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Terminological pitfalls in congenital heart disease

Reappraisal of some confusing terms, with an account of a simplified system of basic nomenclature

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A simplified system of basic nomenclature for congenitally abnormal hearts is outlined. Certain abnormalities in which terminology has had a tendency to be confusing or ambiguous are discussed and ways in which clarity of definition can be maintained are described.

By this system almost all congenitally abnormal hearts can be described fully and accurately, using simple and generally acceptable terms.

During the early part of an analysis of the collection of over 1,000 congenitally abnormal hearts at the Children's Hospital, Birmingham, in 1968, it became apparent to us that the terms in current use by clinicians and pathologists for many cardiac anomalies were ill understood and ambiguous. In order to achieve conformity between ourselves, and to make it easier for others to understand the terms we used, we set out to devise a simplified system of nomenclature for the various abnormalities which seemed to us to present particular problems.

Many previous attempts have been made to do just this (Arcilla and Gasul, 1961; Daves and Pryor, 1970; Lev, 1954; Lev *et al.*, 1968; Rosenbaum, 1964; Van Praagh *et al.*, 1964). None has been wholly successful in resolving the difficulties and the reason for this seems to be that each author or group of authors has continued to use confusing terms which are open to misinterpretation and has sought to render them less ambiguous only by applying to them yet another 'definition' which, though clear to them, has not been acceptable to many others who were familiar with different and earlier definitions of the same terms.

We have therefore attempted to approach the problem from a different angle. We have sought in this system of nomenclature to avoid entirely the use of terms which are ambiguous. Not surprisingly this has not been possible in all respects but when we have felt bound to adhere to terms such as 'corrected transposition', which are so widely used that we could not reject them completely, we have sought to use the most widely acceptable definitions that would fit in with the rest of our nomenclature. In particular our basic policy has been to adhere to the simplest and least ambiguous terms available and to avoid the use of new terms.

Little of this can be claimed to represent original ideas, rather we regard it as a review of terminology with the object of 'tidying up' an extremely difficult and confusing subject.

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Sources and methods

The terms used were chosen by reference to the available published reports and at no time have we used terms in ways in which they have not been widely used previously. We have to a large extent avoided the use of more complex terminology where we think that simple terms can be used to describe the same abnormality.

Necessarily this has meant that some of our terms are less succinct than alternatives used by others (for instance, 'morphological right ventricle' is less succinct than 'dextral') (Rosenbaum, 1964). By the same token however, we believe that our terminology is less ambiguous and hence more easily understandable to other workers in the field.

A comprehensive review of the published material is beyond the scope of this paper. To discuss the relevant arguments at sufficient length to do justice to them would take so much time and space that it would detract from the main purpose, which is to present a clear, concise, and simple account of the ways in which we have attempted

to avoid the terminological pitfalls of congenital heart disease. For this reason lengthy discussion is avoided, and brief reference only is made to the reports in which the full arguments are to be found.

Fundamentals

Classification of congenital abnormalities of the heart depends upon the establishment and clear expression of all the abnormal features of cardiac anatomy. There are obviously considerable limitations in the extent to which certain abnormalities can be assessed in the clinical situation and more precise delineation of anatomy may have to await surgery or even necropsy. None the less any system of classification should seek to present as full a picture as possible, with the available facts.

We feel that in describing any given heart the following facts should be made clear (see Table 1).

1) The presence of laevocardia or dextrocardia.
2) The situs of the viscera and atria (solitus, inversus, indeterminate) (Van Praagh *et al.*, 1964).
3) Type of ventricular loop ('d', 'l', or 'x' loop) (Van Praagh *et al.*, 1964).
4) The relation of the great vessels: (a) normal; (b) inverted; (c) 'd' transposition (Van Praagh *et al.*, 1964); (d) 'l' transposition (Van Praagh *et al.*, 1964); and (e) dextroposition or laevoposition of one or other great vessel.
5) The chamber of origin of the great vessels.
6) The anatomy of the subaortic and subpulmonary conuses (Van Praagh and Van Praagh, 1966).

The above list will adequately define the relation of the chambers and vessels in the great majority of hearts. Clearly the presence of other anomalies, such as atrial or ventricular septal defects, AV canal, abnormalities and atresia of the various valves, persistent truncus arteriosus, and anomalies of the venous return must be defined separately to complete the anatomical description.

TABLE 1 *Facts necessary in describing relations of cardiac chambers and great arteries*

Fact to be established	*Acceptable terms*	
	Normal	*Abnormal*
1) Position of heart	Laevocardia	Dextrocardia/ mesocardia
2) Visceroatrial situs	Solitus	Inversus/ indeterminate
3) Ventricular relation	Non-inverted or 'd' loop	Inverted or 'l' loop/'x' loop
4) Great arteries		
a) Non-transposed	Normal	Inverted/ dextroposition/ laevoposition
b) Transposed		'd' transposition/ 'l' transposition
5) Relation of great arteries to ventricles	Aorta arising from left ventricle; pulmonary artery arising from right ventricle	Double outlet right ventricle/ double outlet left ventricle/ aorta arising from right ventricle, etc.
6) Conal anatomy	Complete subpulmonary conus Incomplete subaortic conus	Complete subaortic conus/ bilateral conus/ Incomplete subpulmonary conus/bilaterally deficient conus

Problematical terms

The abnormalities which we propose to discuss in detail are those which have been associated with terminological pitfalls and include: 1) dextrocardia; 2) inversion; 3) transposition; 4) corrected transposition; 5) dextroposition and laevoposition; 6) persistent truncus arteriosus; 7) bilateral conus anomaly; 8) double-outlet right ventricle; and 9) Taussig-Bing anomaly.

