'Posterior' transposition reconsidered

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Seven cases of transposition with an unusually posterior aorta and mitral-aortic fibrous continuity are described. The cases are similar to four cases published by Van Praagh et al. in 1971. The use of the term 'p' transposition is suggested to distinguish this unusual form of transposition from cases with a subaortic conus ('d' and 'l' transposition). The use of the term 'transposition' is considered justifiable on both semantic and morphogenetic grounds and a possible common embryological mechanism for all forms of transposition is discussed.

In a recent study, Van Praagh and colleagues recorded the details of 4 cases of a previously undocumented cardiac anomaly, which they termed 'Transposition of the great arteries with posterior aorta, anterior pulmonary artery, sub-pulmonary conus and fibrous continuity between aortic and atrioventricular valves' (Van Praagh *et al.*, 1971). Goor and Edwards (1973) also referred to 6 cases of 'd' transposition with posterior aorta, but did not describe them in detail. Van Praagh *et al.* noted that the anomaly did not show all the features which they had formerly regarded as necessary for the application of the term 'transposition', but they concluded none the less that this was a form of transposition and they redefined the term 'transposition' to include the new cases. This radical revision of the nomenclature was criticized on the grounds of the extreme rarity of such cases (Van Mierop, 1971).

During our studies of the cardiopathological collections at the Royal Liverpool and the Birmingham Children's Hospitals, which contain over 2000 specimens, we have encountered 7 further hearts which exhibit the same or closely related abnormalities. In view of the controversy at present surrounding the anomaly, we consider it desirable to document the features of our cases and to review the arguments and conclusions relating to the nomenclature and morphogenesis.

Method

Pathological specimens were studied macroscopically in all cases. Two cases were selected for histological study, particularly in relation to the conducting tissue. A block was taken from the ventricular septum including the area of the ventricular septal defect, the roots of the great arteries, and the distal part of the interatrial

septum. The blocks were embedded in paraffin wax and sectioned in the frontal plane at 10 μ thickness. One section in every 50 was mounted and stained with Masson's trichrome technique; further sections were mounted and stained where indicated after examination of the initial ones.

Clinical data

The clinical features, electrocardiographic, and radiological findings are summarized in Table 1. All except Case 6 had severe heart failure and evidence of pulmonary plethora on the chest x-ray. Case 6 was distinct in that there was no plethora and the clinical course was one of recurrent cyanotic attacks. This child had severe pulmonary stenosis at necropsy. Electrocardiograms showed right atrial hypertrophy in 3 cases (43%), left ventricular hypertrophy in 2 (29%), and biventricular hypertrophy in 1 (14%) (Table 1). The mean frontal QRS axis varied from +60° to +120° in 6 cases, which is approximately the range seen in other examples of transposition with a ventricular septal defect (Burch and De Pasquale, 1967). The electrocardiogram in Case 6 was not available for study, but was reported as showing 'right axis deviation'.

The angiographic features are different from those seen in most patients with transposition and a ventricular septal defect and will be published separately. It is notable that the aortic root, as seen in the lateral projection, lies considerably more posteriorly than in 'classic transposition' (Van Praagh *et al.*, 1971) (Fig. 1). In Case 4 this led to an erroneous angiographic diagnosis of pulmonary atresia.

In Case 7, who was admitted while this study was already in preparation, the initial diagnosis from venous angiography was of double outlet right

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TABLE I Clinical, electrocardiographic, and radiological findings

