A Simplified Basic Classification of Spatial Alignments of the Heart, Its Chambers, and the Great Vessels

By HAROLD D. ROSENBAUM, M.D.

TERMINOLOGY describing spatial arrangements of the heart is variable, inconsistent, and altogether confusing. Accepted precise definitions are not available for such frequently employed terms as dextrocardia, dextroposition, dextroversion, and levocardia. The problem is compounded in instances of so-called corrected transposition of the great vessels where, by the nature of the anomaly, discordance exists in the relationships of the atria and ventricles. Some authors use "right" and "left" to denote spatial location while others use the same terms to indicate the internal anatomy of the cardiac chambers. Functional designations, such as "venous" and "arterial," are not satisfactory nor sufficiently specific or descriptive, since they are not always applicable and do not account for the anatomy or location of the chambers.

Development of a useful classification system presents many problems. Recourse to the literature is singularly unrewarding due not only to a lack of accepted terminology but, more importantly, to incomplete descriptions of clinical and autopsied cases. The disposition of the heart, its chambers, the great vessels, and the abdominal viscera must be considered in a thorough classification of spatial alignments. Descriptions of the internal anatomy of the cardiac chambers are particularly valuable in evaluating case reports but, unfortunately, the literature rarely contains information relative to all these items.

The need for an accepted nomenclature and a system of classification has been noted previously. Although no generally applicable system has appeared in the medical literature, useful classifications in limited areas have been noted. Consequently, the problem has been reviewed and a system of classification, basic in concept, is suggested. Such a classification to be useful must be embryologically sound, clinically applicable, and sufficiently uncomplicated to encourage usage. It is recognized that a classification of this nature cannot at the present time embrace all possible variations, particularly multiple and complicated anomalies. Nevertheless, it is hoped this system represents a superstructure that will be helpful in approaching problems of this nature and upon which a classification of more complicated anomalies and variations can be engrafted.

Location of the Heart

Location of the heart primarily in the right chest with its apex directed toward the right has been designated under various circumstances as dextrocardia, dextroposition, dextroversion, etc. Levocardia, levoposition, and levoposition reasonably could be expected to indicate the opposite of dextrocardia, dextroposition, and dextroversion, although such is not always the case. By one definition levo- cardia exists when there is a left-sided heart with the apex directed toward the left in association with inversion of abdominal viscera. Levocardia, as far as can be determined, is not commonly employed to denote a normally placed heart in the absence of other visceral or chamber malpositions. Although the significance of the location of the heart may be intimately related to the disposition of the abdominal viscera, consideration of this factor in terminology describing the cardiac position may not be necessary and in some circumstances can be confusing. Parenthetically, levo- probably is used more correctly

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as the opposite prefix of dextro- in chemical terminology. Sinistro- appears to be the more appropriate opposite of dextro- for anatomic positions.

To avoid ambiguous terms and to promote clarity, a heart primarily in the left chest with its apex directed toward the left is designated simply a “left-sided” heart. Similarly, a heart primarily in the right chest with its apex directed toward the right is designated a “right-sided” heart. It should be noted that the terms “right” and “left” are always used to indicate location and have no morphologic or functional connotation.

This classification is concerned only with normal and abnormal positions of the heart and its chambers resulting primarily from normal or deranged cardiogenesis. Secondary displacements of the heart due to extracardiac developmental aberrations and postnatal influences are not included in this system.

It is generally thought that the final laterality of the heart is determined by the direction of the bulboventricular loop and whether it undergoes a final apical shift to the opposite side of the thorax in the terminal phases of development. Obviously a “left-sided” heart could result from a bulboventricular loop with a normal right convexity undergoing a final apical swing to the left (the usual normal cardiogenesis), or a bulboventricular loop which initially had its convexity directed toward the left but failed to undergo a final apical shift into the right chest. The reverse of these conditions explains most instances of a “right-sided” heart. A relatively midline or “mesial” heart results when the final apical shift, whether the bulboventricular loop was initially directed toward the right or the left, is incomplete and becomes arrested in the midline. Only rarely does the heart stop precisely in the midline. Usually there is a predominance, albeit at times very slight, toward one side of the thorax, particularly when the location of the apex impulse is considered. With angiocardiography the apex can always be identified as toward one or the other side; hence, laterality is determinable even though the heart be near or in the midline.

Clinical determination of the location of the major heart mass is, of course, very easy. Although percussion, palpation, and auscultation are usually definitive, the heart can be located decisively with a chest film. Only in the rare mesial, or near midline heart, will angiocardiography very infrequently be required.

