Transposition of the great arteries associated with a double left ventricular outflow tract

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SUMMARY A case is described in which, at semilunar valve level, the aorta and pulmonary artery arose from inappropriate ventricles. Despite this, the outflow tracts to both vessels originated from the left ventricle. Embryologically, it is speculated that this anomaly is the result of normal rotation of the proximal conus, without concomitant truncal inversion, and excessive leftward shift of the proximal conus and conal septum or anterior and rightward deviation of the anterior segment of the ventricular septum. Surgical repair using a double conduit between the right ventricle and pulmonary artery and left ventricle and aorta, respectively, was unsuccessful.

Definition of specific outflow tract anomalies is often controversial. For example, transposition of the great arteries has variously been defined as a reversal of the anteroposterior relation of the semilunar valves (Goor and Edwards, 1973), any alteration in the position of the great arteries (Abbot, 1927), and the presence of mitral-aortic valve discontinuity as a result of a subaortic conus (Van Praagh et al., 1967). Recently, several authors have favoured a more literal definition, redefining transposition of the great arteries to mean that both great arteries are 'placed across' the septum (trans-ponere means 'placed across') reversing the connection of the great arteries to the morphological ventricle (Van Praagh, 1973; Shinebourne et al., 1976). Thus both great arteries arise from morphologically inappropriate ventricles. By the same token, double outlet ventricle is defined as a specific ventriculo-arterial connection in which more than one and a half great arteries arise from the same ventricular or outlet chamber (Kirklin et al., 1973). We have accepted this literal definition of outflow tract anomalies.

However, according to the interpretation of embryological events by Goor and Lillehei (1975), the semilunar valves and outflow tracts of each of the major vessels are derived from different embryological segments. It is reasonable to anticipate, therefore, that the ventricle of origin of a major vessel (at semilunar valve level) may be at variance with that of its outflow tract. In this communication a case will be described in which the left ventricle has a double outflow tract (aortic and pulmonary), but the origin of the great arteries at semilunar valve level is clearly from inappropriate morphological ventricles.

Terminology

We have adopted the terminology recently proposed by Anderson et al. (1977) with regard to outflow tract anomalies. The term 'conus' (and its derivatives) is confined to embryological events. In the postnatal, or fully developed heart, the term 'infundibulum' is substituted for 'conus'. Thus the structure interposed between the two semilunar valves is described as the infundibular septum (parietal band of Van Praagh) (Van Praagh et al., 1975). The muscle separating semilunar from atrio-ventricular valves is termed the ventriculo-infundibular fold regardless of whether it exists in the right or left ventricle. The trabecula septomarginalis (septal band of Van Praagh) (Van Praagh et al., 1975) is an extensive right ventricular trabeculation with, superiorly, anterior and posterior limbs, reinforcing the anterior and inferior borders of many septal defects.

Case report

A 5½-year-old boy had been cyanosed since birth. Chest x-ray film showed cardiomegaly and pulmonary plethora while the electrocardiogram

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showed a mean frontal QRS axis of $+60^\circ$, with evidence of right atrial and right ventricular enlargement. Small R waves were noted in the right chest lead. Cardiac catheterisation at age 6 days established the diagnosis of transposition of the great arteries, mild pulmonary stenosis, and a small ventricular septal defect. A Rashkind septostomy was performed with considerable improvement in oxygen saturation.

During the next 3 years increasing polycythaemia developed requiring multiple vensections. Cerebral thrombosis and hemiplegia occurred at age 1$\frac{1}{2}$ years with fairly good recovery. A Blalock-Taussig shunt was performed when the child was 4 years old, with subsequent improvement in oxygen saturation and fall in haematocrit. Recatheterisation at 5$\frac{1}{2}$ years confirmed the previous diagnoses. A systolic gradient of 41 mmHg was measured across the pulmonary outflow tract and the systemic oxygen saturation was 75 per cent. When the left ventricular systolic pressure was 70 mmHg, that in the right ventricle was 80 mmHg. Elective surgery was advised.

Using standard cardiopulmonary bypass techniques, a right ventriculotomy was performed and a 2 to 3 mm ventricular septal defect closed with a single horizontal mattress suture. A valved external conduit (16 mm) was then inserted between the right ventricle and right pulmonary artery. A similar conduit (18 mm) was placed between the apex of the left ventricle and left lateral wall of the ascending aorta. Initially, after discontinuing bypass, the haemodynamics appeared satisfactory. However, just before closing the chest, ventricular fibrillation occurred and resuscitation was not successful.

Necropsy confirmed ventricular non-inversion: the morphological right ventricle was right-sided and the morphological left ventricle was left-sided. The aortic valve was anterior, superior, and to the right, above the right ventricle. The pulmonary valve was to the left of, and slightly posterior to, the aortic valve. Though the pulmonary valve was overriding both ventricles (similar to the aorta in the normal heart), there was no direct communication between the right ventricle and the pulmonary valve or artery. Examination of the interior of the left ventricle disclosed two outflow tracts, separated by a muscular ridge (infundibular septum) approximately in a coronal plane (Fig. 1). The aortic outflow tract was dextro-posterior to the pulmonary outflow tract. The interior of the right ventricle (Fig. 2 and 3a) showed a ventricular septal defect 3 mm in diameter (the surgical suture was removed), triangular in shape, and immediately subaortic. It was bounded by the posterior limb of the trabecula septomarginalis, aortic valve, and, on the left, the anterior ventricular septum. The posteroinferior rim of the defect was separated from the tricuspid valve by a 3 to 4 mm band of muscle. A well-developed ventriculo-infundibular fold separated the semilunar and mitral valves though it was only 2 to 3 mm wide below the aortic valve. The pulmonary outflow tract was mildly stenotic at its origin.

**Discussion**

According to the observations of Goor and Lillehei (1975) and their interpretation of previous embryological descriptions, conotruncal inversion occurs in two stages. Firstly, the conoventricular junction (ostium bulbi) rotates $110^\circ$ counterclockwise (looking downstream) during d-looping of the cardiac tube. This process transfers the proximal aortic
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Fig. 2 Photograph of right ventricle (RV). The anterior wall of the right ventricle has been removed. The ventricular septal defect (VSD) is bounded by the posterior limb of the trabecula septomarginalis (PLTSM), aortic valve (AV), and anterior ventricular septum (AVS).

Fig. 3a Closer view of Fig. 2. Marker through VSD. AO, aorta; TV, tricuspid valve.

Fig. 3b Diagrammatic representation of relation between infundibular septum (IS) and trabecula septomarginalis (TSM). AO, aorta; PL, posterior limb; AL, anterior limb; TV, tricuspid valve.
Incomplete absorption of the subaortic ventriculo-infundibular fold occurred in our case with consequent mitral-aortic fibrous discontinuity. Concomitantly, failure of truncal inversion resulted in the aortic valve and aorta remaining dextroposed and originating completely from the right ventricle.

Although 'posterior' transposition has been previously documented (Van Praagh et al., 1971; Wilkinson et al., 1975), our case is distinctive in that, at semilunar valve level, the aorta was anterior to the pulmonary artery.

References


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