

Repair of the Aortico-left Ventricular Tunnel Originating from the Right Aortic Sinus with Severe Aortic Valve Regurgitation

Masaki Ando, MD,¹ Tsuguo Igari, MD,² Seiichi Ando, MD,¹ Hirono Satokawa, MD,² and Hitoshi Yokoyama, MD²

A 31-year-old adult with an aortico-left ventricular tunnel (ALVT) arising from the right aortic sinus is reported. Preoperative transesophageal echocardiography demonstrated a ruptured sinus of Valsalva with severe aortic valve regurgitation which originated from the right coronary sinus entering the outlet portion of the left ventricular outflow tract. Operation revealed the aortic entrance of the tunnel was above the right coronary sinus. Direct closure of the orifice of the tunnel using three stitches of 4-0 polypropylene with felt and aortic valve replacement (AVR) was performed. At 10-month follow-up the patient is asymptomatic and receiving no oral medications except anticoagulants. We believe this to be the oldest case of ALVT managed with AVR. (Ann Thorac Cardiovasc Surg 2004; 10: 47–50)

Key words: aortico-left ventricular tunnel, ruptured sinus of Valsalva, aortic regurgitation

Introduction

Aortic regurgitation that results from an abnormal communication, aortico-left ventricular tunnel (ALVT) is a rare, abnormal paravalvular communication between the aorta and the left ventricle.¹⁾ Most commonly the tunnel²⁾ is located above the right coronary sinus³⁾ due to the thin left ventricular anterior wall where the right aortic sinus meets the membranous septum.²⁾ We believe this to be the oldest patient reported in the literature with this condition.

Case

A 31-year-old man with no previous cardiac history was referred to a hospital with epigastric pain. A heart murmur was detected and he was transferred to our hospital. On admission, he had no evidence of heart failure; his

blood pressure was 140/84 mmHg and a 3/6 to-and-fro diastolic murmur was heard. An electrocardiogram showed normal sinus rhythm with elevated ST at V₁₋₄ and inverted T wave at II, III, AV_F leads and evidence of left ventricular hypertrophy (RV_s + SV₁ = 6.2 mV). Chest roentgenogram showed no increased heart size (cardiothoracic rate, 0.50). Transesophageal echocardiography showed a rupture of the right coronary sinus with severe aortic valve regurgitation and demonstrated the course of the tunnel, which originated from the right coronary sinus entering the outlet portion of the left ventricular outflow tract (orifice diameter of the left ventricular side, 6 mm). The aortic valve was tricuspid. Doppler echocardiography suggested the presence of anterograde systolic and retrograde diastolic flow across the tunnel, with severe central aortic valve regurgitation (Fig. 1). Angiographic investigation demonstrated a type 3 ALVT, with an intracardiac aneurysm of the septal portion of the tunnel. A 4/4 degree regurgitation and 55% ejection fraction was evident. Cardiac catheterization demonstrated the pulmonary arterial pressure to be 20/8 mmHg, and the left ventricle end-diastolic pressure 18 mmHg. Cardiac index determined by thermodilution was 3.26 L/min/m².

Surgery was performed under cardiopulmonary bypass with 30°C hypothermia and left ventricular venting

From ¹Department of Cardiovascular Surgery, Fukushima Red Cross Hospital, Fukushima, and ²Department of Cardiovascular Surgery, Fukushima Medical University, Fukushima, Japan

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Address reprint requests to Masaki Ando, MD: Department of Cardiovascular Surgery, Fukushima Red Cross Hospital, 11-31 Iriechou, Fukushima-city, Fukushima 960-8117, Japan.



