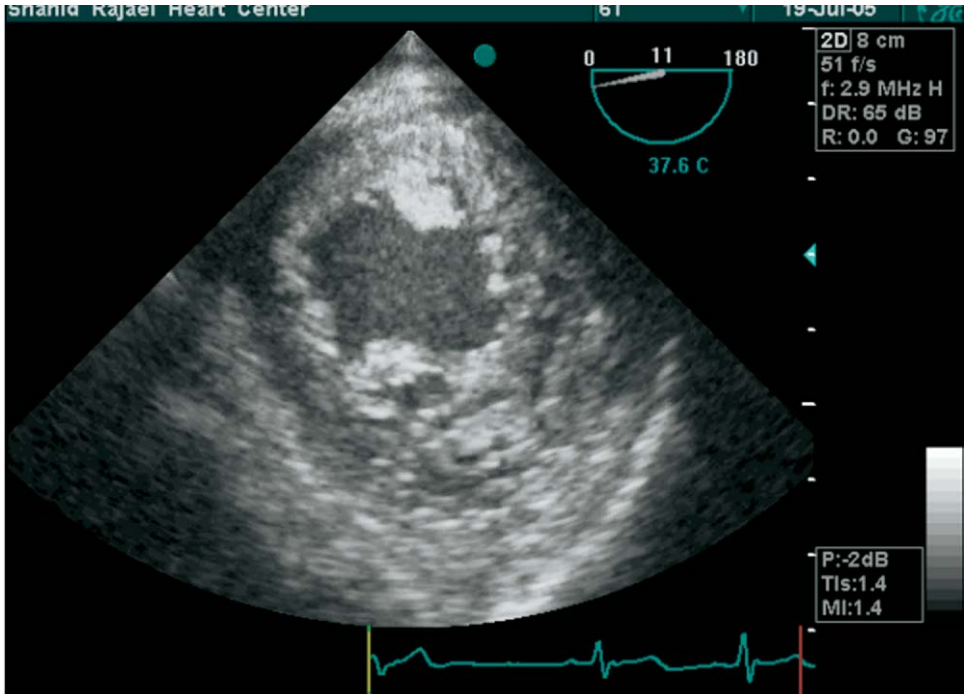

Figure 4 Transgastric short-axis view showing noncompaction left ventricle.

aortic incompetence after operation. After some repairs, there may remain a mild to moderate aortic regurgitation that may result in aortic valve replacement. The operative risk is not low, but results are very encouraging.[6]

ALVT is a very rare congenital anomaly. NCLV or "spongy myocardium" is another rare congenital cardiomyopathy. Vijayalakshmi et al[7] reported the first case in which these two rare anomalies, causing pump failure, coexisted in a 4-year-old boy. The

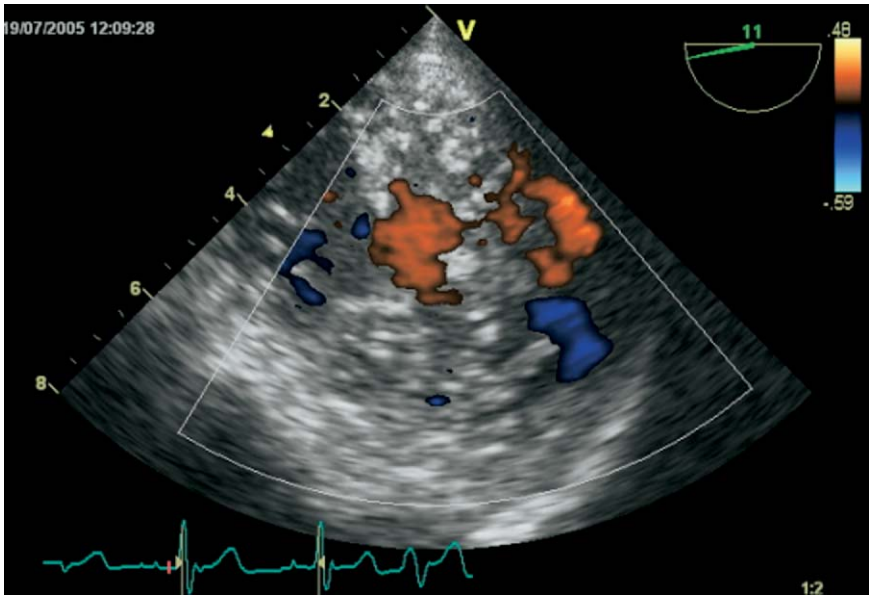

Figure 5 Transgastric short-axis view (2-dimensional and color flow mapping) showing multiple trabeculations suggestive of noncompaction left ventricle.

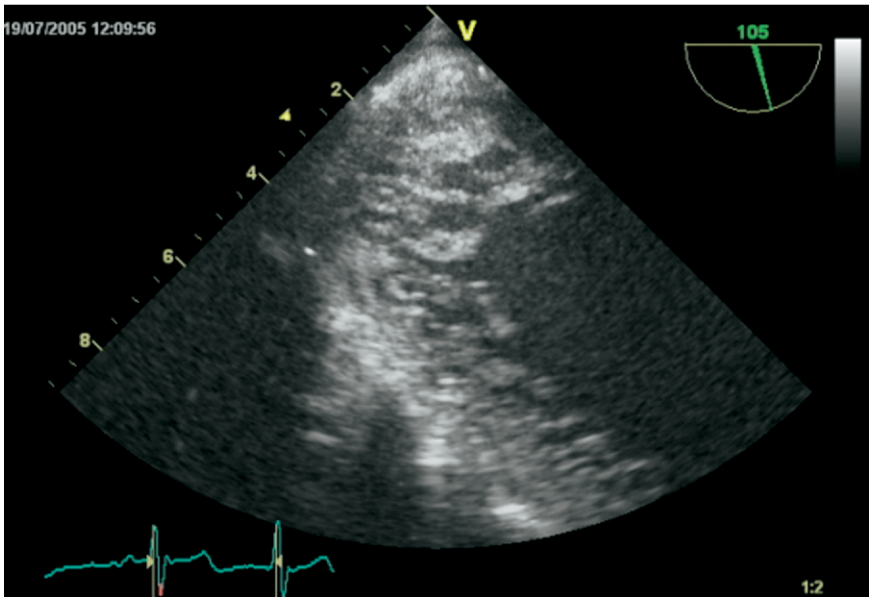

Figure 6 Transgastric long-axis view showing noncompaction left ventricle.

tunnel was successfully treated nonsurgically. Transcatheter closure with the Amplatzer duct occluder was feasible and safe in their case.[8]

Conclusion

ALVT is a very rare congenital anomaly that should be distinguished from other lesions such as aortic regurgitation and sinus of Valsalva fistula. Echocardiography is the diagnostic investigation of choice. ALVT should be treated surgically as soon as possible even in an asymptomatic patient to prevent any damage to the aortic valve and the LV.

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