1) Dextrocardia

Dextrocardia is most commonly understood to mean a heart lying predominantly in the right hemithorax. A heart lying in the left side of the chest is similarly referred to by the term 'laevocardia', and a heart lying in the centre of the chest 'mesocardia' (Van Praagh and Vlad, 1967).

The direction of the ventricular apex may need to be specified separately as it is not necessarily the same as the side of the chest in which the heart lies. It is not, however, our practice to comment on the direction of the apex unless it differs from that expected.

Inversion of the atria or ventricles and alterations in the position of the great vessels must, of course, be noted in addition (see below 'inversion').

Variations in the position of the heart or the direction of the apex can thus easily be distinguished from major alterations in the relation of the chambers or great vessels (Stanger *et al.*, 1968).

These are the only terms which we consider essential to the terminology of dextrocardia. They are simple and unambiguous and we do not believe that the use of more complicated definitions or additional terms is either necessary or desirable (Table 2).

Terms which we prefer to avoid *Dextroposition of the heart* means that a normal heart has been shifted to the right by extracardiac factors

TABLE 2 *Terms relating to dextrocardia and inversion*

	Acceptable terms	*Terms which tend to be confusing*
Dextrocardia	Dextrocardia	Dextroversion/laevoversion
	Mesocardia	Secondary dextrocardia
	Laevocardia	Extrinsic dextrocardia
		Dextroposition
		Dextrorotation
		Mixed dextrocardia
		Isolated dextrocardia
		Pivotal dextrocardia
Inversion		
Ventricles	'd' loop (non-inverted)	
	'l' loop (inverted)	
	'x' loop	
Visceroatrial situs	Solitus	
	Inversus	
	Indeterminate	
Morphology of chambers	Morphological right	Dextral/'right'
	Morphological left	Sinistral/'left'

(Grant, 1958). The term is generally regarded as synonymous with 'secondary dextrocardia' and 'extrinsic dextrocardia' (Harris and Farber, 1939). We consider that it is possible to avoid the use of all these terms without losing clarity.

Dextroversion. This term has been used in many different ways by different authors (Daves and Pryor, 1970; Grant, 1958; Harris and Farber, 1939; Lev *et al.*, 1968; Van Praagh *et al.*, 1964), and again we prefer not to use it at all. (See discussion by Van Praagh *et al.*, 1964.)

'Isolated dextrocardia' is a term which has been used to describe the forms of dextrocardia which are associated with viscero-atrial situs solitus. We feel that the term adds little, if anything, to the terminology of 'dextrocardia', and as it is not generally used we feel that a full description of the position of the viscera, atria, ventricles, and great vessels is more likely to be universally understood.

Dextrorotation (De la Cruz *et al.*, 1959; Gasul, Arcilla, and Lev, 1966), *pivotal dextrocardia* (Lev, 1954), *and mixed dextrocardia* (Daves and Pryor, 1970; Grant, 1958; Lev, 1954; Lev and Rowlatt, 1961). Like dextroversion and isolated dextrocardia, these terms are not universally understood and tend to complicate unnecessarily the terminology of dextrocardia.

Mirror-image dextrocardia. This term is used to refer to dextrocardia with visceroatrial situs inversus. It is most frequently used when the heart is functionally normal and has inverted ventricles and great arteries. Some clinicians and authors use the terms more loosely, however, and include hearts with a variety of other congenital anomalies. It is probably preferable to avoid the term altogether.

2) Inversion

Inversion refers to a reversal in the lateral relation of structures, thus implying that normally right-sided structures are left-sided, and normally left-sided structures are on the right. The term may be used for changes in the arrangements of the viscera, atria, ventricles, or great vessels.

a) Atria Inversion of the atria implies a reversal of the lateral relations of the atria. In order to understand this, it is necessary to appreciate those anatomical characteristics of each chamber, which are relatively constant, even in the presence of septal defects, venous anomalies, or dextrocardia, etc.

The normal right atrium is characterized by:

1) The limbus of the fossa ovalis.
2) The opening of the coronary sinus.
3) Several small orifices for the entry of thebesian veins just above the limbus.
4) The opening of the inferior vena cava.
5) The atrial appendage has a wide opening into the atrium, the blind end of the appendage being broad with a few ill-defined lobulations.

The atrium having these characteristics is called the 'morphological right atrium'.

The normal left atrium is recognized by:

1) The presence of the 'septum primum'. This delicate septum is attached to the septum secundum by a variable number of fibrous strands at its cranial margin on the interatrial wall opposite to the fossa ovalis.
2) This atrium has a smoother parietal wall lining than that of the right atrium.
3) The atrial appendage has a narrow opening from the atrial cavity and the appendage is a slender structure with many clearly defined lobulations and a sharp end.

The atrium having these characteristics may be called the 'morphological left atrium'.

Inversion of the atria may therefore be defined as the situation in which the left-sided atrium has the characteristics of a morphological right atrium and the right-sided atrium has those of a morphological left atrium.

