Echocardiographic-anatomical correlations in aorto-left ventricular tunnel

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Abstract

Objective—To investigate the echocardiographic, morphological, and histological appearances of aorto-left ventricular tunnel observed in four fetal hearts and compare the findings with those reported in older patients with the malformation.

Background—Previous studies have concentrated on clinical features of the malformation from birth to adult life and have speculated on either its embryological formation or its acquisition during late intrauterine life. The presentation of a large series of cases in fetal life is a unique opportunity to study the malformation at an early stage in its natural course.

Methods—A retrospective study was performed of four cases of aorto-left ventricular tunnel discovered among 872 cases of congenital abnormalities diagnosed at a tertiary centre for fetal echocardiography. Detailed echocardiographic and anatomical observations were made of the malformation as identified during fetal life. The precise anatomical arrangement was determined and compared with previous descriptions found in journals published in English.

Results—In fetal life, as after birth, the malformation is characterised by enlargement and hypertrophy of the left ventricle, enlargement of the aortic root, and free regurgitation at the level of the aortic valve. Anatomical abnormalities are found at the aortic ventriculoarterial and sinutubular junctions as well as in the intervening aortic wall. These are unrelated to necrosis, ischaemia, or the presence of mucopolysaccharides.

Conclusions—The lesion is a developmental abnormality that should be reliably diagnosed by fetal echocardiography combined with colour flow Doppler echocardiography during the mid-trimester. The exact anatomical relations clarified by this study are pertinent to diagnosis and subsequent surgical correction.

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Keywords: aorto-left ventricular tunnel; echocardiographic-anatomical correlates; developmental malformation; fetal heart.

An aorto-left ventricular tunnel is an extremely rare lesion. This malformation was first recognised in 1961 and the anatomic features were subsequently emphasised by Levy et al. Since then approximately 71 cases have been reported in the English literature. The clinical profile and echocardiographic features are well described, but the precise anatomy remains difficult to understand.

We have now encountered four cases of aorto-left ventricular tunnel detected by echocardiography during intrauterine life. Careful analysis of this material has permitted us to clarify the morphological arrangement and histology of the lesion, its relation to adjacent cardiac structures, and suggest criteria to permit its future echocardiographic diagnosis even during fetal life.

Material and methods

ECHOCARDIOGRAPHY

Four fetuses (0.5%) from a series of 872 cases referred to our specialist centre between 1980 and 1992 had an aorto-left ventricular tunnel. Detailed examination was performed during fetal life using an Advanced Technical Laboratories Ultramark 3 mechanical sector scanner in one case and an Advanced Technical Laboratories Ultramark 4 and Hewlett Packard 77020 phased array imaging systems with 5 MHz or 3–5 MHz transducers in three. Doppler evaluation was performed in three cases and colour flow mapping in two.

ANATOMICAL AND HISTOLOGICAL ANALYSIS

Complete postmortem examination was carried out in each case. Three fetuses died during gestation, while the fourth patient died during the neonatal period. All external and internal abnormalities were documented, the heart was fixed by inflation with formalin, and dissection subsequently carried out following a standard procedure.

A single representative case (case 2) was selected for histological examination after careful documentation. The heart was imbedded in paraffin wax and sectioned serially at a thickness of 10 μm. All sections were retained according to a standard procedure and initially each 25th section was mounted and stained with the trichome technique. Further sections were mounted and stained as required. Staining with alcian blue was performed to determine the presence of acid mucopolysaccharides. Comparisons were made with serial sections of the aortic root of a normal fetal heart of the same gestational age.
Results

ECHOCARDIOGRAPHIC FEATURES

The mean (range) gestational age at referral to our centre was 23 (18-33) weeks. An initial diagnosis of aneurysm of the sinus of Valsalva with aortic stenosis and incompetence was made in the first patient at 33 weeks. Failure to respond to medical treatment after birth resulted in surgical intervention at 4 months. A tunnel found arising above the right coronary sinus and leading into the left ventricle was closed. The patient died 2 weeks after operation despite full medical support. The findings in the second fetus (seen at 19 weeks) simulated the anatomical configuration of Fallot's tetralogy with aortic regurgitation. In the third fetus (seen at 18 weeks) the leaflets of the aortic valve were thickened, there was dilatation of its orifice, and the aorta was aneurysmically dilated beyond the valve. The overall diagnosis was of profound aortic stenosis and incompetence. The fourth fetus (seen at 22 weeks) was again referred as a case of tetralogy of Fallot. In this fetus, however, the apparently deficient area of the ventricular septum, thought initially to represent a ventricular septal defect with aortic override, was shown to be a tunnel through which turbulent flow passed from the left ventricle to the aorta. The left ventricle in the four fetuses was not echogenic but dilated and thick walled with a decreased shortening fraction.