Case No.	Sex	Age at death	Clinical features				Electrocardiogram			Radiology	
			Systolic murmur grade	Cyanosis	Dyspnoea	Liver enlargement	QRS axis (frontal)	PR (s)	Other	Pulm. vasc.	Angio diagnosis
1	M	1 mth	2/4	From birth	+	+	+90°	0.12	BVH	+	Transposition
2	F	3 wk	3/4	From birth	+	+	+110°	0.10		+	—
3	F	7 wk	2/4	From birth	+	+	+60°	0.10	RA + LV +	+	Transposition
4	M	6 mth	3/4	From 7 wk	+	+	+90°	0.12	LV +	+	Pulmonary atresia
5	M	4 mth	3/4	From birth	+	+	+120°	0.12	RA +	+	Transposition
6	M	5 mth	2/4	Cyanotic attacks + from birth	+	—	'RAD'	0.12	RA +	Normal	Transposition
7	M	3 wk	2/4	From birth	+	+	+110°	0.12		+	Transposition with posterior aorta ('P' transposition)

Abbreviations: BVH=Biventricular hypertrophy; LV+=Left ventricular hypertrophy; RA+=Right atrial hypertrophy; RAD=Right axis deviation.

ventricle, but selective right and left ventricular angiograms enabled the correct diagnosis of transposition with posterior aorta to be reached before death. This we believe to be the first case of this anomaly diagnosed *ante mortem* and confirmed at necropsy.

Pathological findings

The anatomical data are summarized in Table 2. All

cases had left-sided hearts with situs solitus. The ventricular morphology was typical of dextro ('d') looping, and there were no major atrioventricular valve abnormalities. With the exception of Case 6, which will be described separately, all the hearts showed similar features as detailed below. In addition, Case 5 had mild subpulmonary stenosis and a bicuspid pulmonary valve.

Externally the pulmonary artery was anterior and

FIG. 1 a) Lateral frame of selective right ventricular angiogram from Case 3 showing posterior aorta. Pulmonary artery faintly opacified via ventricular septal defect. b) Line diagram drawn from this frame. Ao, aorta; PA, pulmonary artery; RV, right ventricle. c) Anteroposterior frame of same angiogram.

TABLE 2 *Anatomical data on cases with situs solitus, a d-type ventricular loop, and aortic-mitral continuity**

Case No.	Size of VSD (mm)	Great arteries*	Subpulmonary conus	Associated anomalies	Origin of major coronary branches		
					Right M	Circumflex	Ant. desc.
1	5	PA ant.	Complete	—	R	L	L
2†	4	S by S	Complete	—	R	L	R and (small L)
3	7	PA ant.	Complete	—	R	L	R
4	3	S by S	Complete but short	—	R	L	R and (small L)
5†	6	PA ant.	Complete and mild stenosis	Bicuspid pulmonary valve	R	L	L and R
6	8	PA ant.	Complete and severe stenosis	Bicuspid pulmonary stenosis	R	L	L
7	5	PA ant.	Complete	—	R	L	L

S by S=side by side.

Coronary arteries: R, right coronary artery (arising from anterior coronary cusp); L, left coronary artery (arising from posterior coronary cusp); Right M=right marginal branch; Ant. desc.=anterior descending branch.

* Aortic-mitral continuity, assessed grossly, was present in all cases.

† Histology confirmed aortic-mitral continuity.

to the left of the aorta in 5 cases and the vessels were side by side in 2. Internally the aortic valve was seen to be lower than is usual in transposition and partially overriding a small ventricular septal defect. There was macroscopic evidence of mitral-aortic continuity through the ventricular septal defect in all cases. The pulmonary artery arose anteriorly from the left ventricle and possessed a complete muscular conus. The pulmonary and mitral valves were separated by a bar of muscle which formed the posterior part of the subpulmonary conus (see Fig. 2). The pulmonary valve was higher than the aortic valve in all but one case, in which the valves were level. Aortic-tricuspid fibrous continuity was not present in any of the 7 cases.

The coronary arteries were normal in distribution in 3 cases. In 3 cases the circumflex and right marginal branches arose normally, but there were two anterior descending arteries, the major one arising from the right coronary. In one case the anterior descending and right marginal arteries arose from the right coronary artery and only the circumflex arose from the left coronary artery.

