Gross distortion of atrioventricular and ventriculo-arterial relations associated with left juxtaposition of atrial appendages

Bizarre form of atrioventricular criss-cross

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SUMMARY Two pathological specimens are described in which a bizarre distortion of atrioventricular and ventriculoarterial relations existed. In both cases anterior and leftward displacement of the tricuspid valve coexisted with rightward displacement of the infundibular regions and semilunar valves. Abnormal expansion of the AV canal, possibly secondary to malrotation of the cardiac tube, is considered as a possible cause of the anomaly.

One of the hearts showed severe subpulmonary stenosis produced by a valve-like structure which was well differentiated, with a complete annulus and chordae tendineae that inserted in a parachute-like fashion on the infundibular septum. The morphology of this structure was clearly different from that of the fibrous tags described in other anomalies. Its origin is uncertain.

These anomalies have important implications in relation to the nomenclature and classification of congenital heart malformations, and illustrate the value of recent modifications of the segmental approach.

The existence of hearts showing atrioventricular connections inappropriate to the relations of the atria and ventricles (for example, discordant atrioventricular connection in cases in which both morphologically right atrium and ventricle are placed to the right) was originally reported by Lev and Rowlatt (1961). Since then further reports of this anomaly have been published by Wagner *et al.* (1970), Van Praagh (1972), and Anderson *et al.* (1974a).

Such anomalies have interesting implications in relation to the nomenclature and classification of congenital heart malformations, which were reviewed by Anderson *et al.* (1974a). Their morphogenesis remains uncertain, but it has been suggested that these malformations result from abnormal rotation of the ventricular loop (Wagner *et al.*, 1970; Van Praagh, 1972). However the coexistence of other anomalies, which could be related to such malrotation, has not been adequately documented.

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In this report we describe two hearts in which distortion of atrioventricular valve relations produced an effect similar to that reported in 'criss-cross' hearts. However, they also showed gross additional malformations which have not, to our knowledge, been previously reported and which have interesting morphogenetic implications.

Case reports

CASE 1

This man was 25 years old when seen for the first time at Clínica Puerta de Hierro (Madrid), and had been breathless and cyanosed from early infancy. On admission he was severely incapacitated.

On examination he was found to be deeply cyanosed, with clubbing. The liver was 4 cm below the costal margin. Slight cardiomegaly was present with a praecordial thrill. On auscultation, there was a pansystolic murmur (grade 5/6) in the third and fourth intercostal spaces to the left of the sternum, a third heart sound, and a diastolic filling murmur.

The haematocrit was 65 per cent, haemoglobin 18 g/dl, and arterial oxygen saturation 58.5 per cent (breathing 100% oxygen). The electrocardiogram showed counterclockwise rotation of the QRS loop in the frontal plane with an axis of 90°, biatrial enlargement, and left ventricular epicardial patterns in all the praecordial leads. The chest x-ray film showed slight cardiomegaly with absence of the pulmonary arc and increased convexity of the lower left border of the heart; there was decreased pulmonary vascularity with signs of collateral circulation.

Angiocardiograms were interpreted as showing situs solitus and double inlet ventricle with double-outlet outlet chamber (aorta anterior and to the right of the pulmonary artery). There was left-sided juxtaposition of the atrial appendages and moderate regurgitation through the right atrioventricular valve. Severe subpulmonary stenosis was present.

In view of the hypoxaemia and the complexity of the case, an aortopulmonary shunt was attempted. The patient died in the postoperative period.

Necropsy showed viscerotrial situs solitus, with

the cardiac apex pointing to the right (Fig. 1). There were two superior venae cavae of about the same size (the left one emptying into the coronary sinus). All the pulmonary veins drained into a cavity situated, for the most part, above and behind the right atrium, though it communicated only with the left atrium (thus resembling *cor triatriatum*). Left-sided juxtaposition of the atrial appendages was present. The left AV valve was morphologically mitral. It was placed in its normal position, being the most posterior of the 4 cardiac valves (Fig. 2a). The right AV valve was heavily calcified and displaced forwards compared with its normal position, lying on the anterior part of the floor of the right atrium (Fig. 2b), in front of the left AV valve; its morphology was atypical, having two leaflets and three groups of chordae tendineae which were inserted into the free wall of the main ventricular chamber and on to the borders of an anterosuperior outlet foramen (Fig. 3). Both AV valves opened into a large primitive ventricle which communicated with a small, right-sided outlet chamber through two defects. The outlet chamber was heavily trabeculated and gave rise to the aorta, while the pulmonary artery (placed just to the right of the right AV valve) was mainly arising above the septum.

The apical septal defect was situated in the trabeculated septum. The other defect was anterosuperior, produced by a malalignment between the infundibular septum (displaced to the right) and the other septal structures; its limits were the anterior wall of the ventricle, the muscular septum, the pulmonary outlet, and the right AV ring.

