

# An aortico-left ventricular tunnel with aortic stenosis: Diagnosis based on echocardiography in a neonate

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## Abstract

*A very rare case of left-ventricular tunnel is reported. Diagnosis was based on ECHO examination. The differential diagnosis and surgical treatment of aortico-left ventricular tunnel is discussed. (Cardiol J 2007; 14: 193–197)*

**Key words:** aortico-left ventricular tunnel, echocardiography

## Introduction

An aortico-left ventricular tunnel is an extremely rare cardiac malformation characterised by a paravalvular communication between the two structures. This lesion represents about 0.1% of congenital cardiac malformations [1, 2].

The aortic orifice of the tunnel is usually located in the anterior wall of the aorta, above the right sinus of Valsalva (and the right coronary artery ostium). Occasionally it can be found above the left coronary artery ostium. The ventricle orifice of the tunnel opens to the left ventricle (LV) just below the right and left aortic cusps.

This malformation was first described by Levy in 1963. Since then fewer than 100 cases of this

malformation have been reported in the literature [3–5] and its diagnosis is not easy.

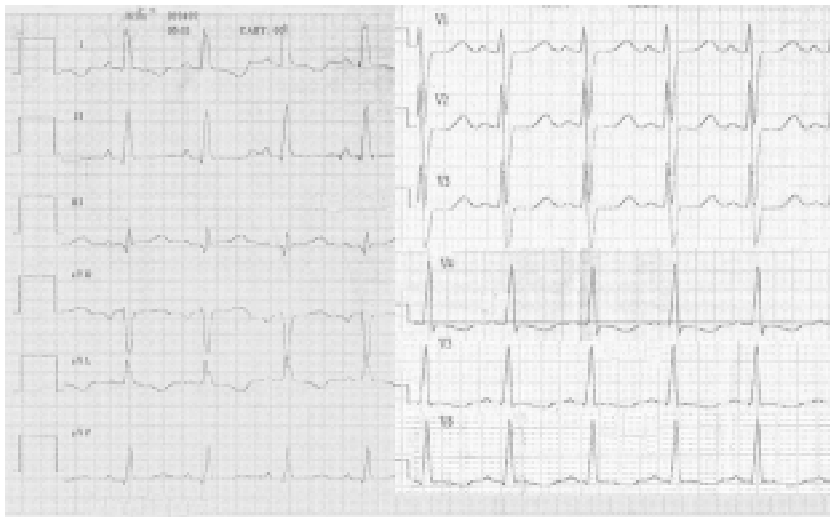
We report a case of aortico-left ventricular tunnel diagnosed in a neonate with the use of echocardiography (ECHO) only. On the basis of this diagnosis the baby underwent successful surgical repair.

## Case report

A female neonate was admitted to our Clinic two hours after birth. The child, from a second pregnancy and second delivery by Caesarean section, was born at 37 weeks of gestation with a birth weight of 3020 g and 9 points in the Apgar scale. Prenatal diagnosis of severe aortic valve stenosis was made in the 36<sup>th</sup> week of gestation and confirmed by ECHO at birth. As a result of this diagnosis the baby received Alprostadil in a dose of 0.02 µg/kg/min, Dopamine in a dose of 2 µg/kg/min and Furosemide in a dose of 1 mg/kg/day after the birth.

On admission the general condition of the neonate was moderately severe. Tachypnoea of 70–80/min was noticed and a grade 3/6 ejection systolic murmur and a low diastolic murmur were heard over the precordium. An ECG showed

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**Figure 1.** ECG on admission.

a normal axis (with no physiological right axis deviation), a normal sinus rhythm rate of 160/min but left ventricular hypertrophy and overload (Fig. 1).

