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Brief Report

Successful repair of aorto-left ventricular tunnel diagnosed prenatally

Jacek Kolcz, Katarzyna Januszewska, Edward Malec

Department of Pediatric Cardiac Surgery, Polish-American Children's Hospital, Collegium Medicum, Jagiellonian University, Kraków, Poland

Abstract Aorto-left ventricular tunnels are anomalous communications between the ascending aorta and the left ventricle. Prenatal diagnosis, followed by successful surgical management, is rare. We describe a fetus diagnosed with aorto-left ventricular tunnel at 35 weeks who achieved a favourable outcome after immediate postnatal surgical repair.

Keywords: Fetal cardiology; aortic root; aortic insufficiency

AORTO-LEFT VENTRICULAR TUNNEL IS AN extremely rare defect, first described by Levy et al.[1] in 1963. It consists of an anomalous, endothelialized communication that extends between the aorta and the left ventricle. The time of diagnosis, symptoms, and signs depend on the size of the tunnel and the severity of aortic regurgitation. Detection of the defect in a routine prenatal ultrasound examination is usually associated with the presence of an evident communication, large enough to produce advanced left ventricular failure. Prenatal diagnosis is rare, and only a few newborns detected antenatally have survived.[2] After delivery, the patients usually present with symptoms of congestive heart failure. Regardless of the time of detection, the only treatment is surgical, the repair preventing the development of heart failure and correcting the aortic insufficiency.[3] In severe forms diagnosed in the neonatal period, surgery is associated with good results if performed soon after the diagnosis. We present a fetus diagnosed with aorto-left ventricular tunnel who underwent an immediate successful repair.

Correspondence to: Professor Edward Malec MD, PhD, Department of Pediatric Cardiac Surgery, Polish-American Children's Hospital, Collegium Medicum, Jagiellonian University, 265 Wielicka St., 30-663 Kraków, Poland. Tel: +48 12 658 10 23/13 25; Fax: +48 12 658 10 23; E-mail: mimalec@cyf-kr.edu.pl

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Case report

A male newborn weighing 3850 grams with aorto-left ventricular tunnel was admitted to our department 26 hours after delivery. The defect had been detected by fetal echocardiography in a 32-year-old woman at 35 weeks of gestation. A routine obstetric ultrasonic study at 27 weeks of gestation had revealed marked polyhydramnios combined with unusual cardiomegaly. Previous routine ultrasonic scans at 20 weeks and 24 weeks of gestation had been assessed as normal. A detailed ultrasonic examination then revealed dilation of the aortic root, along with left ventricular enlargement and severe hypertrophy. Doppler studies showed marked aortic regurgitation. This picture was produced by a large channel passing from the ascending aorta to the left ventricle that bypassed the leaflets of the aortic valve (Fig. 1). The aortic orifice of the tunnel was above the right sinus of Valsalva. Retrograde flow trough the tunnel was identified by colour Doppler scans. The parents of the child were informed about the type of malformation and the detailed plan for postnatal management was outlined with their participation.

The male newborn was spontaneously delivered at 40 weeks of gestation, the delivery proving uncomplicated. On admission, he presented with slight oedema, respiratory distress, and tachycardia. The praecordial area was hyperactive, and loud systolic and diastolic murmurs were heard at the left sternal border. The pulse pressure was wide, with the average diastolic

