Late Outcome After Repair of Aortico-Left Ventricular Tunnel
— 10-Year Follow-up —

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Despite successful surgical repair, patients with congenital aortico-left ventricular tunnel (ALVT) are at risk of developing aortic incompetence in the late postoperative period. Two cases of ALVT were followed for 10 years with special reference to aortic incompetence and geometry of the aortic root. The patients underwent repair of ALVT, one at 4 years of age and the other at 4 months of age. The first patient had a slit-like tunnel (type I) and the aortic orifice was closed with a pericardial patch. The second patient had a large tunnel with an extracardiac aneurysm (type II) and was closed with a pericardial patch at the aortic orifice and a Dacron patch at the left ventricular orifice, thereby completely obliterating the tunnel. The last echocardiographic evaluation showed no residual flow in the tunnel and no aortic incompetence in case 1, but there was mild aortic valvular regurgitation with deformity of the right sinus in case 2. Careful long-term follow-up is necessary because patients with ALVT have some inherent structural abnormalities from the left ventricular outflow tract to the aortic root. (Circ J 2006; 70: 939–941)

Key Words: Aortico-left ventricular tunnel; Aortic root; Congenital heart disease

A ortico-left ventricular tunnel (ALVT), an abnormal communication between the aortic root and left ventricle, is a rare congenital malformation1,2 and despite successful surgical repair, the patients with ALVT are at risk of developing aortic incompetence (AI) even in the late postoperative period3–6. We report 2 cases of ALVT that have been followed for 10 years with a special reference to AI and geometry of the aortic root.

Case Reports

Case 1
A 4-year-old girl with ALVT was referred for surgery. A heart murmur was noted at 10 months of age, and cardiac catheterization revealed the presence of the ALVT. She had been previously asymptomatic, but echocardiography at 4 years of age revealed left ventricular dilatation with the ALVT arising from the right aortic sinus entering the left ventricular outflow tract (LVOT) with trivial AI. A chest roentgenogram showed no cardiomegaly. Aortography showed a simple fistulous channel (type I; Fig 1a). Extracardiac aneurysm, or deformity of the aortic sinuses was not identified.

At surgery, inspection showed no distortion of the aortic cusps. The aortic orifice of the tunnel was 5 mm in diameter, just above the commissure between the non-coronary and right coronary sinuses. The aortic orifice was far from the orifice of the right coronary artery, and there were no abnormalities of the coronary orifices. The aortic orifice was closed with a pericardial patch (Fig 1b). The left ventricular orifice was not detected, and was left open. Postoperative echocardiography demonstrated a trivial residual ALVT and trivial AI. Echocardiography performed 10 years after surgery showed no residual ALVT or AI. Neither deformities in the aortic sinuses nor dilatation of the annulus was identified (Table 1). The patient had been asymptomatic and received no medication.

Case 2
A 1-month-old boy was referred for cardiac evaluation. Cardiomegaly had been detected on chest roentgenogram at 1 week of age, and echocardiography revealed the ALVT arising from the right aortic sinus with an extracardiac aneurysm (type II).6 Cardiac catheterization revealed the ALVT with an extracardiac aneurysm of approximately 40 mm in diameter (Fig 1c). The orifice of the tunnel was 18 mm in diameter at the aortic end and 7 mm in diameter at the left ventricular end. There was massive regurgitation through the tunnel and moderate AI. There was no evidence of left or right ventricular outflow tract obstruction by the aneurysm. The ascending aorta and the right aortic sinuses were dilated, and there was marked dilatation of the left ventricle.

Surgery was performed at 4 months of age. The ascending aorta was transected and the tunnel was opened vertically. There was neither prolapse nor distortion of the aortic valve. The aortic orifice of the tunnel was in the right coronary sinus, approximately 5 mm away from the orifice of the right coronary artery, and there were no abnormalities of the coronary orifices. The tunnel was closed with a pericardial patch at the aortic orifice and with a Dacron
patch at the left ventricular orifice, and 2 additional stitches were placed to completely obliterate the tunnel (Fig 1d). Postoperative echocardiography showed no residual ALVT, but revealed mild AI. Echocardiography performed 10 years after surgery showed mild AI, mild left ventricular dilatation with an end-diastolic diameter of 118% of the normal value, and deformity of the right aortic sinus (Table 1). AI had not developed during the 10-year follow-up period, but the aortic annulus remained dilated at 140% of normal. The patient had remained asymptomatic and had not received medication.

### Discussion

The incidence of AI in patients with ALVT following surgery ranges from 16% to 60%, and the requirement for aortic valve replacement ranges from 0% to 50%.3–8 In our series of cases, there was no further exacerbation of AI during the follow-up period of 10 years, although case 2 still had mild AI at late follow-up, presumably because of the dilated aortic annulus and deformed right aortic sinus. The potential causes of late AI include coexistent aortic valve disease, dilatation of the aortic root and sinuses, and distortion of the aortic cusps at surgery.4–6 Tuna et al suggested that deviation of the unsupported segment of the aortic annulus leads to inappropriate coaptation and subsequent AI.2 Therefore, Horvath et al have suggested that 2 patches be used to close both orifices of the ALVT and that the tunnel be obliterated with additional stitches in order to support the aortic annulus.5 Because case 2 had a large ALVT with an extracardiac aneurysm, it is possible that severe deviation of the unsupported segment of the aortic annulus resulted in residual AI. Patients with ALVT have some inherent structural abnormalities from the LVOT to the aortic root,2 necessitating careful follow-up to assess for the development of AI and changes in geometry of the aortic root and annulus, irrespective of lesion type or surgery. In addition, aortic root remodeling combined with aortic valve repair would be a useful option in patients with ALVT who present late AI following surgery if the AI predominantly results from aortic root deformity.

The surgical approach for ALVT consists of suture or patch closure of the aortic and ventricular orifices, and obliteration of the tunnel. Some studies, using different surgical techniques, have shown comparable long-term outcomes, but it is still controversial whether or not direct closure of the aortic orifice causes distortion of the aortic cusps.4–6 Meldrum-Hanna et al, who used direct closure of the aortic orifice, showed that 3 of 6 patients (50%) required aortic valve replacement after a mean period of 10 years following repair.4–6 In contrast, in a recent series using patch closure, there were excellent early and late outcomes.7–8 Martins et al, who used direct closure in 7 of 9 patients, had no cases of aortic valve replacement during a median follow-up period of 5 years, but 5 of 10 (50%) patients had mild AI at follow-up.6 We prefer to use patch closure to minimize the adverse effects on the aortic cusps, irrespective of the size of the orifice, especially when repairing ALVT in small infants. In addition, early surgery at the time of diagnosis is recommended because tunnel-related turbulence and/or progressive dilatation of aortic root and the annulus may affect long-term aortic valve function.
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References