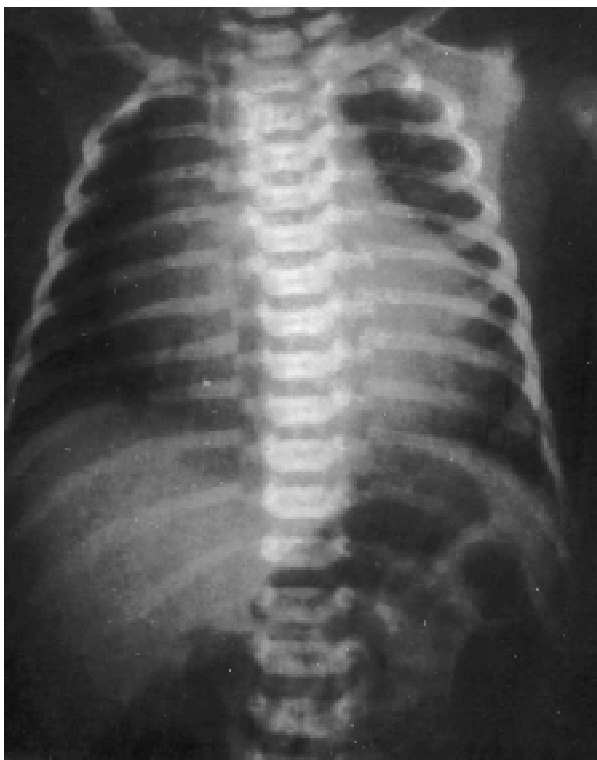
An X-ray showed that the heart size was distinctly increased, while the pulmonary vasculature was normal. There were no focal condensations in the lung (Fig. 2).

The ECHO examination revealed significant left ventricular chamber enlargement with a thick-

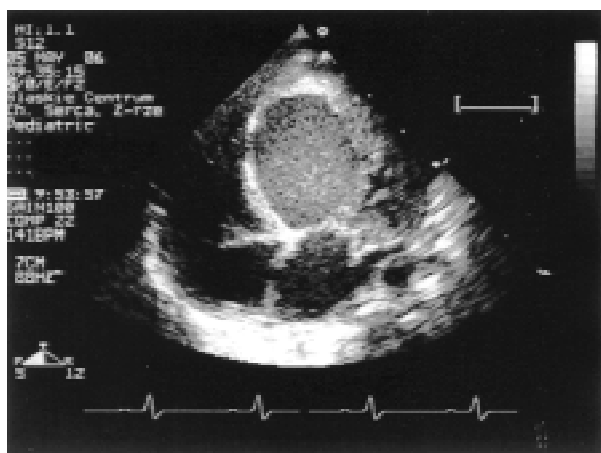
ened wall and decreased contractility (FS 18%) (Fig. 3).

In the parasternal long-axis view the presence of aneurismal dilatation above the aortic valve annulus was noticed.

Colour Doppler echocardiography demonstrated a significant regurgitant flow leading to the LV, although this flow was located above the aortic annulus. Regurgitant flow came from the canal, which opened to the LV chamber just below the aortic valve. This canal (in the central part aneurysmatically dilatated to 7–8 mm) opened to the aorta above the right sinus of Valsalva and above the right coronary artery ostium (Figs. 4, 5). The aortic valve annulus was narrow, measuring about 5 mm in diameter; the aortic valve was thickened and narrowed, with altered leaflets but without regurgitant flow.



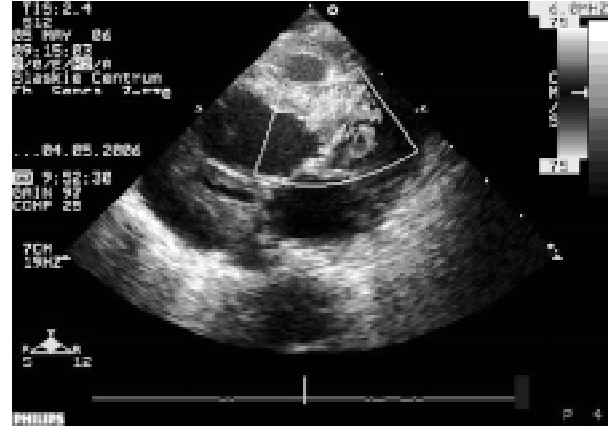
**Figure 2.** Chest X ray on admission.