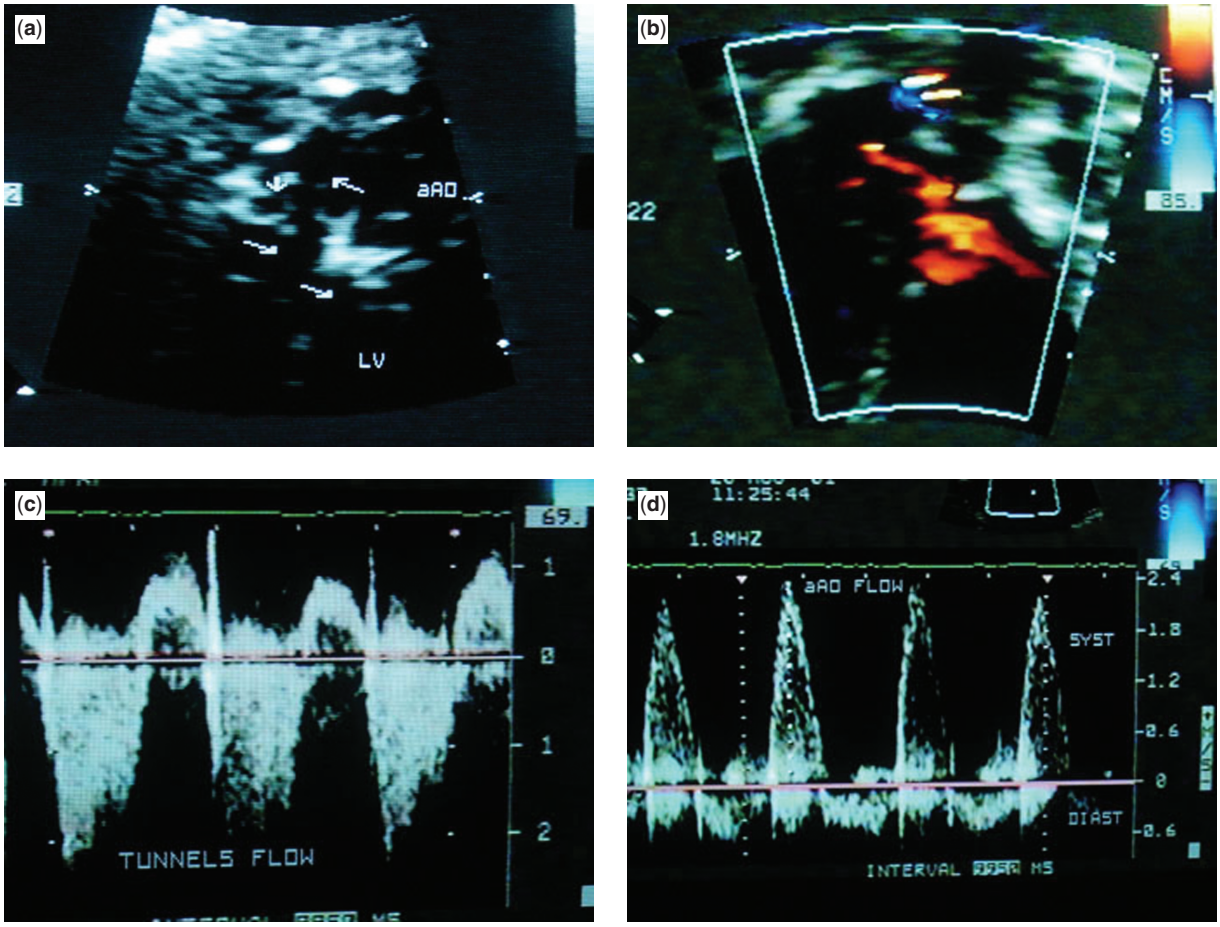

Figure 1.
*The prenatal echocardiogram reveals (**a**) a tunnel extending from the left ventricle (LV) to the ascending aorta (aAO) tunnel, with colour Doppler demonstrating flow through the tunnel in antegrade (**b**) and retrograde (**c**) directions, the retrograde flow (**d**) also being seen in the ascending aorta.*

pressure at 25 millimetres of mercury, and with a range from 18 to 28 millimetres of mercury. Left ventricular enlargement, and dilation of the ascending aorta, was visible on the chest radiograph. Cross-sectional echocardiography confirmed the prenatal diagnosis of an aorto-left ventricular tunnel, showing the channel to have a diameter of 7 to 8 millimetres, with central aneurysmal dilation. The left ventricle was dilated, hypertrophied, and hypokinetic. The colour Doppler study showed antegrade systolic flow and massive diastolic regurgitant flow trough the tunnel.

The neonate underwent surgery 28 hours after delivery. The operation was performed using cardiopulmonary bypass with a short period of deep hypothermic circulatory arrest.

The tunnel was situated on the anterior wall of the ascending aorta. It was a thin-walled canal, with a lumen of 7 millimetres and aneurysmal dilation in its central part (Fig. 2). After cross-clamping of the aorta and temporary clenching of the tunnel, cold crystalloid cardioplegia was infused and the aorta was opened transversely. The aortic valvar leaflets, although thickened, were competent. The aortic orifice was confirmed as originating above the right sinus of Valsalva, and the tunnel was traced to the left ventricle just below the aortic valve. The aortic origin was closed with a Gore-Tex patch using a continuous suture of 6.0 Prolene. The tunnel itself was then opened, and the ventricular orifice was closed with another Gore-Tex patch using a continuous suture, additionally suturing the tunnel with continuous 5.0 Prolene. There was no evidence of aortic insufficiency noted after the procedure. The mean diastolic blood pressure within the first 48 hours was 51 millimetres of mercury, with a range from 45 to 56. The echocardiogram showed slightly improved left ventricular function, albeit that hypertrophy of the left ventricle persisted. The patient was weaned from respiratory support on the third postoperative day. He was discharged from hospital after 24 days. Left ventricular dimensions and function were then normal on echocardiography. Follow-up after 24 months revealed normal development, echocardiography with colour Doppler confirming closure of the