Case 3 showed certain additional features which were of particular interest. Immediately below the pulmonary valve, in the left ventricle, a small blind-ended sinus in the septum passed downwards and to the right towards the apex of the right ventricle. On the right aspect of the ventricular septum, anterior to the septomarginal trabecula, a second blind-ended sinus passed upward and to the right towards the first. These sinuses seemed to us to

demarcate the line of fusion between conus septum and ventricular septum (Fig. 3). Similar features, though less well marked, were present in Case 7.

Case 6 was different in that the pulmonary outflow tract from the left ventricle was very stenosed (Fig. 4). The pulmonary artery was two-thirds the diameter of the aorta and lay anteriorly and to the left. The pulmonary outflow tract was 1 cm in length and 0.4 cm in diameter and was orientated superiorly and to the right so that the bicuspid pulmonary valve lay above the ventricular septum. A small blind-ended sinus was present in the right side of the septum, anterior to the septomarginal trabecula, and pointing up towards the pulmonary artery. The posteriorly placed aorta overrode a ventricular septal defect but arose predominantly from the right ventricle.

Histological findings

Examination of the conducting tissue was undertaken in Cases 2 and 5. In both cases, the bundle of His ran along the posteroinferior margin of the ventricular septal defect before dividing into left and right bundle-branches. The distribution of the main bundles was normal.

The presence of fibrous mitral-aortic continuity was confirmed in both cases (Fig. 5a).

Sections taken in Case 2 through the anterior part of the ventricular septum below the pulmonary artery, showed that a clear line of demarcation existed between the ventricular septum and the muscle which comprised the right wall of the sub-

FIG. 2 Heart of Case 2. Abbreviations: Ao, aorta; PA, pulmonary artery; CVF (R), right part of conoventricular flange; VSD, ventricular septal defect; AoV, aortic valve; TV, tricuspid valve; MV, mitral valve; CVF (L), left part of conoventricular flange. a) Anterior view showing relatively normally related great arteries. b) Right ventricle opened. c) Left ventricle opened. Probe in ventricular septal defect. Note conoventricular flange forming prominent bar of muscle between mitral and pulmonary valves. d) View of heart from below with the apices of both ventricles removed. Probe through ventricular septal defect. Note conal muscle (conoventricular flange) anterior to both mitral and tricuspid valves.

pulmonary conus. This conal muscle extended down into the right ventricle, overlapping the ventricular septum, with which it was fused, and lying as it did between aortic and pulmonary roots, comprised the conus septum (Fig. 5b).

Discussion

The morphological feature of the 7 hearts described are similar to those documented by Van Praagh and his co-workers (1971). Haemodynamically these hearts function as 'classic complete transposition', with a ventricular septal defect. On the other hand their anatomical features distinguish them from the

vast majority of hearts with transposition of the great arteries. The distinguishing features are: a complete subpulmonary conus above the left ventricle, an unusually posterior origin of the aorta from the right ventricle, and fibrous mitral-aortic continuity through a ventricular septal defect. Before considering these morphological features in greater detail we will review briefly the problem of nomenclature and classification.

Definitions of the term 'transposition' are many and varied. They have been excellently reviewed by Harris and Farber (1939) and more recently by Van Praagh, Vlad, and Keith (1967). Most workers have stressed as important diagnostic criteria either

FIG. 3 Heart of Case 3. Abbreviations: As for Fig. 1. SRV, sinus in right ventricle; SLV, sinus in left ventricle (see text). a) Anterior view. b) Right ventricle opened showing sutured ventricular septal defect. c) Left ventricle opened; note subpulmonary sinus in ventricular septum (SLV). d) Right ventricle with sutures removed from ventricular septal defect (probe in ventricular septal defect). Note sinus in right side of ventricular septum (SRV).