The atrial position is independent of the position of the ventricles or the great vessels, but it is always

related to the visceral situs (Van Praagh *et al.*, 1964), thus:

1) *Visceroatrial situs solitus* is the usual, hence normal, placement of atria with the liver on the right side and the spleen and stomach on the left.
2) *Visceroatrial situs inversus* is the apparent mirror-image of situs solitus with atrial inversion, liver on the left, and stomach and spleen on the right.
3) *Visceroatrial situs uncertain or indeterminate* is characterized by uncertain anatomical arrangements of the atria with visceral heterotaxy and an abnormal degree of bilateral symmetry. Asplenia or polysplenia is present in over 80 per cent of cases. Many cases of laevocardia with a right-sided stomach and dextrocardia with a left-sided stomach are examples of this 'syndrome' (Ivemark, 1955; Keith, Rowe, and Vlad, 1967).

b) Ventricles The normal right ventricle is characterized by:

1) A coarse trabecular interior (particularly the septal surface).
2) The presence of a crista supraventricularis.
3) The presence of a trabecular septo-marginalis with one or more septal papillary muscles.
4) A well-defined muscular outflow tract (infundibulum).
5) A tricuspid atrioventricular valve (*N.B.*: the normal tricuspid valve not infrequently only has two cusps).
6) The trabeculae are separated by deep troughs extending almost to the epicardium.

The ventricle having these characteristics may be called the 'morphological right ventricle'.

Similarly the left ventricle is characterized by:

1) Smooth septal surface.
2) Absence of crista supraventricularis.
3) Absence of septal papillary muscles.
4) Absence of a muscular outflow tract.
5) A bicuspid AV valve.
6) The AV valve is in fibrous continuity with the aortic root in the absence of transposition.
7) The trabeculae of the left ventricle are separated by shallow troughs which do not extend deep into the myocardium.

The ventricle having these characteristics may be called 'morphological left ventricle'.

The 'morphological right ventricle' is embryologically derived from the 'bulbus cordis' and the 'morphological left ventricle' from the primitive 'ventricle' of the heart tube (Davis, 1927; De Vries and Saunders, 1962) (see Fig. 1).

Inversion of the ventricles is present when the right-sided ventricle has the characteristics of a 'morphological left ventricle' and the left-sided ventricle those of a 'morphological right ventricle'.

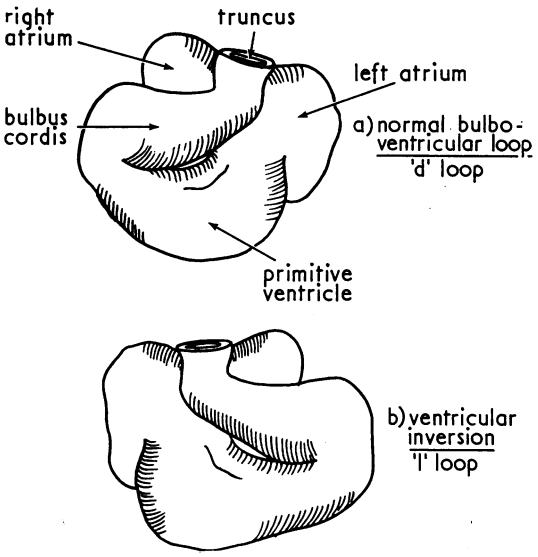

FIG. 1 *Diagram showing normal and inverted ventricular loop of embryo heart at approximately 4·5 mm stage.*

It should be emphasized that in many abnormalities associated with ventricular inversion (and some without), several of the features regarded as characteristic of ventricular morphology may be ill defined and ambiguous. None the less in the vast majority of hearts the anatomy is sufficiently clear to allow a decision to be made as to whether ventricular inversion is present or absent.

c) Inversion of the great arteries This refers to reversal in the lateral relation of the great vessels *without* transposition. Thus the aorta lies posterior and to the *left* of the pulmonary artery (Fig. 2) (Van Praagh *et al.*, 1964).

Inversion of the great arteries is the 'normal' position in dextrocardia with visceroatrial situs inversus (see discussion of 'mirror image dextrocardia', page 1168).

d) Other terms relating to 'inversion' *Ventricular loop* Normally the heart tube forms with the primitive ventricle lying towards the left and the bulbus cordis to the right (Davis, 1927; De Vries and Saunders, 1962). Such a 'loop' is termed a 'd' loop (Van Praagh *et al.*, 1964), i.e. with the morphological right ventricle to the right. When the loop is 'inverted' the 'morphological right ventricle'

lies to the left and this may be termed an 'l' (laevo) loop (see Fig. 1).

It may be seen clearly therefore that in a heart with a 'd' loop the morphological right ventricle lies to the right of the morphological left ventricle, as in a normal heart, and in a heart with an 'l' loop the morphological right ventricle lies to the left of the morphological left ventricle, i.e. the ventricles are 'inverted'.

The term 'ventricular inversion' is thus synonymous with an 'l' loop.

The use of the term 'd' and 'l' loops refer solely to embryological considerations of the bulboventricular area of the heart. The atria and the great vessels must be regarded independently as they do not necessarily follow the pattern that might be supposed from the relation of the ventricles.

In cases in which it is impossible to differentiate with certainty which ventricle is which on morphological grounds and in cases where the ventricles lie in a completely anteroposterior relation the 'loop' is called an 'x' loop. This merely means that the direction of the loop is unknown (Van Praagh *et al.*, 1964).