The pulmonary trunk had a diameter about three-quarters that of the aorta and straddled the septum, arising mainly from the outlet chamber, slightly behind and to the right of the right AV valve. The pulmonary valve was tricuspid and one of the leaflets was smaller than the other two. A raphe originated from one of the commissures and inserted in the wall of the pulmonary trunk.

There was another valve-like structure just below the pulmonary valve (Fig. 4). It was composed of tissue similar to the right AV valve with which it had fibrous continuity. It had three fibrous bands that had the appearance of fused commissures but which differed in position from the pulmonary commissures. There was an orifice of about 2 mm in the valve-like structure leading from the primitive ventricle to the pulmonary artery. Chordae tendineae arose from the leaflets of this valve and came together, like the strings of a parachute, in one main stem that inserted on the infundibular septum. This was the only structure producing stenosis in the subpulmonary tract.

Fig. 1 External view of the specimen of case 1, showing left-sided juxtaposition (RA, right atrial appendage; LA, left atrial appendage). The pulmonary trunk (P) origin is lower than the aortic (Ao). The descending or delimiting anterior artery is the boundary (IVA) between the outlet chamber and the primitive ventricle.

Fig. 2a Case 1. Diagram to illustrate relation of ventricular chambers, atrioventricular valves, and semilunar valves. A, aorta; P, pulmonary artery; T, right AV valve; M, left AV valve; OC, outlet chamber; V, primitive ventricle. Fig. 2b Case 1. Opened right atrial appendage shows the displacement of a heavily calcified right AV valve (T). Arrow points to the communication with the left atrium (CIA).

Fig. 3 Case 1. Left ventricular septal aspect. Arrow points to the superior VSD (IVF). Right AV valve chordae tendineae are inserted upwards into the free ventricular wall, and downwards onto the septum; calcification of this valve is noteworthy. The mitral valve (M) is normal. S, septum; T, right AV valve; RA, right atrium; LA, left atrium.

Fig. 4a Case 1. Location and relations of the subpulmonary valve-like structure and the right AV valve. IS, infundibular septum.

Fig. 4b Case 1. Pulmonary outflow tract has been opened, showing the pulmonary valve (PV) and a subpulmonary valve-like structure (SPV), whose leaflets are in continuity with the right AV valve (T). Subpulmonary valve has been cut and arrow points to chordae tendineae (CT), arising like strings of a parachute from a main stem which passes rightwards, posterior, and inferior, and arises from the infundibular septum.

CASE 2

This premature infant with a birthweight of 1.9 kg presented in the neonatal period with cyanosis and cardiac failure. Chest x-ray film showed dextrocardia in the presence of visceral situs solitus, and the electrocardiogram showed right atrial hypertrophy with P axis $+90^\circ$ and QRS axis 0° . All praecordial leads showed qR complexes with tall R waves (30 mm). She deteriorated despite vigorous medical treatment and died before cardiac catheterisation and angiocardiography could be performed.

Necropsy showed atrial situs solitus, with the apex pointing to the right (Fig. 5). Left-sided juxtaposition of the atrial appendages was present. The right atrium received superior and inferior venae cavae and coronary sinus; a large secundum atrial septal defect was present. The left atrium received 4 pulmonary veins.

The right atrium was connected to an anterior ventricular chamber via a tricuspid valve which straddled the inflow septum by about 25 per cent, the septal cusp being attached to the crest of the septum. The inflow portion of the anterior ventricle lay on the left anterior shoulder of the ventricular mass. The trabeculated zone and outflow portion of the anterior ventricle lay on the right anterior shoulder of the ventricular mass, and both great arteries took origin from this ventricle, with a bilateral muscular infundibulum (Fig. 6). The tricuspid valve had septal, left, and right anterior cusps. In front of the latter was the ventriculo-infundibular fold separating it from the pulmonary valve. To the right of the pulmonary outflow tract was a well-formed infundibular septum with prominent septal and parietal extensions, which formed well-defined muscle bands around the

Fig. 5 Anterior view of specimen from case 2 showing juxtaposed appendages. The transverse ventricular incision opens into the anterior morphological right ventricle. Dots delineate right ventricle.

ventricular chamber, producing some obstruction to the aortic outflow tract. The latter lay further to the right and more posteriorly on the right shoulder of the ventricular mass and consisted of a trabecular zone with short infundibular segment. The aortic valve lay to the right of, and slightly posterior to, the pulmonary valve. There was a hypoplastic aortic arch and a persistent ductus arteriosus.

The left atrium connected with a posterior ventricle via a mitral valve (Fig. 7) and this ventricle formed the apex of the ventricular mass which was directed to the right. The posterior ventricle was a morphological left ventricle. The mitral valve was supported by multiple small papillary muscles on the parietal aspect (lying to the left). The septal cusp was orientated to the right and anteriorly. The posterior ventricle was connected to the anterior ventricle via two septal defects, the more leftward of which was an atrioventricular canal type of defect allowing some straddling of the tricuspid valve. The more rightward defect was a muscular defect which opened into the anterior ventricle just to the right of the infundibular septum and into the aortic outflow tract.