anteroposterior reversal in the relation of the aorta and pulmonary artery (Harris and Farber, 1939; Cardell, 1956; Shaher, 1964), or the presence of a subaortic conus (Van Praagh *et al.*, 1967). The possibility of reversal of the relation of the vessels to the ventricles without either a subaortic conus or anteroposterior reversal has not been included in

such definitions largely, we believe, because it had not previously been encountered. None the less the definition used by Abbott (1927) does allow for such cases, viz: 'Any alteration in the position of the two great vessels relative to the ventricles of the heart, or to each other at their origin, so that they either spring from reversed ventricles, the aorta from the

FIG. 4 Heart of Case 6. Abbreviations: VSD, ventricular septal defect; POT, pulmonary outflow tract. a) Anterior view. b) Right ventricle opened. Note prominent ridge of muscle (right part of conoventricular flange) separating tricuspid and aortic valves. c) Left ventricle opened. Note very narrow opening to pulmonary outflow tract.

right and the pulmonary artery from the left chamber, or from the ventricle to which they usually belong but in reversed relationship.'

Van Praagh *et al.* (1967) stressed the presence of a muscular subaortic conus in transposition, and divided transposition into two types: 'd' transposition or 'l' transposition, depending on whether the

aorta arose to the right, or to the left, of the pulmonary artery.

However, when discussing their 4 cases of transposition with a posterior aorta, Van Praagh and his colleagues concluded that the presence of a subaortic conus could no longer be regarded as an essential criterion of 'transposition'. They, there-

FIG. 5 Histology. a) Coronal section of aortic root and ventricular septum in Case 5 passing through ventricular septal defect and showing aortic-mitral fibrous continuity. b) Oblique coronal section through aortic root and ventricular septum of Case 2 just anterior to ventricular septal defect showing clear line of demarcation between conus septum and ventricular septum. Abbreviations: Ao, aorta. RV, right ventricle; VSD, ventricular septal defect; LBB, left bundle-branch; SL, septal leaflet (of mitral valve); LV, left ventricle; VS, ventricular septum; AM, atrial myocardium; CS, conus septum; CVF (L), left part of conoventricular flange.

fore, redefined transposition to mean that both the great arteries are 'placed across' the septum – reversing the relation of the great arteries to the morphological ventricle. This definition corresponds to the earliest use of the term (Van Praagh, 1973). They introduced the terms 'd' and 'l' malposition to refer to 'any positional anomaly of the great arteries'. These latter terms thus included 'transposition' and also encompassed a great variety of conditions such as double outlet right ventricle, Taussig-Bing anomaly, and 'anatomically corrected transposition', which had previously been regarded as forms of 'transposition' but were excluded by the new definition (Van Praagh *et al.*, 1971).

This change in the nomenclature has been criticized on the grounds that the cases in point are extremely rare (Van Mierop, 1971) and scarcely justified such a revision of terminology. It was also argued that these cases were possibly not related embryologically to other forms of transposition, and should, therefore, perhaps be excluded from the term 'transposition'.

Our 7 cases suggest that the anomaly is not as rare as the small number of published cases might sug-

gest. We agree with Van Praagh *et al.* (1971) that they do represent a form of transposition, but in view of the distinctive anatomical features we consider it justifiable to apply a new term to the anomaly: 'p' transposition. This term would refer solely to the condition in which the aorta rises from the morphological right ventricle, but retains fibrous aortic-mitral continuity and the pulmonary artery arises from a muscular conus above the morphological left ventricle. The terms 'd' and 'l' transposition would thus continue to refer to cases exhibiting a complete subaortic conus.

The terms 'd' and 'l' malposition (Van Praagh *et al.*, 1971) appear at first sight to be somewhat imprecise. None the less there is a need for a simple term which may be used in conjunction with other descriptive terms to indicate the relation of the great arteries in complex malformations such as double outlet right ventricle and 'anatomically corrected transposition', where the term 'transposition' is clearly of doubtful accuracy. We would, therefore, commend the use of the terms 'd' and 'l' malposition in such instances, believing that in conjunction with a full description of cardiac anatomy they are