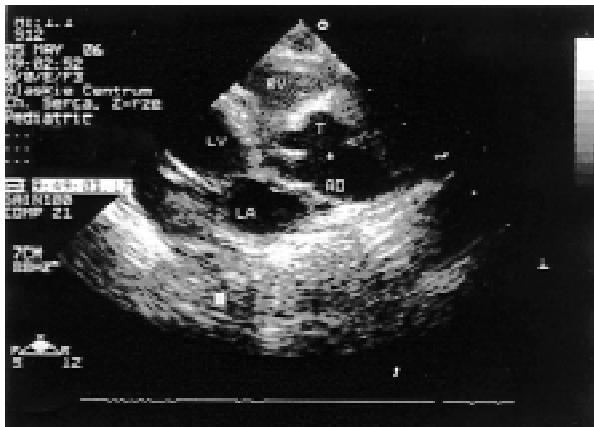
**Figure 3.** ECHO examination, apical four-chamber view.



**Figure 4.** ECHO examination, parasternal long-axis view. LV — left ventricle, LA — left atrium, RV — right ventricle, T — tunnel, Ao — aorta.



**Figure 6.** ECHO examination, colour Doppler, parasternal long-axis view. Two outflow tracts of blood from the left ventricle are visualised.



**Figure 5.** ECHO examination, modified parasternal long-axis view. Junction of tunnel (T) to aorta (Ao) — arrow; LV — left ventricle, LA — left atrium, RV — right ventricle.

The maximum systolic pressure gradient through the aortic valve was 40–50 mm Hg. In the colour Doppler echocardiography examination two separate “outflow tracts” for the exit of blood from the left ventricle were observed: one, in an anterior position, led to the right sinus of Valsalva and the second, in a posterior position, led to the aorta through the aortic valve. The two “outflow tracts” joined above the right coronary artery ostium, where the tunnel opened to the ascending aorta (Fig. 6).

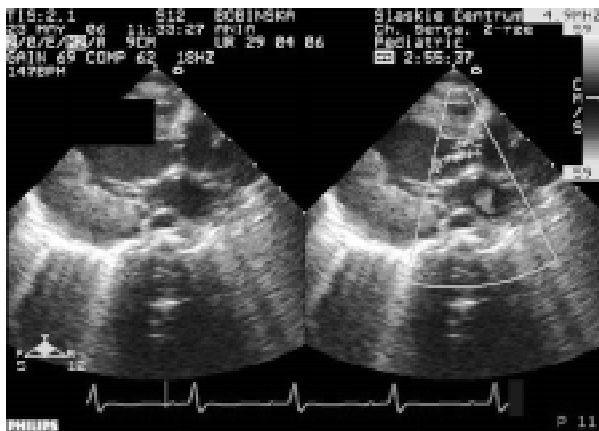
The diagnosis of aortico-left ventricular tunnel with associated aortic stenosis was made by echocardiographic examination. The ECHO images were so convincing that we decided not to perform cardiac catheterisation.

The child was scheduled for surgical correction, which was performed with extracorporeal circulation and hypothermia of 20°C. Patent ductus arteriosus was ligated. The ascending aorta was cross-clamped and cardioplegia was administered. After a transverse incision of the aorta above the sinus of Valsalva the aortic orifice of the tunnel was exposed next to the right coronary artery ostium. The diameter of the aortic orifice of the tunnel was 7 mm × 5 mm and the LV orifice diameter was about 3–4 mm. The ventricle orifice was closed with mattress sutures using a pericardial pad support, whereas the aortic orifice was closed by a pericardial patch stiffened in glutaraldehyde. The aortic valve was found to be tricuspid with a leaflet separation of about 4 mm. All three fused commissures of the aortic valve were incised up to about 0.5–1 mm and an improvement in leaflet separation was achieved, a Hegar 6 being easily passed through.

The course of the operation and the postoperative period were uncomplicated. The postoperative echocardiographic control examination showed aortic insufficiency and tight closure of the tunnel into the LV (Fig. 7). Left ventricular contractility remained decreased, as before (FS 18%, EF 36%), as a result of endocardial fibroelastosis. As a result, in the postoperative period catecholamine (Dopamine) and Milrinone infusion were used for 8 days.