Concordance and discordance The relation between the atria and the ventricles may be understood clearly by the use of the terms 'concordance' and 'discordance' of the cardiac loop (Rosenbaum, 1964; Van Praagh *et al.*, 1964). In the presence of a concordant cardiac loop the morphological right atrium opens into the morphological right ventricle and the morphological left atrium opens into the morphological left ventricle. A discordant loop indicates that the morphological right atrium opens into the morphological left ventricle and the morphological left atrium into the morphological right ventricle.

It may be seen that in the presence of a concordant loop the atria and ventricles are either both 'inverted' or else neither is inverted. In a discordant loop there is always inversion of *either* the ventricles *or* the atria but never both.

Conclusion The terms so far discussed in this section – 'inversion', 'd' and 'l' loops, 'concordance' and 'discordance' – are all sufficiently simple and useful to have a place. Though the terms overlap to a certain extent all are sufficiently unambiguous and explicit to be of value (see Table 2).

Terms which we prefer to avoid *'Dextral' and 'sinistral':* these terms have been used to provide a short alternative for the terms 'morphological right' and 'morphological left', in relation to the atria or ventricles (Rosenbaum, 1964). For those who can accept the use of these terms in this way they provide a useful means of avoiding the words 'right' and 'left', when discussing the morphology of the ventricles or atria. In Rosenbaum's terminology the words 'right' and 'left' thus refer solely to anatomical position. These terms are not widely accepted or understood and, for this reason, we prefer not to use them.

'Left' and 'right'. Some workers in discussing inversion have used the words 'left' and 'right' on their own to refer to the morphological characteristics of chambers, rather than their anatomical position. This increases the ambiguity by allowing such phrases as: 'the pulmonary veins drained into the *left* atrium', without further indication of whether *left* refers to morphology or position. The only advantage of using the words thus is brevity and this we feel is far less important than clarity.

3) Transposition[1]

Several terms have come to be used in relation to transposition. These are: *complete; inverted; partial; corrected* (a) *anatomically*, (b) *physiologically; 'd' transposition; and 'l' transposition.*

Various definitions have been proposed before for the term 'transposition' and it is difficult to suggest any clear and succinct usage which will be universally acceptable.

The term has been used most frequently to refer to anomalies in which the aortic root is abnormally placed (see below), but the position of the pulmonary artery, relative to the aorta or to the ventricles, has not been regarded as important in deciding whether transposition is present or not. Though it is debatable whether this usage is terminologically precise enough or etymologically accurate (Van Praagh *et al.*, 1971), it seems to us that the most widely acceptable and the most useful way of defining the term is the conventional one, which follows.

Basically *'transposition'* refers to an abnormality of the development of the bulbo-truncal areas such that the aorta arises in an area removed from its normal site of origin in relation to the mitral valve. The most characteristic feature of the anomaly, pathologically, is loss of the fibrous continuity which normally exists between the mitral valve and the root of the aorta (see discussion on 'double outlet RV' and 'bilateral conus anomaly').

Some descriptions of the anomaly have stressed a reversal of the normal anteroposterior relation of the aorta and pulmonary artery.

In fact, while a change in this relation is universal, the aorta does not necessarily lie *anterior* to the pulmonary artery, but may lie lateral to it (or possibly even slightly posterior). The essential change is

[1] In the following discussion we use the terms 'aorta' and 'pulmonary artery' to refer to the position of the root of the vessels.

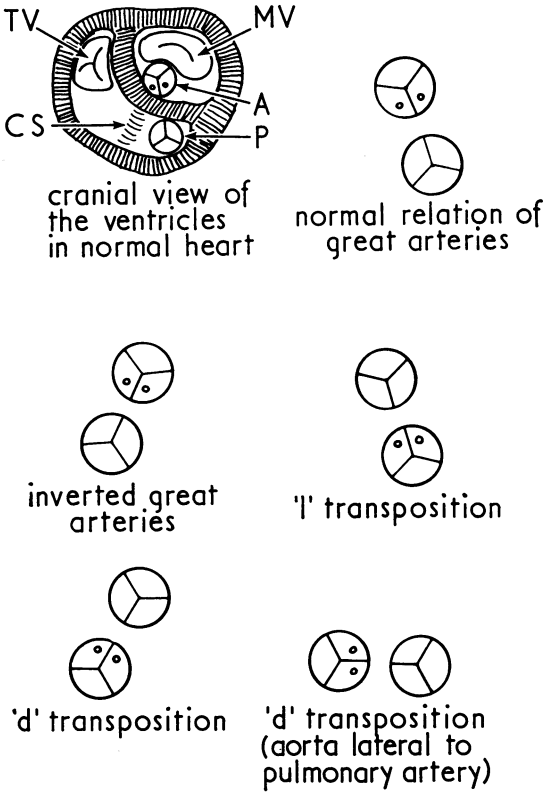

FIG. 2 *Diagram of the relation of the great arterial in the normal heart, hearts with inverted great arterial and in various forms of transposition.*

that the aorta is separated from the mitral valve by muscle which forms part of a ring of muscle or 'subaortic conus' from which the aorta arises. In most cases the presence of this 'conus' carries the aorta anteriorly so that it lies lateral or anterior to the pulmonary artery. The ultimate relation of the great vessels depends, however, on the site of origin of the pulmonary artery and the manner in which the heart lies in the chest.

There are thus three characteristic pathological features of transposition.

