

Case Reports

AORTO-LEFT VENTRICULAR TUNNEL. A CASE REPORT WITH AORTIC PRESSURE AND ECHOCARDIOGRAPHIC FINDINGS

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ABSTRACT

A seven-year-old girl with clinical diagnosis of aortic insufficiency underwent echocardiographic and hemodynamic evaluation and referred for surgery with the same diagnosis. However, at surgery, aorto-left ventricular tunnel was found and repaired successfully. Review of echocardiogram and catheterization data revealed interesting findings to differentiate this rare entity from congenital aortic regurgitation.

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CASE REPORT

A seven-year-old girl was admitted because of a heart murmur. She had a good general appearance, bounding pulses, a lateral and lower point of maximum impulse, markedly overactive heart, left ventricular lift, diastolic thrill and a loud diastolic murmur at the left sternal border and apex. Chest radiography showed marked cardiomegaly, left ventricular enlargement, and a wide mediastinum especially a dilated ascending aorta. Electrocardiography had signs of left ventricular hypertrophy with strain pattern. Echocardiography revealed left ventricular enlargement, overactivity, and dilated aortic root with an accessory channel in front of the aorta (Fig. 1,2). Diastolic pressure was higher in the aortic root than in the descending aorta (Table I). Cardiac angiography demonstrated left ventricular overactivity and dilated ascending aorta which projected anterior to the right aortic sinus.

At surgery a tunnel was found with an entrance above the right coronary sinus. The orifice was closed without a patch. The heart murmur and signs of aortic run off disappeared post-operatively.

DISCUSSION

In this extremely rare condition of congenital aortic

run off an endothelialized vascular channel exists between the aorta and left ventricle.¹⁻⁶ Morgan and Mazur suggested that formation of the tract was a consequence of a primary separation of the aortic annulus from the fibrous skeleton of the heart.^{6,7} The tunnel is thought to be present at birth. Its aortic origin is above the right sinus of valsalva and right coronary ostium, both of which are intact but displaced.³ There is usually a sharp ridge separating the sinus below and the orifice above. The aorto-left ventricle tunnel bypasses the aortic valve behind the infundibulum of the right ventricle and through the anterior and upper part of the ventricular septum and terminates in the left ventricle

Table I. Pressure Data of the Patient

Site	Pressure			O ₂ Saturation	Blood Gas
	S	D	M		
DAO	140	24	60	94%	PH=7.53
SVC				75%	PO ₂ =76
RA	a=6 v=2		2	74%	O ₂ Sat=96.5%
RV	48	2		78%	PCO ₂ =27.5
PA	42	12	20	77%	HCO ₃ =22.7
LV	110	6		92%	Total CO ₂ =23.7
Aortic root	110	62	80		BE=2.7

Cardiac output: 2.7 Cardiac index: 3.4 Ratio: 0.3
systemic vascular resistance: 10.6 pulmonary vascular resistance: 3

Aorto-Left Ventricular Tunnel

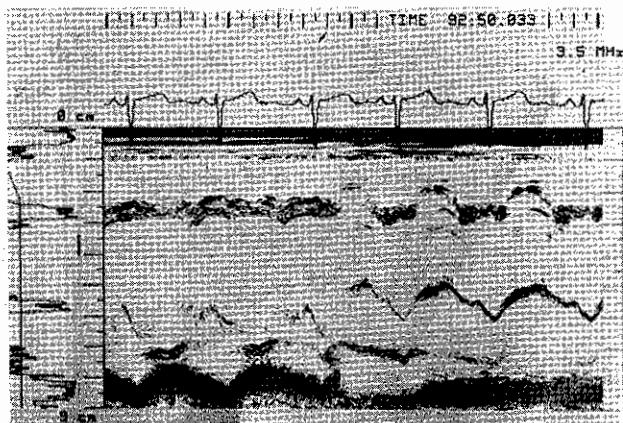


Fig 1. Pre-op, M-Mode echocardiography reveals ventricular dilation, fluttering of mitral valve and accessory channel, A.C., anterior part of aorta.

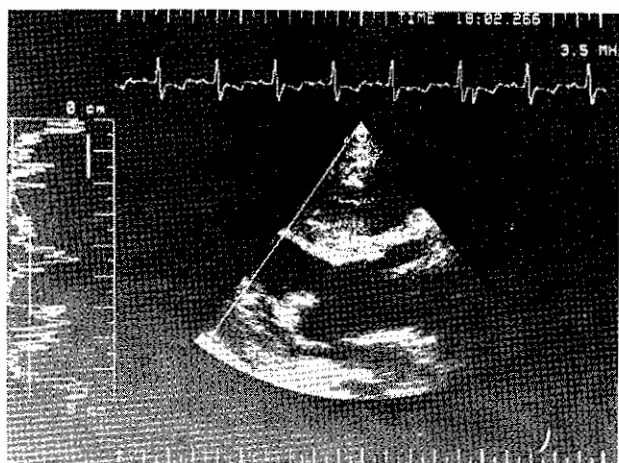


Fig 2. 2D echo-cardiography reveals accessory channel, A.C., anterior part of aorta, inspite of distal part of aorta.

just below the right and left aortic cusps.⁸ It is usually short and direct, though it may show aneurysmal dilatation.²

The diagnosis of aorto-left ventricular tunnel should be considered in any infant or young child presenting with clinical findings of severe aortic incompetence. Clinical picture includes wide pulse pressure, loud systolic and diastolic murmurs, usually with a momentary interval separating them, marked left ventricle enlargement and overactivity, and a dilated ascending aorta. Somerville suggested that the earlier the age at which these signs are found, the more likely the diagnosis of aorto-left ventricular tunnel, particularly so if the murmur is loud in the first months of life.⁴

15 patients of 20 reported cases by Okoroma and associates had the characteristic to-and-fro murmur during the first week of life.³ Nine patients developed congestive heart failure within the first year of life and only two patients were asymptomatic. Most of the patients (70%) were male.

Chest radiography may show dilatation of the

ascending aorta, left ventricular hypertrophy, and occasionally disproportionate dilatation of the right aortic sinus. Supportive information is the presence of a dirotic notch in the aortic pressure pulse indicating valve functions, despite the presence of the signs of aortic insufficiency. Ascending aortography will confirm the diagnosis. As pointed out by Somerville and associates,⁵ the upper part of the right aortic sinus is almost always large, eccentric, and projects anteriorly. The interventricular portion of the tunnel is frequently not clearly visualized. Central aortic valve regurgitation secondary to valve ring dilatation may occur. Levy, et al. first reported the entity as a congenital defect requiring surgery in early life.¹ Medical treatment is usually inadequate. Surgical intervention has been very successful in some of the reported patients. In each case, the sole procedure was closure of the aortic ostium of the tunnel. Early closure of the tunnel should prevent aortic valve deformity secondary to ring dilatation and should obviate the need for aortic valve replacement, so as to preserve the support of the right coronary cusp without distortion.^{2,4,5}

The differential diagnosis should include ventricular septal defect with aortic regurgitation, patent ductus arteriosus, aortic incompetence accompanying a bicuspid aortic valve, rupture of sinus of valsalva, and coronary AV fistula.⁴ Congenital absence of the pulmonary valve may have a to-and-fro systolic and diastolic murmur but the signs of aortic run off will be lacking and bronchial compression may dominate the clinical picture.⁴

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