Fig. 6 Case 2. Anterior view of right ventricle opened showing outflow tracts to aorta and pulmonary artery, tricuspid valve, and two ventricular septal defects.

Fig. 7 Case 2. Posterior view of heart with morphological left ventricle opened. Probe is passed through a ventricular septal defect into outflow tract of right ventricle. Open arrow pointing behind anterior leaflet of mitral valve indicates position of other ventricular septal defect.

Fig. 8 Case 2. Specimen viewed from above with atria opened, showing relation of tricuspid and mitral valves.

The tricuspid valve lay more or less directly anterior to the mitral valve (Fig. 8, 9) and was directed anteriorly and to the left while the mitral valve was directed inferiorly and to the right. This produced a criss-cross effect involving the inflow areas of the ventricles, but the outflow tract of the anterior ventricle extending to the right tended to mask this effect.

Discussion

Both specimens showed striking morphological similarities which suggest similar pathogenetic

Fig. 9 Case 2. Diagram to illustrate relation of ventricular chambers, atrioventricular valves, and semi-lunar valves. Infundibular septum indicated by light cross-hatching.

mechanisms. Conspicuous distortion of AV valve relations was present in both hearts. Though its cause is uncertain, malrotation of the ventricular loop can be inferred from the presence of a right-sided apex and auricular juxtaposition.

The abnormal situation of the AV valves could be related to abnormal expansion of the AV canal. In normal cardiogenesis a number of morphogenetic movements, well described by Goor *et al.* (1972) and Anderson *et al.* (1974b), lead to a situation in which the posterior great artery is placed in front of and between both AV valves, with fibrous continuity among these structures. However, in our cases both great vessels remained to the right of the abnormally anterior right AV valve; this abnormal relation could explain the presence of double outlet right ventricle or outlet chamber in our cases (because the connection of any great artery to the left ventricle would be prevented by the mechanical obstacle of the right AV valve). The abnormal relations between the valves in these two cases do not occur during normal stages of cardiogenesis, and thus it seems likely that the abnormal location of the right AV valve (the most clearly pathologically situated in our cases) is a vital part of this anomaly. This abnormal location cannot be explained by a simple failure in rightwards expansion of the AV canal, and it is probable that an abnormally directed expansion took place. Whether this abnormal expansion is produced (or conditioned) by a primary anomaly of the AV canal, or is induced by other anomalies cannot be deduced with certainty, but this second possibility would be supported by the presence of other anomalies (of earlier genesis) suggestive of malrotation (right-sided apex, auricular juxtaposition).

The existence of fibrous structures ('fibrous tags') in the outflow tract of a ventricle or outlet chamber has been reported in association with several anomalies (mainly conotruncal malformations) (Levy *et al.*, 1963; Rastelli *et al.*, 1969; Riemen-schneider *et al.*, 1969; Anderson *et al.*, 1975; Otero Coto *et al.*, 1978). It is unusual for such structures to produce significant obstruction. However, in our first case a valvular structure, supported by chordae tendineae arranged in a parachute fashion, with a small central opening, was present below the pulmonary valve, severely obstructing this outflow tract. This type of obstruction has not been previously reported to our knowledge. The origin of this structure is uncertain. It was not part of the right or left AV valves from which it was separated and had its own annulus just below the pulmonary valve. Nor does it seem to have been a derivative of the membranous septum, because the presence of double inlet ventricle argues against the existence of

that structure and neither the location nor the morphology of the abnormal valve supports such a possibility. Its origin might be related to the differentiation of tissue derived from the cardiac jelly of Davis present in the bulbus and the bulbo-ventricular junction, which has been said to be involved in the formation of part of the normal tricuspid valve (Ugarte *et al.*, 1976).

We have found incomplete absorption of the common pulmonary vein into the left atrium in several cases with a large left superior vena cava draining into the coronary sinus (Otero Coto, 1977). Thus we think that the abnormal morphology of non-restrictive cor triatriatum found in case 1 may be related to the presence of a large coronary sinus receiving a left superior vena cava. This fact could have interest in angiocardigraphic evaluation of cases with this anomaly.

The existence of the reported anomalies has important implications in relation to the nomenclature of congenital heart disease. They offer, in our view, further evidence that any useful segmental approach must include both external relations between the segments and their connections. We believe that recent modifications (Otero Coto *et al.*, 1976; Otero Coto and Quero Jiménez, 1977; Shinebourne *et al.*, 1978) to the segmental approach of Van Praagh *et al.* (1964), whose initial limitations were recognised and largely overcome by Van Praagh himself in a later paper (1972), are a helpful advance in this field.

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