Fig. 1. Long-axis transesophageal echocardiographic view demonstrating the course of the ALVT (a) and interrogation by Doppler (b).

through the right upper pulmonary vein. After aortic cross clamping and cardioplegic arrest, the aortic valve was inspected. All cusps were atrophied without signs of previous endocarditis. The aortic entrance of the tunnel was identified below and just to the left side of the right coronary ostium (16×6 mm) (Figs. 2, 3). Closure of the aortic entrance of the tunnel was accomplished using three stitches of 4-0 polypropylene with felt. Then, the aortic valve was replaced with bileaflet mechanical prosthesis (23 mm, CarboMedics®, CarboMedics Inc., Austin, TX). Doppler interrogation by transesophageal echocardiography and aortography after surgical repair demonstrated a complete closure of the ALVT. The post-operative course was uneventful. The patient was discharged home, receiving anticoagulants. The excised aortic valve showed a fibrous thickening and partially myxoid degeneration.

Discussion

ALVT is a rare congenital malformation that usually results in severe aortic regurgitation in infancy and childhood and it was first repaired in 1963.¹⁾ An abnormal communication between the aorta and the left ventricle may be secondary to a rupture of a sinus of Valsalva aneurysm. It has been described that the point of difference is the tunnel originated from or joined the ascending aorta above the sinotubular junction and a normal sinus in ALVT compared with the ruptured sinus of Valsalva.¹⁾ From this viewpoint, our case is ALVT. In the ruptured sinus Val-

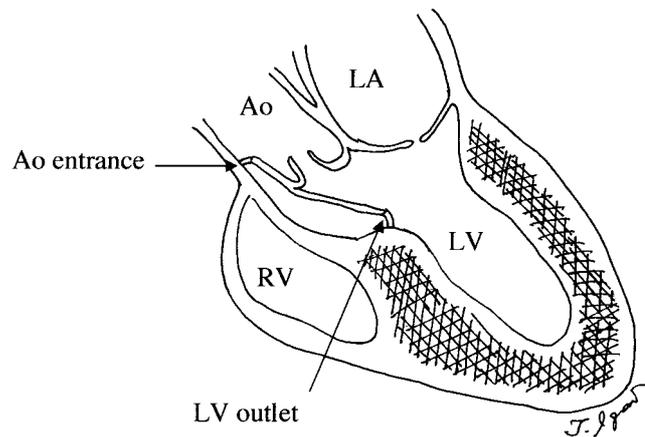


Fig. 2. Schematic sagittal view of the ALVT. Ao, aorta; LA, left atrium; RV, right ventricle; LV, left ventricle.

salva, only three (5.8%) of 52 aneurysms had ruptured from the right sinus into the left ventricle.⁵⁾ Regarding the position of aortic opening of the tunnel, in 14 of the 37 patients it was above the RCA orifice, nine were below as in our case, six were at the same level and six were unknown.⁴⁾

The exact pathogenesis of ALVT is unknown and controversial. The histological sections suggested that the ALVT was an abnormal coronary artery.¹⁾ The origin of one coronary artery from the tunnel supports the theory of ALVT as an example of a coronary fistula.⁶⁾ Persistence of embryonic rests of the fifth arch, intrauterine ruptured aneurysm of sinus Valsalva,⁴⁾ and early aortic dissections as in Marfan syndrome⁷⁾ have been proposed as