1) Loss of fibrous continuity between the aorta and mitral valve.
2) A muscular conus around the aortic root.[1]
3) A *change* in the anteroposterior relation of the great vessels with the aorta lying lateral or anterior to the pulmonary artery.

[1] This muscle forms a complete conus in all but a very few instances in which it is deficient posteriorly and allows fibrous continuity between the aorta and the tricuspid valve – a situation in which there is often a bilaterally deficient conus.

It will be seen from this that the ultimate criteria for the diagnosis of transposition are anatomical and it is true to say that some forms of transposition may be impossible to categorize with accuracy during life. None the less, a large number of cases can be distinguished during life by specific clinical features, *x*-ray, cardiac catheterization, and angiocardiography.

It should also be noted that this definition of 'transposition' is, in practice, a definition of 'transposition of the aorta' and *not* of 'transposition of the great arteries'. Indeed the latter term can only be applied with accuracy when the transposed aorta arises from the venous ventricle (right ventricle in visceroatrial situs solitus and left ventricle in visceroatrial situs inversus), and the pulmonary artery arises from the arterial ventricle. This definition is similar to, but not identical with, that currently used by Van Praagh and his co-workers (1971).

This is the situation which applies in the commonest form of transposition as shown in Fig. 3, but the forms of transposition shown in Fig. 10c and 10d (Taussig-Bing anomaly and double outlet right ventricle) are distinct and, though they show 'transposition of the aorta', they should not be regarded as examples of 'transposition of the great arteries'.

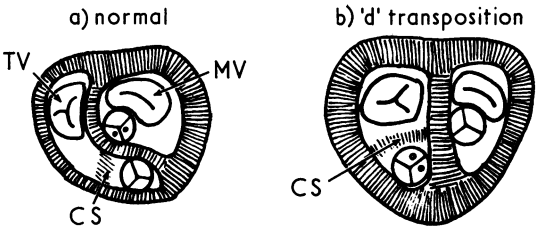

FIG. 3 *Diagram of the relations of the great arteries, AV valves, and ventricular septum in the normal heart and in the commonest transposition variant. MV, mitral valve; TV, tricuspid valve; CS, crista supraventricularis.*

According to Van Praagh and Van Praagh (1966) the common embryological feature of all forms of transposition, as defined on these lines, is the excessive growth of the subaortic conus, which thus carries the aortic root ventrally and away from the mitral valve.

Transposition thus defined has, therefore, distinct anatomical and embryological features which make the use of the term rational.

3a) Other terms relating to transposition *'d' and 'l'* For the sake of simplicity of description Van Praagh has introduced the terms 'd' trans-

position and 'l' transposition to indicate the lateral relation of the vessels in transposition (Paul, Van Praagh, and Van Praagh, 1968; Van Praagh *et al.*, 1964).

'd' transpotition is that in which the aorta lies to the *right* of the pulmonary artery, and 'l' transposition that where the aorta lies to the left of the pulmonary artery (Fig. 2).

It should be emphasized that the term 'transposition' does not in itself imply that each great vessel arises from a separate ventricle and there is a very large number of described and undescribed possibilities for the relation of the ventricles and the vessels to one another in transposition.

Terms which we prefer to avoid (Table 3) *Inverted transposition* Inverted transposition as used by Spitzer refers to any form of transposition occurring in an inverted ventricular loop (Spitzer, 1951) (see 'inversion'). It has also been used to indicate 'l' transposition in dextrocardia (Lev *et al.*, 1968) – the term is thus confusing and is better abandoned.

TABLE 3 *Terms relating to transposition and corrected transposition*

	Acceptable terms	*Terms which tend to be confusing*
Transposition	'd' transposition	Partial transposition
	'l' transposition	Inverted transposition
Corrected transposition	'Corrected transposition' – only acceptable when transposition is physiologically corrected (see text)	Anatomically corrected transposition

Incomplete or partial transposition This term is ambiguous and has been used to refer variously to Taussig-Bing anomaly, double outlet right ventricle, Fallot's tetralogy, persistent truncus, and any kind of overriding aorta. We consider that transposition is either present or absent and the term 'partial' transposition is vague and unhelpful. In the Taussig-Bing anomaly and some cases of double outlet RV, the aorta shows the features of transposition as defined previously. Fallot's tetralogy, overriding aorta, truncus, and other cases of double outlet RV, however, do not show these features (see dextroposition of aorta; truncus arteriosus; Fallot's tetralogy; double outlet RV).

4) Corrected transposition

The term 'corrected transposition' is confusing but, in view of its now generally accepted and time-honoured usage, it may be employed to a limited extent. Its use should probably be restricted to those cases of transposition which are corrected *physiologically*, i.e. in which pulmonary venous blood passes via the 'morphological left atrium' through a ventricle to a transposed aorta and systemic venous blood via the 'morphological right atrium' through another ventricle to the pulmonary artery.

There are four conditions that fit this description (Fig. 4). They may be classified as follows:

1) Situs solitus, ventricular 'l' loop, 'l' transposition.

2) Situs inversus, ventricular 'd' loop, 'd' transposition.

3) Situs solitus, ventricular 'd' loop, 'l' transposition.

4) Situs inversus, ventricular 'l' loop, 'd' transposition.

