Double outlet left ventricle

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A case, the sixth, of double outlet left ventricle is reported, the diagnosis being made by cardiac catheterization and angiocardiography. Though the existence of such a malformation has been questioned on embryological grounds, double outlet left ventricle must now be regarded as a recognized cardiac anomaly. The embryological significance of this case is discussed and it is suggested that the malformation is easily explained by the hypothesis of differential conal absorption.

Although the existence of double outlet left ventricle has been doubted on embryological grounds (Grant, 1962; Van Mierop and Wigglesworth, 1963), 5 cases have recently been reported (Sakakibara et al., 1967; Paul et al., 1970; Kerr et al., 1971; Pacifico et al., 1972) and it is now a recognized congenital cardiac malformation. In this paper, a further case of double outlet left ventricle is described and some of the diagnostic problems and possible surgical approaches relating to this rare condition are discussed as are the embryological events which may produce the anomaly.

Case report

The patient was a 5-year-old Iranian boy with no family history of heart disease. There was no known exposure to x-rays, potentially teratogenic drugs, or viruses during pregnancy, and his birth was normal at term. Persistent central cyanosis appeared during the first 10 days of life, and, latterly, mild effort dyspnoea had been noted. He had not been greatly limited by these symptoms, and there had been no feeding difficulties, squatting episodes, or tendency to recurrent respiratory infections.

On examination, he was small (height and weight on tenth centile) with pronounced clubbing and central cyanosis. Peripheral arterial and jugular venous pulses were normal, and oedema was not present. The cardiac impulse was normal. On auscultation, there was a grade 3/6 ejection systolic murmur maximal at the left second intercostal space, the second sound was single, and no ejection click was audible. No abnormality was found elsewhere. The electrocardiogram showed sinus rhythm, a mean frontal QRS axis of $-150^\circ$, and a dominant 15 mm $R$ wave in V$4_R$ and V$1$ (Fig. 1). The plain chest x-ray revealed only slight cardiac enlargement and the pulmonary vasculature appeared normal. The haemoglobin was 17.1 g/100 ml. The differential diagnosis was tetralogy of Fallot, double outlet right ventricle with pulmonary stenosis and endocardial cushion defect with pulmonary stenosis.

At cardiac catheterization, the right side of the heart was studied from the right axillary vein and the left side from the right saphenous vein via the foramen ovale which was patent. The catheter crossed a ventricular septal defect to enter the aorta; the pulmonary artery was entered from the left ventricle using a Swan-Ganz balloon-tipped flow-directed catheter. The pressures and oxygen saturations obtained are shown in the Table. A large fall in oxygen saturation was apparent between left ventricle and aorta, and a similarly large increase between right ventricle and pulmonary artery, so that the oxygen saturation in the pulmonary artery was almost equal to that in the aorta. Right and left ventricular pressures were identical. There was valvar pulmonary stenosis (systolic gradient of 83 mmHg), but calculated pulmonary blood flow was slightly greater than systemic blood flow.

Serial angiocardiograms were performed with injections of contrast medium in the aorta, right ventricle, and left atrium. The right ventriculogram (Fig. 2) revealed that the anterior, morphological right ventricular cavity ended blindly superiorly and appeared to

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure (mmHg)</th>
<th>Oxygen saturation (%)*</th>
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<tbody>
<tr>
<td>Right atrium</td>
<td>Mean 3</td>
<td>61</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>95/1-6</td>
<td>65</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>12/9 mean 11.1</td>
<td>81</td>
</tr>
<tr>
<td>Left atrium</td>
<td>mean 7</td>
<td>99</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>95/2-6</td>
<td>99</td>
</tr>
<tr>
<td>Aorta</td>
<td>95/75</td>
<td>82</td>
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* Measured by reflection oximetry. Qp/Qs = 4.2/3.9 l/min per m.$^2$
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**FIG. 1** Electrocardiogram.