Parents of the last three fetuses elected to interrupt the pregnancy and the heart was subsequently obtained for study and comparison with the echocardiographic scans.

ANATOMICAL FINDINGS

Two fetuses were male. There were no external dysmorphic features and the only extracardiac anomaly found was a Meckel's diverticulum (case 2).

All hearts showed similar external appearances. There was pronounced enlargement of the left ventricle, which was approximately two to four times the size of the right ventricle and most notable in the neonatal heart. The aorta was enlarged in each fetal specimen (cases 2-4) with a diameter of 160-200% greater than mean normal values for the respective gestational ages. In general, the ascending aorta was uniformly dilated at its root but, in one fetal specimen (now 19 weeks gestation by foot length, case 3) there was an additional and distinctive aneurysmal bulge at the site of the right coronary sinus (fig 1(A)). The aortic end of the tunnel in the neonatal specimen had been closed during operation by resection of the aneurysmal dilatation over the right coronary sinus and suturing of the aortic wall to the fibrous bar supporting the right coronary aortic leaflet. There was also noticeable bulging of the ventricular septum into the posterior portion of the right ventricular outflow tract in this specimen.

All hearts showed normal connections of the cardiac segments and usual venoatrial connections. The origin of the tunnel in each case was at the site of the interleaflet triangle between the attachments of the valvar leaflets in the left and right coronary aortic sinuses. It then extended to varying degrees inferiorly beneath the right coronary aortic leaflet, which arose from a fibrous bar spanning the aortic root. The inferior aspect of the opening showed varying degrees of dilatation (figs 1(B) and 2). The tunnel itself was located in the fibro-fatty tissue plane separating the aortic and pulmonary trunks, with its right side bulging into the posterior portion of the right ventricular infundibulum. As the tunnel spiralled superiorly and to the right, it bypassed the hinge point of the right coronary leaflet of the aortic valve arising from the fibrous bar within the aortic root. In each case the tunnel led into the ascending aorta above the sinutubular junction of the right coronary sinus. The mid-point of the bar supporting the aortic...
leaflet extended in one case to a point level with the top of the aortic commissure. In this case the orifice of the right coronary artery arose within the fibromuscular bar, passed through it, and then extended posteriorly to reach the epicardial surface of the heart. The right coronary artery arose from the wall of the tunnel itself in the remaining cases. The fibrous bar supporting the aortic valvar leaflet in these hearts was smaller, with its mid-point not extending to the level of the commissural apices, thus producing a shallow right aortic sinus. The left coronary artery arose normally in each case. There was no fusion between adjacent leaflets of the aortic valves, and nodular dysplasia of the leaflets was only present in the neonate. In the second fetal specimen (now 22 weeks' gestation by foot length), however, there was moderate thickening of the valvar leaflets while, in the fourth (also 22 weeks' gestation), valve-like mucoid tissue was found on the superior aspect of the fibrous bar that separated the tunnel from the right coronary sinus (fig 2).

The ventricular origin of the tunnels was anteroccephalad to the area of the membranous septum separated from it by an area of muscular septum, which was largest when the ventricular opening of the tunnel was smallest. In case 2, which was studied histologically (see later), the left bundle branch could be traced arising from the lower aspect of the membranous septum and took a normal course down the ventricular septum, being unrelated to the ventricular orifice of the tunnel.

HISTOLOGICAL FINDINGS
The fibrous triangle separating the coronary leaflets of the aortic valve in the normal heart has an elastic media and is of similar thickness to the wall of the ascending aorta. At the base of the fibrous triangle, the aortic wall is joined to the ventricular myocardium by a layer of collagen (the anatomical ventriculoarterial junction). The right coronary leaflet attaches along this junction and rises to the apex of the interleaflet triangle (fig 3). In case 2 reported here the aortic wall beneath the right coronary leaflet was deficient and as a consequence the ventriculoarterial junction was anomalous. The right coronary leaflet itself was fixed on its upper surface to the fibrous bar spanning the aortic root (fig 4(B)). The leaflet and its supporting fibrous bar, therefore, formed the roof of the entrance of the tunnel into the left ventricle, the floor being formed by left ventricular myocardium (fig 4A and B).