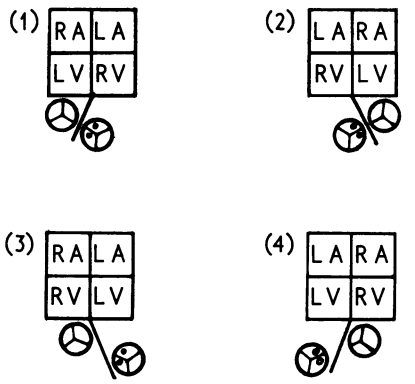

FIG. 4 *Schematic diagram showing relation of the chambers and great arteries in the four possible variants of 'corrected' transposition.* N.B. *The various forms of so-called 'anatomically corrected transposition' are not included – see text. (Adapted from Harris and Farber, 1939.)*

The presence of these four possibilities makes it clear that the term 'corrected transposition' should not be used in isolation and hence, if it is to be used at all, it must be accompanied by a full anatomical description.

It will be seen that the essential characteristics of 'corrected transposition' are the presence of 'l' transposition in situs solitus or 'd' transposition in situs inversus.

Cases where the term 'corrected transposition' should be avoided (Table 3) The presence of associated anomalies which interfere with the physiological pathways of venous and arterial blood to such an extent as to distort materially the physiological 'correction' – as defined above, preclude the use of the term 'corrected transposition' (Malers *et al.*, 1960). Thus the presence of lesions such as anomalous venous drainage, AV valve atresia, semilunar valve atresia, or large septal defects, with free mixing of arterial and venous blood will make the use of this term inaccurate (De la Cruz *et al.*, 1959). Under such circumstances the various defects must be classified individually.

The term 'anatomically corrected transposition' is not in general use and is poorly understood by many. The conditions which have been so labelled are of great rarity, and there is considerable variation from case to case (Harris and Farber, 1939; Van Praagh and Van Praagh, 1967). For the sake of simplicity we prefer not to use the term at all. All such hearts may be described clearly by other terms.

Before leaving the discussion on transposition it should be made clear that though it is common for 'd' transposition to occur in the presence of a ventricular 'd' loop (normal loop), and for 'l' transposition to be associated with an 'l' loop (inversion of the ventricles), this is not invariably so.

There has been a tendency for some clinicians and radiologists to use the term 'd' loop and 'l' loop when only the relation of the great arteries is known with any certainty. The assumption implicit in the use of the terms in this way, is by no means always accurate.

5) Dextroposition and laevoposition of the great arteries

Dextroposition and laevoposition of the aorta or pulmonary artery are commonly understood to mean displacement of the origin of a great artery to right or left over a septal defect.

When the terms are used in relation to the aorta, however, a more restricted meaning has developed and it has become implicit that the terms transposition and dextroposition (or laevoposition) are mutually exclusive. The essential characteristic of dextroposition and laevoposition is that the normal fibrous continuity between the aorta and the mitral valve is maintained. Thus, in hearts in which both great arteries arise from the right ventricle a distinction is drawn between those in which the aorta is transposed and those in which it is extremely dextroposed (Neufeld *et al.*, 1961a, b). This distinction is of considerable embryological significance and it is, therefore, rational to restrict the use of the term 'dextroposition of the aorta' to cases in which mitral-aortic fibrous continuity is present. Similarly, laevoposition of the aorta may be used in the presence of ventricular inversion when the same criterion is met (Fig. 5).

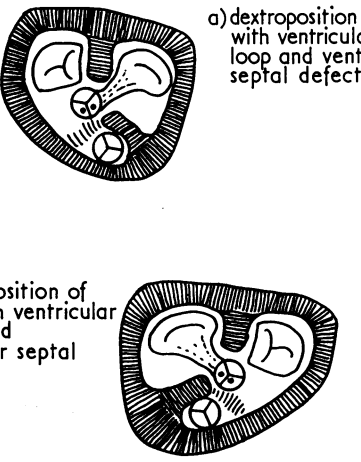

FIG. 5 *Dextroposition and laevoposition of aorta. Schematic diagram to show the relations of the great arteries, AV valves, and ventricular septum.* N.B. *there is fibrous continuity between the aorta and the mitral valve in both situations.*

In relation to the use of the terms laevoposition or dextroposition of the pulmonary artery no such strictures exist, but the terms are vague and imprecise and we prefer to avoid them altogether.

6) 'Persistent truncus arteriosus' (Table 4)

This term refers to failure of septation of the great vessels at their roots. Thus a single orifice is present

TABLE 4 *Terms relating to persistent truncus arteriosus and anomalies which are confused with it (see Fig. 6 and 7)*

	Acceptable terms	*Terms which tend to be confusing*
Truncus	Truncus type 1 Truncus type 2 Truncus type 3	Pseudotruncus Truncus type 4
Other anomalies	Pulmonary atresia Sixth arch agenesis (see text)	

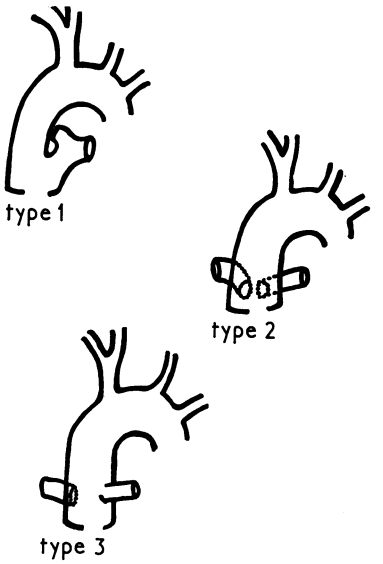

FIG. 6 *Diagram of the three types of persistent truncus (adapted from Collett and Edwards, 1949). N.B. The so-called 'type 4' truncus is not shown here, as it is not universally accepted as a form of truncus (see text and Fig. 7).*