**FIG. 2** Right ventricular injection, lateral projection. There is opacification of the left ventricle via a ventricular septal defect. The pulmonary artery fills from the left ventricle and pulmonary stenosis is demonstrated.
lack an infundibulum. All the contrast passed posteriorly through a large ventricular septal defect into a morphological left ventricle, and thence into both aorta and pulmonary artery. A subpulmonary conus was visible, the pulmonary valve was small and slightly thickened, and the pulmonary vessels were all somewhat reduced in size. The left atrial injection (Fig. 3) confirmed that both pulmonary artery and aorta arose from the morphological left ventricle and mitral-aortic continuity was clearly demonstrated. The pulmonary valve lay to the left, posteriorly and superiorly in relation to the aortic valve. No contrast passed into the right atrium or right ventricle during this angiogram. An aortogram showed a left-sided aortic arch with some dilatation of the ascending aorta. There was slight opacification of the pulmonary arterial tree via moderately enlarged bronchial arteries, but no evidence of any other communication between aorta and pulmonary artery. The final catheter diagnosis was double outlet left ventricle with a ventricular septal defect, valvar pulmonary stenosis, and a persistent foramen ovale.

A palliative aorta to right pulmonary artery shunt was performed by Mr. M. Paneth. At operation, the external appearances of the heart were found to be compatible with the catheter diagnosis. Postoperative progress was uneventful and the patient was discharged with a view to performing total correction at some future time.

Discussion

Double outlet left ventricle may be defined as the origin of both pulmonary artery and aorta from the morphological left ventricle. Malformations where the great arteries arise from the morphological right ventricle, which is left-sided by virtue of associated ventricular inversion, are best regarded as a variant of double-outlet right ventricle (Fragoyannis and Kardalinos, 1962; Ruttenburg et al., 1964; Kiser et al., 1968). The patient presented in this paper thus has true double outlet left ventricle as defined above.

The existing case reports of double outlet left ventricle, though few in number (Sakakibara et al., 1967; Paul et al., 1970; Kerr et al., 1971; Pacifico et al., 1972), clearly demonstrate that wide variation in anatomy is possible. The major differences are with respect to the presence or absence of a ventricular septal defect and of pulmonary stenosis, the distribution of conal musculature, and the interrelations of the great arteries. Thus, in the case of double outlet left ventricle reported by Sakakibara et al. (1967), there was no pulmonary stenosis, there was a large subpulmonary conal septal defect, and the aorta and pulmonary artery were normally related. Paul et al. (1970) described a case of double outlet left ventricle with no pulmonary stenosis, but an intact ventricular septum. Conal tissue was bilaterally hypoplastic and the semilunar valves lay side by side equally posteriorly, the aortic valve to the right and the pulmonary valve to the left at approximately the same height. In contrast, in the patient of Kerr et al. (1971) double outlet left ventricle was associated with a subpulmonary ventricular septal defect and severe pulmonary stenosis, though the relative positions of aorta and pulmonary artery were similar to those in the previous case (Paul et al., 1970). These three examples of double outlet left ventricle demonstrate quite distinct anatomy, and form the basis of a classification of double outlet left ventricle suggested recently by R. Van Praagh (1973, personal communication). The lesions in our case appear to be very similar to those found by Kerr et al. (1971), except that the pulmonary valve was slightly posterior and superior to the aortic valve.

There is corresponding diversity in the clinical features. In the cases of Sakakibara et al. (1967) and Paul et al. (1970), in whom pulmonary stenosis and definite cyanosis were absent, the diagnoses were ventricular septal defect with pulmonary hypertension and tricuspid atresia with d-transposition respectively. In the patient of Kerr et al. (1971) where severe pulmonary stenosis and pronounced cyanosis and clubbing were present, the initial diag-
nosis was tetralogy of Fallot. The correct diagnosis was not appreciated at cardiac catheterization in two of the reported cases (Sakakibara et al., 1967; Kerr et al., 1971) and only at repeat catheterization in another (Paul et al., 1970). Commenting on the difficulty of diagnosis, Kerr et al. (1971) point out that the condition may be missed if left-sided injections of contrast medium are not made. However the origin of the pulmonary artery from the left ventricle was clearly visible during the right ventriculogram in our patient, and was seen in retrospect in the case of Kerr et al. (1971).

While left-sided injections may not be performed routinely in patients suspected of having tetralogy of Fallot, it is noteworthy that in our patient and in those reported cases for whom figures are available (Sakakibara et al., 1967; Paul et al., 1970), oxygen saturations in the aorta and pulmonary artery were almost identical. Such findings are incompatible with a diagnosis of tetralogy of Fallot and should indicate the true diagnosis to the operator and alert him to the need for a left-sided injection of contrast medium.