## Discussion

An aortico-left ventricular tunnel may occur as an isolated lesion or in association with other malformations (in 30% cases). In association it is more frequently combined with bicuspid aortic valve and



**Figure 7.** ECHO examination after cardiosurgical operation, colour Doppler, parasternal long-axis view. Visualised trivial aortic regurgitation.

aortic valve stenosis and rarely with ventricular septal defect (VSD), patent ductus arteriosus, pulmonary valve stenosis or coronary artery anomaly [1].

The mechanism of aortico-left ventricular tunnel formation has not entirely been explained. Turley et al. [5] and Spooner et al. [6] considered this malformation to be the result of rupture of the sinus of Valsalva, occurring during intrauterine life or soon after birth. According to Levy et al. [3], the tunnel is an additional abnormal coronary artery, a fact that has been confirmed by histopathological examinations [4, 7]. Cooley et al. [8] explain that the pathogenesis of the tunnel is the result of delamination of the aortic wall, similar to that in Marfan's syndrome. Moreover, this suggestion has been supported by the existence of an increased amount of mucopolysaccharide substances in the aortic and tunnel wall [1, 4, 9].

A diagnosis of aortico-left ventricular tunnel should be considered whenever significant, often symptomatic, aortic insufficiency is present in the early period of life. This is the situation faced in the case reported by us. At first only severe aortic stenosis was recognised in the neonate, but the presence of haemodynamically significant aortic insufficiency (because of the existence of the tunnel to the LV) and the morphology of the lesion enabled us to make the correct diagnosis by ECHO only. Some authors recommend the cardiac catheterisation procedure for making this diagnosis.

An aortico-left ventricular tunnel requires differentiation from the ruptured aneurysm of the sinus of Valsalva, coronary artery arteriovenous fis-

tula and subaortic VSD with associated aortic insufficiency. In the last case the ventricular orifice of the tunnel may suggest the presence of a subaortic defect in the intraventricular septum and the regurgitant flow through the tunnel to the LV may be misinterpreted as aortic insufficiency associated with VSD. However, the application of colour Doppler showing the absence of a left-to-right shunt through the intraventricular septum and the presence of aortic insufficiency above the aortic valve made the diagnosis unequivocal.

In a ruptured aneurysm of the sinus of Valsalva the aortic orifice of communication is located below the coronary arteries, whereas in the case of aortico-left ventricular tunnel it is placed above the coronary arteries (usually above the right coronary artery) [3, 10]. In coronary artery arteriovenous fistula the characteristic coronary artery dilatation is noticed, a differential factor usually found in this case.

An aortico-left ventricular tunnel is a lesion which has a high mortality rate. According to the data from literature, sudden death may occur in 20% of the cases [2]. Surgical correction requires closure of the tunnel orifices. Moreover, the patch closure of both orifices is recommended [9, 11].

From literature data on the long-term follow-up it appears that after aortico-left ventricular tunnel surgical repair aortic insufficiency arises and becomes more intense with age. This is the effect of the postoperative distortion on the aortic root.

Direct suture of the base of the right coronary cusp to the aortic wall causes a shortening and dislocation of the cusp edge, resulting in valvular insufficiency. Patch closure of the aortic orifice of the tunnel stabilises the right coronary cusp to secure its optimal function. Thus, if possible, patch closure of both orifices of the tunnel is recommended [7, 11, 12].

In our case aortic insufficiency was noticed immediately after the operation, but this was the effect of the valvulotomy performed as a result of the presence of aortic valve stenosis.

In spite of early surgical treatment the prognosis for our patient should be cautious. This is so not only because of aortic valve malformation but also because of the persistent weakening of contractility of the significantly thickened LV wall.

In the further follow-up after three months we found mild aortic insufficiency (++) in the child. At present her condition is stable and she is putting on weight steadily.

## Conclusions

1. An aortico-left ventricular tunnel poses a difficult clinical problem.
2. The diagnosis may be based on a detailed echocardiographic examination, which eliminates the necessity of performing cardiac catheterisation.

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