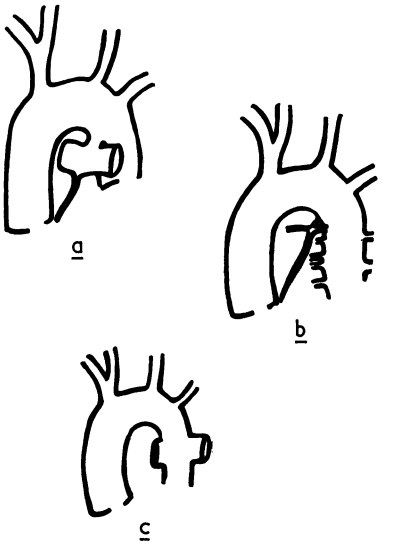

FIG. 7 *Diagram of three conditions commonly confused with truncus.* (a) *'Pulmonary atresia with persistent ductus arteriosus' sometimes called pseudotruncus, a term which should be avoided. Even if no vestige of a main pulmonary artery exists, this is not a form of truncus (4).* (b) *Pulmonary atresia with circulation via small collaterals from the descending aorta.'* (c) *'Sixth arch agenesis with pulmonary circulation via large collaterals from the descending aorta.'* N.B. *This is the anomaly sometimes called 'type 4' truncus. The embryogenesis of the abnormality is, however, quite different from the true forms of 'persistent truncus'.*

and a single vessel emerges from the heart, directly supplying the coronary, the systemic, and the pulmonary circulation. This condition is invariably accompanied by a ventricular septal defect or a common ventricle. There are three main anatomical types of truncus (Fig. 6).

Pulmonary atresia presents certain similarities to 'persistent truncus' and has been called 'pseudotruncus arteriosus'. In this condition a single functional great vessel (the aorta) arising from the heart supplies the coronary and systemic circulation and usually the pulmonary circulation also – via a persistent ductus arteriosus. This condition may be described as 'pulmonary atresia with persistent ductus arteriosus' (Fig. 7a).

In cases of pulmonary atresia where the ductus arteriosus does not persist the lungs may receive their sole blood supply from collaterals. Two variants of this situation occur – those in which the main pulmonary artery can be demonstrated but is extremely hypoplastic, and those in which no vestige of the main pulmonary artery or its branches is present (see Fig. 7a). It is this latter anomaly that has caused the greatest difficulty and confusion in the use of the term truncus. It has been called truncus (type 4) (Collett and Edwards, 1949), but the embryological implications are entirely different from those of the other forms of truncus. In this condition 6th arch agenesis is probably responsible, whereas the deficiency that results in persistence of the truncus is failure of truncal septation (Collett and Edwards, 1949; Edwards *et al.*, 1965).

To our knowledge, no satisfactory term exists whereby this anomaly may be adequately described. We suggest, therefore, that the term 'sixth arch agenesis with pulmonary circulation via collaterals' might be appropriate.[1]

The other abnormality in which, as has been mentioned, the ductus does *not* persist, but in which a hypoplastic pulmonary artery can be demonstrated, should be called 'pulmonary atresia with pulmonary circulation via collaterals' (Fig. 7b).

7) Bilateral conus anomaly

The normal pulmonary artery arises from the muscular infundibulum of the morphological right

[1] Debate still exists about the nature of these collaterals but it seems probable that they are not equivalent to the bronchial arteries'.

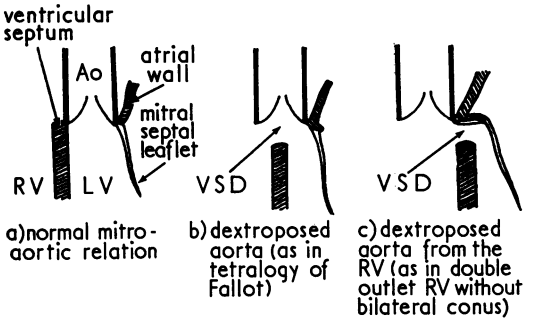

FIG. 8 *Schematic diagram to show the relation of the aorta to the ventricular septum, septal leaflet of the mitral valve, and atrial wall. The diagram represents a vertical section through the aortic root taken at right angles to the plane of the ventricular septum.*

ventricle. On the other hand, the aortic root is related in part to the muscle of the ventricular septum and in part to the atrial wall and the septal cusp of the mitral valve. No ventricular muscle intervenes between the aorta and the atrial wall and there is fibrous continuity between the root of the aorta and the septal cusp of the mitral valve (Fig. 8).

In the great majority of congenital malformations of the heart this relation is still present and clear cut. In the presence of transposition of the great arteries the pulmonary artery is usually in a similar relation to the mitral valve and atrial wall. In the presence of dextroposition or laevoposition of the aorta (with overriding of a VSD), mitral-aortic

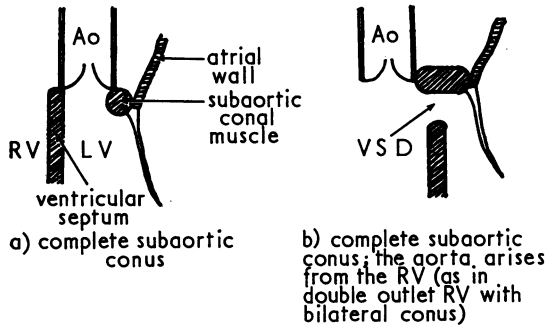

FIG. 9 *Schematic diagram to show the mitral-aortic relation in the presence of a subaortic muscular conus. The diagram represents a vertical section through the aortic root, taken at right angles to the plane of the ventricular septum. (Fig. 9a is a theoretical situation. We have not seen an example of a complete subaortic conus in the presence of an intact ventricular septum as shown here.)*

fibrous continuity is still present and there may sometimes be tricuspid-aortic fibrous continuity in addition.

