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Brief Report

Valvar aortico-ventricular tunnel: an insight into the development of the great arteries

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Abstract Aorto-left ventricular tunnel is a rare congenital heart lesion, with an incidence of <0.1% of all CHD. We present a unique case of a valvar aorto-left ventricular tunnel in a neonate, in belief that our findings may shed some light on the developmental origins of this lesion.

Keywords: Aorto-left ventricular tunnel; CHD; cardiac development

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AORTO-LEFT VENTRICULAR TUNNEL IS A RARE congenital heart lesion, in which a paravalvar communication exists between the tubular aorta and, most commonly, the left ventricle, bypassing the hinges of the aortic valve leaflets. The first description was provided in 1963 by Levy et al.[1] Since then, >130 cases of this anomaly have been reported with variable features. In this study, we present a unique case of a valvar structure within the paravalvar channel. This finding may shed some light on the developmental origins of this lesion.

Case

A term neonate presented at 4 days of life for evaluation of a murmur. He was born after an uncomplicated gestation of 41 weeks, with a weight at birth of 3710 g. A murmur was heard at 12 hours of age, although he was asymptomatic, and he had no tachypnoea or feeding intolerance. His blood pressure was 75/30, and the distal pulses were bounding. The murmur, graded at 3/6, was of decrescendo nature and holodiastolic, being heard from the right upper sternal border down to the apex.

Transthoracic echocardiography showed a dilated left ventricle, with preserved systolic function and a fractional shortening of 34%. The morphology of the aortic valve was normal, but a large paravalvar tunnel was noted between the ascending aorta and the cavity of the left ventricle, permitting both systolic flow and diastolic flow. The aortic opening of the tunnel was above the right coronary sinus of Valsava, with the right coronary artery originating at the lateral aspect of the tunnel. An echogenic structure was seen within the tunnel. This was later evaluated by a preoperative transoesophageal echocardiography, which showed a rudimentary valvar structure at the level of the arterial valves (Fig 1 and Supplementary Videos 1 and 2). The consensus was to pursue surgical repair because of the size of the tunnel and the long-term concerns of aortic valve distortion.

On gross inspection of the heart in the operating room, there was a visible bulge passing along the anterior aspect of the aorta towards the right ventricular outflow tract. After establishing cardiopulmonary bypass and following antegrade cardioplegic arrest, a transverse aortotomy was made at the level of the sinutubular junction. A large opening was present at the level of the right coronary sinus. The orifice of the right coronary artery could be seen at the edge of the tunnel itself, with the artery coursing rightward to its normal location in the atrioventricular groove. Rudimentary myxomatous valvar tissue was present in the cavity of the tunnel. The aortic valve itself was trifoliate and normal.

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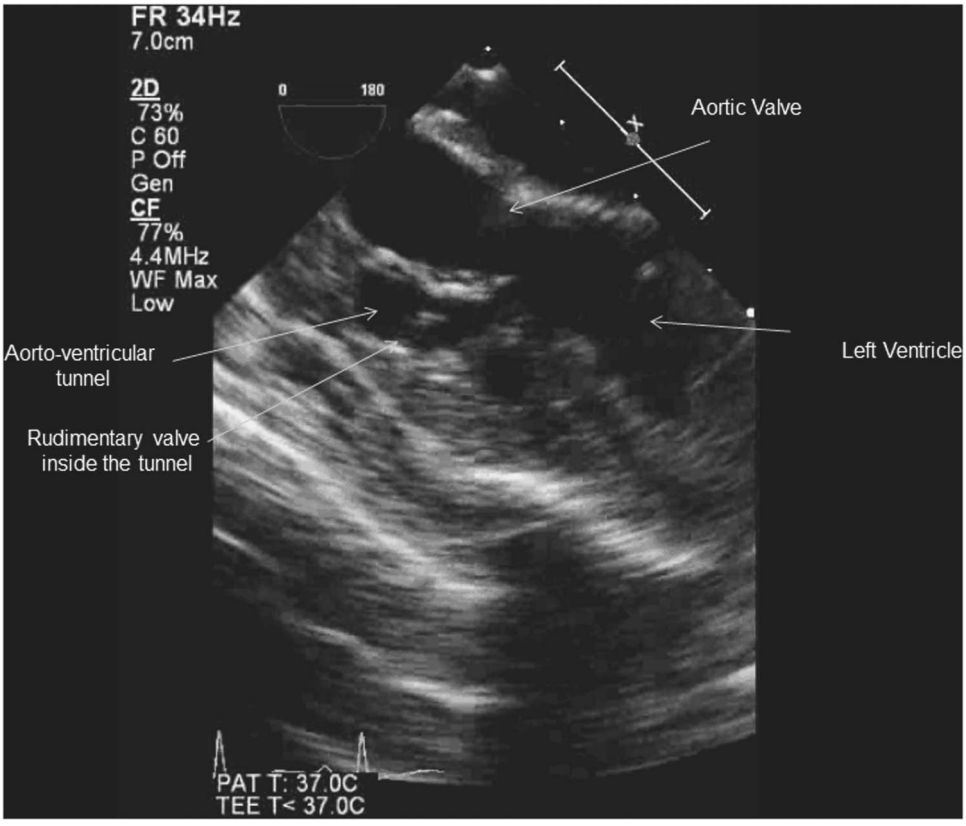