TABLE 3 *Uses of the terms transposition and malposition*

<i>Terms</i>	<i>Examples of some situations in which terms are both appropriate and useful</i>
'd' transposition/'l' transposition	1) Classic complete transposition 2) Classic corrected transposition 3) Single ventricle with outlet chamber
'p' transposition	Transposition with unusually posterior aorta and mitral-aortic continuity
'd' and 'l' malposition	1) Double outlet right ventricle 2) Double outlet left ventricle 3) Anatomically corrected transposition 4) Common ventricle with anterior aorta

accurate and helpful (see Table 3). This use of terms is not drastically different from the sense in which we have used them previously (Wilkinson and Acerete, 1973). It does, however, imply two modifications – namely, the introduction of 'p' transposition and the adoption of 'd' and 'l' malposition to include some examples of 'd' and 'l' transposition of the aorta, in which the pulmonary artery is not also transposed. We now restrict the use of the term 'transposition' to cases in which *both* vessels are 'placed across the septum' (Van Praagh *et al.*, 1971; Van Praagh, 1973).

Unlike Van Mierop (1971) who considered that transposition with a posterior aorta, as presently described, was unrelated to classic 'transposition', we believe that a single morphogenetic concept can account for all forms of transposition (Goor and Edwards, 1973; Anderson *et al.*, 1974a, b). Before considering it further, however, it is necessary to discuss the anatomical features in greater detail.

In the ensuing discussion the term 'p' transposition refers to the presently described cases and classic transposition refers to 'classic complete transposition'.

In 'p' transposition the main anatomical features, which distinguish it from classic transposition, are the presence of a complete subpulmonary conus and the absence of a complete subaortic conus.

In the normal heart the subpulmonary conus is composed of conus septum posteriorly and to the right, anterior interventricular septum to the left, and parietal wall of the infundibulum anteriorly and to the right (see Fig. 6a). The crista supraventricularis, which separates the pulmonary and tricuspid valves, is formed in part by conus septum (Van Mierop *et al.*, 1963; Los, 1968; Goor, Dische, and Lillehei, 1972) and also by an area of right ventricular wall – the anterior tricuspid ledge, which is derived from the right part of the conoventricular flange (Lubkiewicz, 1969; Anderson and Ashley, 1974) and which is referred to by Van Praagh as 'conal free wall' (R. Van Praagh, 1973, personal communication).

In 'p' transposition the subpulmonary conus is composed of conus septum to the right and posteriorly, left ventricular free wall anteriorly, and a bar of muscle posteriorly and to the left which separates the mitral and pulmonary valves. This bar of muscle was referred to by Van Praagh and his colleagues as 'a left ventricular crista supraventricularis' (Fig. 2 and 6). It is probable that this muscle is embryologically derived from the left part of the conoventricular flange but, as it is separate from the conus septum, it is not analogous with the normal crista.

The normal subaortic conus consists of the muscular conus septum anteriorly and a small area of muscular conoventricular flange anterior and to the left. The remainder of the subaortic conus is attenuated and fibrous and consists of left fibrous trigone, septal cusp of mitral valve, central fibrous body, and membranous septum (see Fig. 6a). This fibrous area is formed by absorption of the central portion of the conoventricular flange, which serves to bring the aorta over the left ventricle and away from its embryonic position over the bulbus cordis (Goor *et al.*, 1972; Anderson and Ashley, 1974).

In 'p' transposition the aortic conus also has muscular and fibrous portions. The muscular parts comprise the conus septum to the left, the right part of the conoventricular flange posteriorly, and parietal wall of the right ventricle anteriorly. The fibrous part lies posteriorly and to the left, and here the aortic root is in fibrous continuity with the mitral valve via the central fibrous body (Fig. 6b).

As will be apparent from this description, the orientation of the conus septum in 'p' transposition is sagittal instead of transverse as in the normal heart (see Fig. 6a and b), and anteriorly it is fused with the ventricular septum, thus separating the pulmonary artery from the right ventricle.

In order to relate these anatomical features to morphogenesis, we must first consider certain events in normal cardiac embryology.