In all these cases it may be said that the anterior great artery has a 'complete muscular conus' or 'infundibulum' and that the conus of the posterior great artery is incomplete in that it lacks muscle in the area where the artery is related to the mitral valve.

When both great arteries arise from a complete conus, i.e. a complete ring of ventricular muscle, then a 'bilateral' or 'combined' conus is present (Van Praagh and Van Praagh, 1966). In such an abnormality the relation of the posterior vessel with the mitral or tricuspid valve is abnormal and is characterized by the presence of ventricular muscle between the root of the great vessel and the atrioventricular valves (Fig. 9).

A bilateral conus occurs in a number of unusual transposition complexes (Van Praagh and Van Praagh, 1966). It is of sufficient interest and importance because of its embryological implications to be specified separately and in addition to the abnormalities already discussed.

8) Double-outlet right ventricle

This term signifies that both great arteries arise *completely* from the morphological right ventricle and that the only outlet, if any, for the morphological left ventricle is a VSD (Neufeld *et al.*, 1961a, b).

Double-outlet right ventricle may or may not be associated with a 'bilateral conus' anomaly.

Cases with extreme dextroposition of the aorta in a heart otherwise resembling the tetralogy of Fallot may sometimes have the characteristics of a double-outlet right ventricle. This can only be the case when there is no overriding of the septal defect and the aorta arises completely from the right ventricle (Neufeld *et al.*, 1961a). The absence of overriding must be taken as the differentiating feature between the tetralogy of Fallot and double-outlet right ventricle (Fig. 10).

It may be seen that in the double-outlet anomaly the distance between the aortic root and the septal cusp of the mitral valve is increased. This area may be formed merely by atrial muscle lined by endocardium (which is usually thickened) or by conal muscle, in which case a bilateral conus is present.

The position of the great arteries, relative to one another and to the septal defect, is very variable.

9) Taussig-Bing anomaly

The Taussig-Bing anomaly refers to a heart with a transposed aorta arising from the right ventricle and

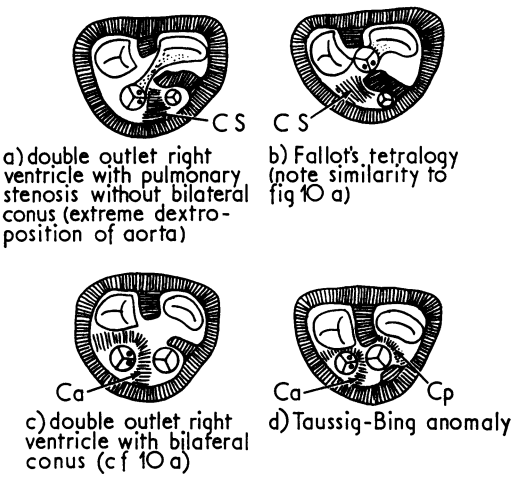

FIG. 10 *Diagram to show the relations of the great arteries, AV valves, ventricular septum, and canal musculature in two situations with dextroposition of aorta (upper) and two with 'd' transposition and a bilateral conus (lower). CS, crista supraventricularis, Ca, subaortic canal muscle; Cp, subpulmonary canal muscle.*

to the right of (and sometimes either slightly anterior or slightly posterior to) the pulmonary artery. The pulmonary artery is large and arises mainly from the right ventricle, overriding a ventricular septal defect to a variable extent. There is a bilateral conus (Van Praagh, 1968).

It has been argued that the Taussig-Bing anomaly is a form of double-outlet right ventricle (Van Praagh, 1968). The basis of this argument is that the pulmonary artery, though overriding the ventricular septum, never overrides the cavity of the left ventricle. This argument is, we think, invalid on two grounds. First, we have in our material an example of the Taussig-Bing anomaly in which the pulmonary artery quite clearly does override the cavity of the left ventricle and, secondly, the difference between Fallot's tetralogy and double-outlet right ventricle without a bilateral conus depends on whether the aorta overrides the septal defect or not. By definition the Taussig-Bing anomaly has an overriding pulmonary artery (Taussig and Bing, 1949; Van Praagh, 1968; Beuren, 1960). We therefore consider that the term 'double-outlet right ventricle' is inaccurate and should not be used for this condition.

Conclusion

The vast majority, indeed almost all, abnormalities of position of the heart, its chambers, and the great arteries can be accurately and precisely described by the use of the simple and unambiguous terms defined here. In the very small number of cases where unusual anomalies defy classification on these lines, it is clearly mandatory that a full and careful description of all abnormalities present should be given, using always the simplest available anatomical terms.

We wish to thank Dr. C. G. Parsons, Dr. S. P. Singh, Dr. R. Astley, and Dr. H. Cameron for helpful advice and criticism, and Dr. H. Cameron for allowing us to study the collection of pathological specimens in his department.

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