The intercoronary aortic interleaflet triangle in the normal fetal heart is bordered anterio-

ly by the wall of the pulmonary trunk, the two structures being separated from one another by a fibro-fatty tissue plane (fig 3). In the abnormal heart the tunnel passed from the left ventricle into this fibro-fatty tissue plane. It did not pass through the wall of the right ventricular infundibulum, nor through the outlet septum, but merely displaced the wall of the right ventricular outflow tract (fig 4(A)). Thus, the inferior border of the tunnel was formed by the septal myocardium. The roof of the tunnel, however, was more arterial in composition (fig 4(B)). Its epicardial aspect consisted of loose vascularised collagen and sparse myofibres—similar structures being seen in the epicardial region of the normal right ventricle adjacent to the intercoronary aortic commissure. Internally, the roof was...
Figure 4 (A) Schematic representation of aorto-left ventricular tunnel, taken in the same plane as in fig 3, indicating the position of the histological sections in (B) and (C). (B) At the left ventricular opening of the tunnel (closed arrow) the right coronary leaflet is fixed only on its upper surface to the fibrous bar (F) separating the tunnel (T) from the aortic valve. The ventriculoarterial junction beneath is abnormally formed. The floor of the tunnel consists of infundibular myocardium, while the roof is arterial in structure. (C) This arrangement persists until reattachment of the right coronary leaflet to the left ventricular myocardium (closed arrow) from which point the entire tunnel is arterial in structure. R and N as in fig 1. Open arrow, opening of tunnel into aorta; pulm, pulmonary; other abbreviations as given in Fig 1. (Elastic van Gieson’s stain).

formed by a thick layer of fibrous tissue formed by intermingled collagen and elastic fibres. This configuration, of an arterial roof and a muscular floor, persisted along the tunnel until it reached the point of reattachment of the right coronary leaflet of the aortic valve to the left ventricular myocardium. From this region the tunnel was completely arterial in structure (fig 4(C)). At the junction of the tunnel with the ascending aortic wall the elastic fibres were continuous, well laminated and showed no signs of disorganisation. The right coronary artery joined the wall of the tunnel in this region. Mucopolysaccharides were not detected at any point within the tunnel.

Discussion
Aorto-left ventricular tunnel is a rare anomaly. In our review of the English literature, we have found only 71 cases reported in the 30 years since the original description by Edwards and its elaboration by Levy et al. The four cases reported here form, to the best of our knowledge, the largest detailed pathological study of this entity. As well as including, as far as we know, the only cases reported during fetal life, our experience has also enabled us to elucidate the anatomical features since before our study we had not totally understood the existing anatomical descriptions.

ANATOMICAL FEATURES
Existing descriptions and diagrams of the anatomical relation of the tunnel to the aortic valve and its supporting structures are, at best, ambiguous. Indeed, descriptions of the aortic and left ventricular origins are given in only six of 43 reports. Furthermore, such descriptions are usually confused by reference to an aortic “annulus” or “ring”.

The leaflets of the aortic valve, however, do not join the aortic wall in the form of a circle, but have semilunar attachments. Such an arrangement gives rise to spaces on the ventricular aspects of the leaflets, which can be termed the interleaflet triangles. The base of these triangles is formed by the anatomical ventriculoarterial junction and the apex by the peripheral attachment of the commissures at the level of the sinutubular junction. The triangles themselves are filled by an elastic wall of comparable thickness to the walls of the adjacent aortic sinuses.

In the tunnel studied histologically (case 2) the major disorder was found within the interleaflet triangle between the left and right coronary sinuses. Review of the literature shows this region to be described as the site of the ventricular opening in a further 11 (of 29) cases with tunnels. In the remaining 18 cases the origin is stated to be immediately below the attachments of the aortic valve, although its exact site is not specified. In our case the tunnel was an expansion of the interleaflet triangle that bulged into the tissue plane between the front of the aorta and the back of the subpulmonary infundibulum. This expansion had the effect of separating the attachment of the right coronary leaflet of the aortic valve from its ventricular support, the leaflet being attached to a dense fibrous bar that spanned the root of the right aortic sinus. The walls of the tunnel were made of fibroelastic tissue, but the base was the crest of the muscular septum. The tunnel itself, therefore, did not extend through the ventricular septum, as has previously been reported. Instead, it occupied the tissue plane between the subpulmonary infundibulum and the right aortic sinus, deforming the sinus by its position, but lying on the ventricular musculature. It is this expansion of the tunnel within the tissue plane which, by impinging on the infundibulum, produces subpulmonary narrowing, a feature reported in 31 of 71 reviewed cases. The normal anatomical relation of the aorta, pulmonary trunk, and subpulmonary infundibulum, however, are conserved in the presence of a tunnel.
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Surgical implications
Results of our study suggest that the difficulty in performing surgical closure without distortion of the aortic valve is a feature emphasised in several previous descriptions. 

Echocardiographic diagnosis
The diagnosis of aorto-left ventricular tunnel has frequently been made in postnatal life by cross sectional echocardiography and colour flow imaging. 

Aetiology
Our finding of aorto-left ventricular tunnel in the early mid-trimester confirms the suspicions of Levy et al. 

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3 Cooley RN, Harris LC, Rodin AE. Abnormal communication between the aorta and left ventricle—aortico-left ventricular tunnel. Circulation 1965;31:564-71.
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