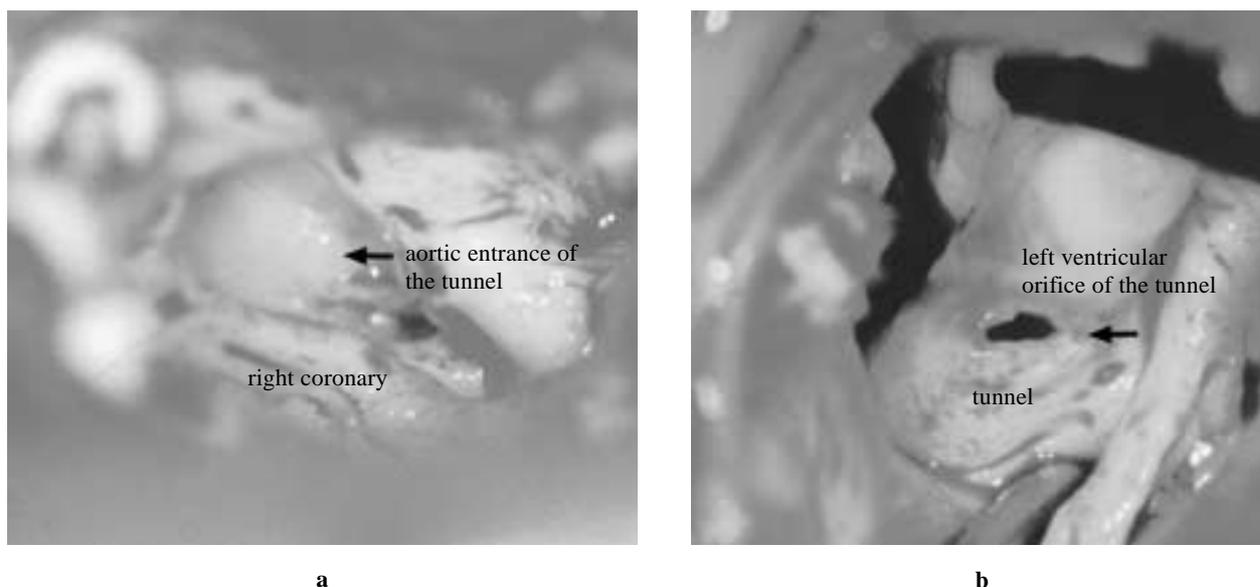


Fig. 3. Operative view of the aortico-left ventricular tunnel from the aortic root. a: Entrance of the tunnel (arrow). b: Orifice of the tunnel at the left ventricular side (arrow).

alternative pathogenic theories of ALVT origin.

The clinical differential diagnosis of ALVT includes aortic valve regurgitation and ruptured aneurysm of sinus Valsalva. In an ALVT the ascending aorta seems to be dilated compared with aortic regurgitation. The aortic dilatation in ALVT is quite similar to Marfan syndrome.¹⁾ In our case, the dilatation of the aortic root was not so prominent. ALVT was classified into four anatomic subtypes from the review of 34 surgically treated cases reported. The incidence of type 3, of which this case is classified, is 23%.⁴⁾

Clinical presentation frequently occurs early in infancy (60% at birth) because of rapid aortic run-off, leading to cardiomegaly and congestive heart failure. Our case is very rare, he having reached age 31, without symptoms. The early diagnosis of this anomaly could be facilitated by two-dimensional and Doppler echocardiography^{8,9)} and transesophageal echocardiography. However, diagnosis should be confirmed by cardiac catheterization and angiography, associated cardiac anomalies should be detected prior to surgery.¹⁰⁾

In our case, the incidental finding of a heart murmur during hospital admission for epigastric pain in an otherwise asymptomatic man is a quite unusual presentation for this rare lesion. The echocardiography finding of an aortic opening of the tunnel in the right aortic sinus represents a very usual anatomic finding for ALVT.¹¹⁾ In our case, long-term obstruction of the right ventricular out-

flow tract by an aneurysmal tunnel may cause degeneration of the aortic valve. The inherent weakness at the junction between the membranous portion of the ventricular septum and the aortic annulus is suggested as the most important factor leading to aortic valve regurgitation in ALVT.²⁾

The ALVT leads to heart failure. No long-term survival has been reported with medical treatment alone; therefore, surgical management is the only therapeutic option.^{4,11,12)} Surgical correction reduces left ventricular volume overload and the progressive annular dilatation.¹¹⁾ A transaortic approach was employed, with or without control at the cardiac end of the tunnel. Transaortic determination the proximity of the tunnel to the aortic valve is superior to distal control through the cardiac end of the tunnel. Surgical correction in our case included direct closure of the aortic opening of the tunnel. This choice, although controversial, was selected because the aneurysm of the tunnel was intracardiac. The main problem after surgery is aortic regurgitation,³⁾ can develop late after surgery. In our 31-year-old case valve replacement was carried out. We believe that direct closure of the aortic end of the tunnel may be tolerated and would be preserved.

Conclusion

We have reported the oldest patient in the literature to have an ALVT, originating above the right aortic sinus,

with severe aortic regurgitation, having successful surgical treatment.

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