The Atria

Laterality of the atria relative to one another usually is quite distinct. They do not tend to overlap so much, nor be so variable relative to one another, as do the ventricles. The terms “right” and “left” and “inverted” and at times “noninverted” have been used to describe the positions of the atria. Unfortunately, “right” and “left” are used by some to describe the relative locations of the atria, and by others to indicate their basic internal anatomy, regardless of their spatial relationship. “Inverted” atria usually indicate that the normal anatomic dextral atrium is located on the left regardless of the location of the heart as a whole or laterality of the other viscera, although this is not invariable nor always clear in the literature. Confusion may therefore result. Atrial inversion, defined in this manner, would be expected in total situs inversus but not expected (perhaps nonexistent) in situs solitus.

In this classification an effort is made to avoid terms that have been used in the past with varying or vague meanings. Consequently, an atrium with the characteristics of a morphologic right atrium is designated as “dextral.” Similarly, an atrium with the internal anatomy of a left atrium is designated as “sinistral.” The use of “dextral” and “sinistral” is determined solely by internal anatomic considerations and has no reference to spatial laterality.

A meaningful designation of the atria in regard to their positions relative to the location of the major heart mass obtains from additionally describing the atria as “concordant” or “discordant.” With a left-sided heart, concordant atria indicate that the sinistral atria...
um is on the left and the dextral atrium is on the right. Conversely, a left-sided heart with discordant atria has the sinistral atrium on the right and the dextral atrium on the left. Similarly, a right-sided heart with concordant atria has the sinistral atrium on the right.

According to de la Cruz, laterality of the atria is not influenced by the original direction of the bulboventricular loop. Embryologic influences during cardiogenesis lead to a posterior location and decreased mobility of the atria. Consequently the atria act as anchors and are not spatially dislocated to a marked degree by influences that affect the location of the future ventricular, bulbar, and truncal regions of the heart.

There appears to exist a relationship between the lateral disposition of the atria and the noncardiac abdominal viscera. Generally speaking, atrial laterality parallels abdominal visceral laterality. The literature does not provide many cases in which these details are covered, although in almost every instance the dextral atrium is on the right when the stomach is on the left. Exceptions are noted in isolated inversion of the stomach where there is no cardiac anomaly, and Lev and Rowlatt reported a case (no. 13) that appears to contradict this general statement. An occasional case of congenital heart disease associated with asplenia (Ivemark's syndrome) appears not to fit this generalization, although this syndrome may be a special situation. It has been suggested that Ivemark's syndrome results in abnormal symmetry and reflects a condition in which laterality of the viscera, including the heart, fails to develop properly in the expected manner. It should be noted that Ivemark does not specifically describe the internal anatomy of the atria in his cases and apparently uses "right" and "left" to denote location.

Problems arise in determining whether abdominal viscera showing partial malrotation represent deranged or incomplete visceral situs inversus rather than deranged or incomplete visceral situs solitus. The location of the stomach appears to be as good an indicator as is available. It certainly has the advantage of being determined easily by clinical means.

Differentiation of the dextral atrium from the sinistral atrium is very easy on pathologic examination (table 1). The dextral atrium presents the limbus fossa ovalis and usually the openings of the inferior vena cava and coronary sinus, while the sinistral atrium is recognized by the presence of the septum primum derivative. Clinical differentiation of the atria is less easy although it presents no insurmountable problem (table 2). With

<table>
<thead>
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<th>Table 1</th>
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Anatomic Characteristics of the Cardiac Chambers (Presence of Septa Required for Definite Identification)

<table>
<thead>
<tr>
<th>Dextral atrium (morphologic right atrium)</th>
<th>Sinistral atrium (morphologic left atrium)</th>
<th>Dextral ventricle (morphologic right ventricle)</th>
<th>Sinistral ventricle (morphologic left ventricle)</th>
</tr>
</thead>
</table>
| Presence of limbus fossa ovalis and usually openings of inferior vena cava and coronary sinus | Presence of septum primum derivative | 1. Usually tricuspid AV valve
2. Trabeculated septal surface
3. Presence of crista supraventricularis and conus
4. Semilunar valve, anterior
5. AV and semilunar valves somewhat separated | 1. Usually bicuspid AV valve
2. Smooth septal surface
3. Absence of crista supraventricularis and conus
4. Semilunar valve, posterior
5. AV and semilunar valves close together and connected with a fibroplastic continuum
6. Muscle fibers stream parallel to blood outflow |

