Aortico–Left Ventricular Tunnel Arising From the Noncoronary Sinus Associated With a Ventricular Septal Defect

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Aortico-left ventricular tunnel is a very rare congenital heart disease. The aortic orifice of the tunnel usually situates in the right or left coronary sinus. Here we present the case of one child who had an unusual tunnel arising from the noncoronary sinus in addition to an unrestrictive perimembranous ventricular septal defect. Surgical repair was performed at 2 months of age and there was no postoperative residual lesion.


Aortico-left ventricular tunnel (ALVT) has occasionally been reported in the last 50 years. Most of ALVT patients have the aortic orifice in the right or left coronary sinus [1, 2]. Here we present a child who has an unusual ALVT arising from the noncoronary sinus, as well as a ventricular septal defect.

We report the case of a 2-month-old boy who had prenatal diagnosis of ventricular septal defect. He was asymptomatic in the first month but started having signs of congestive heart failure after that. The follow-up transthoracic echocardiography incidentally found a small ALVT, as well as an expected ventricular septal defect. The ALVT was around 2-mm wide and 8-mm long. The aortic orifice was in the noncoronary sinus and it was close to the left noncoronary commissure. The left ventricular orifice was around the right fibrous trigone area. There was diastolic flow regurgitation through the ALVT, with the jet flowing toward the anterior leaflet of the mitral valve (Figs 1A, 1B). The perimembranous ventricular septal defect was unrestrictive, mainly shunting left to right (Fig 1C). There was no aortic stenosis or insufficiency and the functions of the other valves were good. The ventricular function was normal. On physical examination, there was a 2/6 grade systolic murmur and a 3/6 grade diastolic murmur in the precordium area with a loud P2.

The patient underwent ALVT repair and ventricular septal defect closure. During the operation, a standard aortotomy was performed. The aortic orifice of the ALVT was confirmed at the level of the sinotubular junction in the noncoronary sinus, and it was near the left noncoronary commissure. A probe was put through the ALVT and it ended in the left ventricular outflow tract.
The ventricular orifice of the ALVT was in the interleaflet triangle between the left and noncoronary cusps (Fig 2A). The aortic orifice was closed by 2 interrupted pledgeted sutures that were tied outside of the aortic wall (Fig 2B). The ventricular orifice had reasonable distance away from the margin of the perimembranous ventricular septal defect (Fig 2C), and it was closed by a continuous Prolene stitch. The ventricular septal defect was closed with a patch through the right atrium. The intraoperative and follow-up echocardiography at 1-year post operation demonstrated no residual ALVT, no residual ventricular septal defect, and no aortic insufficiency.

Comment

Aortico–left ventricular tunnel is an extracardiac channel between ascending aorta above the sinotubular junction and left ventricle. It represents 0.1% to 0.5% of congenital heart disease, and two thirds of ALVT patients are male [1]. The ALVT should be distinguished from the sinus of Valsalva aneurysm rupture. In ALVT patients the aortic orifice is located at the level of the sinotubular junction instead of the Valsalva sinus. The ALVT also needs to be differentiated from aortic insufficiency, especially those eccentric regurgitations.

As reported in previous cases, most ALVTs are located in the right or left coronary sinus. The associated diseases consist of aortic stenosis, aortic insufficiency, left ventricular outflow obstruction, and pulmonary stenosis [1, 3]. Our patient did not have a similar anatomy to those presented in previous literature. He had a combination of an unusual noncoronary sinus originating ALVT with a ventricular septal defect. The coronary anatomy in most ALVT patients is normal; however, 1 of the coronary arteries may originate from the ALVT, which makes surgery more complicated [4].

Most ALVT patients are diagnosed by either trans-thoracic or transesophageal echocardiography [5, 6]. Prenatal diagnosis has been reported in recent years [5]. Unfortunately, the prenatal ALVT diagnosis was missed in this case. Magnetic resonance imaging and angiography are supplemental diagnostic modalities.

Almost all the reported ALVT patients had surgical repair, except 1 patient who was conservatively observed after an aortic balloon valvuloplasty, and very few patients had device closure [1, 6–8]. The most popular surgical repair is double patch closure of both the aortic and the ventricular orifices. Direct suture closure of the ALVT has also been reported [1]. In our patient, because of the small size of the tunnel and the orifices, the ALVT was closed with interrupted pledgeted sutures. Postoperative echocardiography showed a good short-term result without residual shunt and aortic insufficiency but

Fig 2. Intraoperative images. (A) The probe was put through the aortico–left ventricular tunnel (ALVT, the star marker), which is in the noncoronary sinus and close to the left noncoronary commissure. (B) Pledged stitch was put to close the aortic orifice of the ALVT (arrow), and was tied outside the aortic wall (the triangle symbol). (C) A diagram illustrates the relationship between the ventricular septal defect (VSD) and the ALVT. (LCC = left coronary cusp; NCC = noncoronary cusp; RCC = right coronary cusp; RFT = right fibrous trigone.)
long-term follow-up is needed for observation of ALVT recurrence and aortic valve function. According to the reported cases, the operative risk is low and the surgical outcome is good.

In summary, we reported a surgical case of ALVT arising from the noncoronary sinus associated with a ventricular septal defect. Echocardiography is an effective diagnostic modality. Surgical repair is the main option of treatment and it achieves a good outcome with low operative risk.

References