The different haemodynamic abnormalities in these cases have also resulted in different surgical approaches. In the acyanotic patients (Paul et al., 1970; Kerr et al., 1971) the problem was one of left-to-right shunting, since both the pulmonary artery and aorta were exposed to left ventricular pressure while pulmonary vascular resistance had not yet risen above systemic. Palliative pulmonary artery banding was therefore performed in Paul's patient (Paul et al., 1970), while in Sakakibara's case (Sakakibara et al., 1967) total correction was achieved, since intraventricular diversion of blood from the right ventricle to the pulmonary artery was readily accomplished using a patch to repair the conal septal defect. Conversely, in our patient and in that reported by Kerr et al. (1971) with severe pulmonary stenosis and pronounced cyanosis, palliation was achieved by anastomosis between the pulmonary artery and aorta. Total correction was later performed in the latter patient after two shunts had failed to function, by creation of an artificial tunnel from right ventricle to pulmonary artery, roofed anteriorly with a free pericardial graft (Kerr et al., 1971).

Alternatively, total correction may be achieved with an extracardiac conduit from the right ventricle to the pulmonary artery as described recently in two further patients by Pacifico et al. (1972). While palliation by means of either pulmonary artery banding or an aorto-pulmonary anastomosis is possible, the results of total correction in double outlet left ventricle appear particularly encouraging. All four reported patients in whom this has been carried out (Sakakibara et al., 1967; Kerr et al., 1971; Pacifico et al., 1962) were alive and well at 3 months, 1 year, 3½ years, and 6 years.

Embryology

When distinguished authorities expressed the opinion that double outlet left ventricle was an embryological impossibility (Grant, 1962; Van Mierop and Wigglesworth, 1963), it was generally held that transposition of the great arteries was the result of normally spiral bulbar ridges fusing in straight fashion (de la Cruz and da Rocha, 1956). Using the same criteria, Van Mierop and Wigglesworth (1963) also stated that only the classically complete and corrected forms of transposition existed. Since that time the careful studies of Van Praagh and Van Praagh, 1966; Van Praagh, 1973 have shown that many forms of transposition exist which cannot be explained on the 'straight septum' hypothesis. For this reason, Van Praagh and Van Praagh (1966) propounded their concept of differential conal growth. This hypothesis was also not ideal, since at no stage of normal embryonic growth is the pulmonary artery ever above the primitive ventricle, an essential premise of Van Praagh's concept. This fact has been elegantly demonstrated by the study of Goor, Dische, and Lillehei (1972) and endorsed independently by Anderson and associates (Anderson, Wilkinson, and Arnold, 1974; Anderson et al., 1973). As Goor has pointed out, these studies are not original since previous studies are extant in the German literature; however they clearly show that the aorta is taken above the left ventricle by a process of differential conal absorption. Furthermore, an additional essential part of normal growth is that bulbar rotation must occur, so that the presumptive aortic conus is carried posteriorly to the presumptive pulmonary artery. The vital part of Goor's work shows that if this rotation does not occur then the aorta and pulmonary artery would be situated in the same frontal plane, with the aorta to the right. This is the precise position of the great arteries in most cases of double outlet right ventricle, where the bilateral conus indicates the absence of conal absorption. In an equally elegant paper, Goor and Edwards (1973) have shown that examination of pathological specimens provides evidence to show a spectrum of abnormalities between double outlet right ventricle and complete transposition depending upon the degree of absorption of the subpulmonary conus. This investigation has also been endorsed independently by Anderson and associates (Anderson and Ashley, 1973). In complete transposition therefore, the ventricular septal defect is not a typical
membranous septal defect, but represents the gap between the ventricular and conal septa. If this gap is large it is also possible for partial absorption of the aortic conus to occur through it. This produces posterior displacement of the aorta, and the so-called ‘posterior transposition with mitral-aortic fibrous continuity’ (Van Praagh et al., 1971). Clearly if this absorptive process continued, then both conuses would be absorbed above the primitive ventricle, producing double outlet left ventricle. Since there is usually a pulmonary conus in posterior transposition, it would be expected that in double outlet left ventricle, while the aortic conus would be deficient, the pulmonary conus could be either absent or present. This supposition is borne out by the published cases. The one case that is difficult to explain is that with an intact septum, but then cases of double outlet right ventricle with intact septum are equally difficult to explain (Oppenheimer-Dekker and Gittenberger-De Groot, 1971). However, in our experience (Anderson et al., 1973 and unpublished data), microscopical evidence of a bulboventricular foramen is always present, and it is likely that the intact septum is purely ventricular in origin. Thus the previous cases together with our own attest to the existence of double outlet left ventricle, and we believe that the anomaly is easily explained by the differential conal absorption hypothesis.

References


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