Figure 1.
Intraoperative two-dimensional transoesophageal echocardiogram showing the valvar structure inside the aorto-left ventricular tunnel.

The tunnel was closed by suturing a small patch of bovine pericardium in its mid portion. The patient was subsequently weaned from cardiopulmonary bypass with good haemodynamics on no inotropic support. A transoesophageal echocardiogram confirmed lack of flow through the tunnel. He had good ventricular function and normal flow within the right coronary artery. His postoperative course was uneventful, and he was discharged on the third postoperative day. Transthoracic echocardiograms at the 3- and 6-month follow-up showed no residual flow in the tunnel. The left ventricular dimensions and function were normal.

Discussion

Aorto-left ventricular tunnel is an extremely rare lesion, varying from 0.5% in congenital malformations identified by fetal echocardiogram[2] to between 0.05 and 0.1% of lesions identified in clinical series;[3] two-thirds of such patients are male.[4] The majority of tunnels represent communications between the aorta and the left ventricular cavity. Only one-eighth of the reported cases have involvment of the right ventricle. Their exact embryological basis remains unknown. Fetal echocardiogram has established that the lesion is congenital. Speculations of the developmental origin include an anomalous coronary artery opening in the left ventricle,[1] in utero rupture of a sinus of Valsava,[4] malformation of the outflow tract,[5] and degeneration of the anterior aortic wall, comparable with in utero cystic medial necrosis.[6] The origin of the distal part of the tunnel from the tubular aorta above the sinuses of Valsava and the normal coronary anatomy in many of these patients make the first two theories unlikely. It is well established that the histological appearance of the two ends of the tunnel differ markedly.[1,8] The aortic origin consists of fibrous tissue with smooth muscle cells and elastic fibres, whereas the ventricular end consists of myocardial tissue similar to the subpulmonary infundibulum. The most logical explanation is that the lesions represent an abnormal formation of the arterial valvar sinuses and leaflets.[7] The early developmental origin of this lesion has also been suggested by Bove's case series, where accessory valve tissue was seen at the aortic end of the tunnel in one of the patients.[8]

During early embryogenesis, the entire wall of the outflow tract is composed of myocardium.[9] As the division process progresses, the walls of the distal outflow tract are re-modelled to become arterial structures.

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The distal ends of the cushions that divide the outflow tract, along with the intercalated cushions, are themselves re-modelled to become the valvar leaflets as the myocardial cuff surrounding them regresses.[10] The disappearance of the muscular cuff causes the tissue plane developing between the infundibulum and the aortic sinuses to lie in communication with the extracardiac space. It is within this tissue plane that abnormal development will produce an aorto-ventricular tunnel, which persists as an anomalous channel joining the distal and proximal parts of the initial solitary outflow tract. The unique finding in our patient may have resulted potentially from the fusion of the right cushion not only with the left cushion as expected but also with the intercalated cushion, producing division of the aortic root.

The clinical presentation depends largely on the diameter of the tunnel, as well as the presence of compression of the coronary arteries and obstruction of either the right or the left ventricular outflow tract. The presence of a rudimentary valve in our patient likely limited the aortic diastolic runoff, which explains his minimal clinical symptoms on presentation. This unique finding in our patient gives insight to the developmental origins of the lesions.

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Conflicts of Interest

None.

Supplementary material

To view the supplementary material for this article, please visit http://dx.doi.org/10.1017/S1047951116001888.

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