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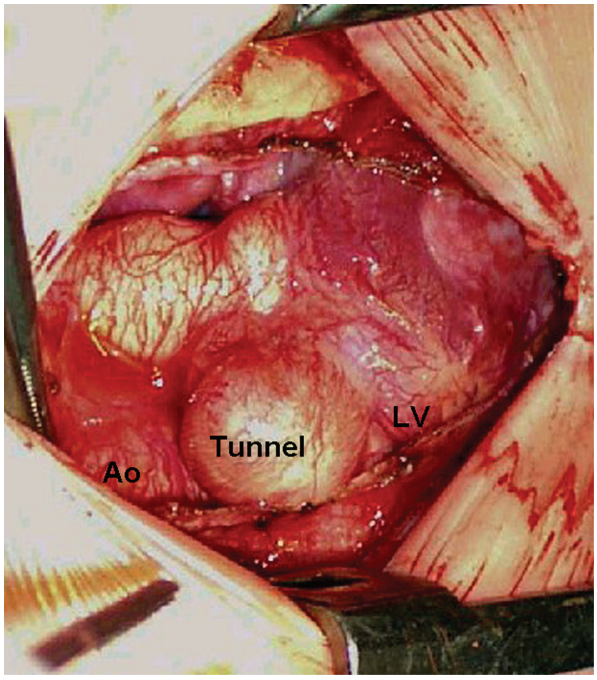

Figure 2.
The surgical view shows the tunnel running between the left ventricle (LV) and the aorta (Ao).

tunnel, with no regurgitation nor obstruction of the aortic valve. The left ventricle was slightly hypertrophied and the ascending aorta insignificantly dilated.

Discussion

The aorto-left ventricular tunnel presenting at birth, and requiring surgery in infancy, was first reported by Levy et al.[1] As with many other congenital cardiac malformations, the lesion can now be detected prenatally, albeit that antenatal diagnosis usually accompanies severe aortic regurgitation, whereas diagnosis after delivery is more likely to indicate a milder form of the defect.[2,4,5] The earliest prenatal detection of which we are aware has been reported at 19 weeks of gestation, subsequent to observation of a dilated aortic root and left ventricle.[4]

Prenatal diagnosis of the cardiac defect can be clinically important because of the potential postnatal implications. It offers the possibility of special care for the fetus and its parents, optimal delivery, and immediate postnatal care. It results in avoidance of haemodynamic compromise, reduced systemic dysfunction, and reduced surgical delay. It can also prevent discharge to home without diagnosis and subsequent sudden death. Early identification of a defect allows for planned delivery at a site where services for neonatology, paediatric cardiology, and congenital cardiac surgery are readily available. It is associated with the possibility of preparing an appropriate strategy with the participation of the family.[6] The prenatal diagnosis of a severe example of an aorto-left ventricular tunnel in our patient allowed for all these preparations.

Only a small number of neonates, to the best of our knowledge, have survived surgical correction following correct antenatal diagnosis of the malformation. Since the lesion is usually severe, such fetuses are usually characterized by generalized oedema, severe left ventricular dysfunction, and aortic incompetence.[2] In our patient, the most important observation when making the diagnosis was the presence, on colour Doppler echocardiography, of marked aortic regurgitation, enlargement and hypertrophy of the left ventricle, and dilation of the aortic root.

Accurate postnatal diagnosis can be also problematic, be it based on angiography or echocardiography.[7,8] This is possibly because the more common milder variants produce a less intensified clinical picture. Non-invasive diagnosis with cross-sectional and colour flow Doppler echocardiography after delivery is now preferred, especially in neonates in critical condition, since invasive diagnostic procedures such as cardiac catheterization and angiography may lead to deterioration of the neonate and increase the subsequent risk of treatment.

In the majority of cases, the tunnel originates above the right sinus of Valsalva and the orifice of the right coronary artery, sometimes with the artery originating from the tunnel. Sporadically, it may originate above the origin of the left coronary artery.[9] Associated cardiac defects include a bicuspid aortic valve, aortic stenosis or regurgitation, patency of the arterial duct, ventricular septal defect, pulmonary stenosis, and infundibular right ventricular obstruction.[4] In our patient, the aortic origin of the tunnel was typically located above the right sinus of Valsalva, close to the zone of apposition between the right and non-coronary aortic valvar leaflets. Both the right sinus of Valsalva and the right coronary artery remained normal, albeit that the tunnel was dilated aneurysmally in its central part. There were no other intracardiac or extracardiac anomalies.

There is no survival without surgery, but early operative correction can prevent the negative results of aortic incompetence and congestive heart failure, and avoid sudden death.[2,10] There are different techniques of surgical repair. Some authors advocate direct closure of the aortic opening using mattress sutures alone.[1] In case of larger defects, it is usual to close the aortic end with a patch.[7] In our patient, we closed both ends of the tunnel with Gore-Tex patches, additionally placating the tunnel itself with a continuous suture. This technique avoids distorting either the aortic valve or the orifice of the coronary

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artery, and prevents turbulent flow within the blind recess created by the surgical repair. Aortic regurgitation is reported in up to three quarters of patients after repair,[10] and may necessitate reoperation on the aortic valve. In the follow up of our patient, however, the function of the aortic valve was good 24 months after surgery. The patient will continue to be followed-up carefully to detect any potential aortic insufficiency.

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