After bending of the heart tube the truncus has been shown to arise from the bulbus. As the bulbar

FIG. 6 Diagrammatic representation of cross-section through the valve rings with conus septum and conoventricular flange superimposed. a) Normal heart. b) 'p' transposition. c) Classic 'd' transposition. Abbreviations: CVF, conoventricular flange; Crista SV, crista supra-ventricularis; Ao, aorta; PA, pulmonary artery.

ridges form they divide the distal bulbus into an anterior outflow tract to the pulmonary artery and a posterior path to the aorta (Van Mierop *et al.*, 1963; Los, 1968; Goor *et al.*, 1972; Anderson *et al.*, 1974b). Furthermore, at this stage the aorta is separated from the left (primitive) ventricle and the atrioventricular canal by the conoventricular flange (Van Mierop *et al.*, 1963; Anderson *et al.*, 1974b). As the bulbus is septated the aortic component migrates posteriorly over the left ventricle and the middle portion of the conoventricular flange is absorbed (Goor *et al.*, 1972; Anderson *et al.*, 1974b). Attenuation of this area results in shortening of the aortic conus and the formation of the fibrous conus and fibrous trigones.

During abnormal development Van Mierop and Wigglesworth (1963a) suggested that absorption of the conoventricular flange might be incomplete, as in double outlet right ventricle. They also proposed that malorientation of the conus septum was a feature of Fallot's tetralogy and double outlet right ventricle. On the other hand, they proposed straight fusion of the truncal ridges as the cause of transposition (Van Mierop and Wigglesworth 1963b). Van Praagh and his colleagues have criticized this latter suggestion on the grounds that it does not explain the conal morphology in transposition. They, in

turn, originally proposed abnormal 'differential conal growth' as an explanation of transpositions, finding little evidence to support the rather similar theory of differential conal absorption (Keith, 1909; Paul, Van Praagh, and Van Praagh, 1968). Recent studies, however, lend strong support to the latter concept (Goor *et al.*, 1972; Anderson *et al.*, 1974a, b).

The common anatomical feature of 'p' transposition and of classic transposition is the origin of the pulmonary artery from the left ventricle. In normal development, the pulmonary artery is firmly walled off from the left ventricle from an early embryonic stage. This results from the fusion of the sinistro-ventral conal ridge, which underlies the left part of the conoventricular flange, with the anterior inter-ventricular septum. Deficiency of this ridge may allow the pulmonary artery to override the left ventricle. Malorientation of the conal ridges would similarly allow migration of the pulmonary artery over the left ventricle, and later fusion of conus septum with ventricular septum would separate the pulmonary artery completely from the right ventricle. Such a concept, which is a logical extension of Van Mierop and Wigglesworth's (1963a) views on development of double outlet right ventricle, allows for a spectrum of abnormalities, including

double outlet right ventricle, Taussig-Bing anomaly, and complete transposition depending on the degree of malorientation of the conus septum, the extent of absorption of the conoventricular flange, and the site at which fusion of conus septum with conoventricular flange or ventricular septum takes place (Goor and Edwards, 1973; Anderson *et al.*, 1974a). 'p' transposition, while not part of this spectrum, is closely related and, in our view, similarly produced. The essential abnormalities are malorientation of the conus septum and malfusion of the conus septum with the ventricular septum. The presence of a posterior aorta is, we would postulate, related to absorption of the subaortic rather than the subpulmonary conus. This absorption is only partial and produces mitral-aortic continuity while the aorta remains above the right ventricle, as in some forms of extreme Fallot's tetralogy. However, it is a logical extension of this hypothesis that more complete absorption of the subaortic conus would produce a form of double outlet left ventricle.

The mechanism of conal absorption and the factors determining which conus is absorbed are not known. Though the orientation of the outflow tracts at the time of their septation and the form of the bulboventricular loop are important, there may be other factors, less obvious morphologically, which have an important bearing on this problem.

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