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Table 2

Clinical Differentiation of the Cardiac Chambers

<table>
<thead>
<tr>
<th>Dextral atrium (morphologic right atrium)</th>
<th>Sinistral atrium (morphologic left atrium)</th>
<th>Dextral ventricle (morphologic right ventricle)</th>
<th>Sinistral ventricle (morphologic left ventricle)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Catheter and opaque media from inferior vena cava usually enter this atrium</td>
<td>1. Pulmonary veins usually enter sinistral atrium</td>
<td>1. Opaque studies usually show more trabeculated boundaries, crista supraventricularis, and infundibulum</td>
<td>1. Opaque studies show smoother boundaries, no crista supraventricularis or infundibulum</td>
</tr>
<tr>
<td>Stomach on the left, and upright P wave in lead I and inverted P wave in leads II and III usually means dextral atrium is on the right</td>
<td>3. Atrioventricular and semilunar valves somewhat separated</td>
<td>3. Atrioventricular and semilunar valves relatively close together</td>
<td>4. No separate inflow and outflow tracts</td>
</tr>
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<td></td>
<td>4. Usually more elongated than sinistral ventricle with definite inflow and outflow tracts</td>
<td></td>
<td>5. Semilunar valve usually anterior</td>
</tr>
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</table>

rare exceptions \(^{(14,15)}\) a catheter introduced from the inferior vena cava enters the dextral atrium. Although exceptions exist, \(^{(16,17)}\) a negative P wave in lead I and positive P waves in leads II and III are helpful in indicating that the dextral atrium is on the left and the sinistral atrium is on the right. \(^{(17,18)}\)

Angiocardiographic appearances are useful in this differentiation, since the sinistral atrium is characteristically more oval and less elongated in its supero-inferior axis than the dextral atrium. Also, the progression of opaque medium from the cavae is almost always into the dextral atrium. Finally, location of the stomach on the left from examination of the chest film or barium studies is strong evidence that the dextral atrium is on the right and the sinistral atrium is on the left.

The Ventricles and Great Vessels

The ventricles and great vessels are considered together, since their embryogeneses, although incompletely understood, are intimately entwined. This developmental relationship causes spatial derangements of the ventricles to affect the alignment of the great vessels and vice versa. Absence of a mesentery for the bulboventricular loop in early cardiogene sis allows great mobility for this portion of the primitive heart tube, resulting in variability in the final definitive locations of the ventricles. In normal individuals absolute spatial laterality of one ventricle relative to the other is much less definite than is true of the atria. In fact, the dextral ventricle is more strikingly anterior than to the right of the sinistral ventricle. Congenital anomalies further cloud this relationship, if they lead to selective hypertrophy or dilatation of one ventricle, particularly if there is an associated element of cardiac rotation.

The term “ventricular inversion” has been commonly used to indicate that the dextral ventricle is located in the proper position for the sinistral ventricle and vice versa. “Inversion,” in general, has been employed to indicate reversal in the usual lateral locations of a pair of cardiac chambers. This is to be contrasted to “transposition,” which is usually, although not invariably, defined as reversal in the anteroposterior relationship of structures being described. Usually neither definition is completely nor totally descriptive, since there are many borderline cases, and frequently there are elements of relative displacement in both the sagittal and the coronal.
planes. When these terms, particularly inversion, are used in relation to reported cases they often are not defined with precision. It may be particularly difficult to fathom the author's meaning when there is associated dislocation of the atria or the entire heart. The important consideration is whether the ventricles are translocated in relation to their feeding atria.

"Complete transposition of the great vessels" and "corrected transposition" are in common use and enjoy a considerable degree of general agreement and understanding as to the anomalous situations being described. It has been suggested that "congenital" should be added to the term "corrected transposition" to indicate that whatever correction exists resulted from embryologic influences. "Corrected transposition" has been further criticized, since, in most instances, the blood flow is not totally corrected because of associated defects that adversely affect the circulation.

The possible courses of blood flow through the heart in various alignments of the chambers and great vessels merit discussion at this point. Only total translocations are considered (fig. 1). A dextral atrium can empty into either a dextral ventricle or a sinistral ventricle. Similarly, a dextral ventricle can empty into either the aorta or the pulmonary artery. It is obvious that the sinistral ventricle has an identical potentiality. These considerations presume an oppositely counterposed course being followed by blood from the sinistral atrium, although it is recognized that many variations and alterations of this basic pattern exist in complicated anomalies.

The four possible basic routes the blood can follow as it passes from the dextral atrium can be designated as pursuing courses determined by the following ventriculotrunical alignments. In cases designated "normal" the flow of blood is from the dextral atrium to the dextral ventricle to the pulmonary artery. In "noninverted transposition" the course of blood is from the dextral atrium to the dextral ventricle to the aorta. This is classical complete transposition of the great vessels. In "inverted transposition" the flow of blood is from the dextral atrium to the sinistral ventricle to the pulmonary artery. This is the anatomic alignment in so-called corrected transposition of the great vessels. In "isolated ventricular inversion" the flow of blood is from the dextral atrium to the sinistral ventricle to the aorta. This is a very rare condition.

By this terminology "ventricular inversion" signifies translocation of the two ventricles, or discordancy of the ventricles, relative to the feeding atria. In "ventricular inversion" ventricular spatial laterality varies inversely with the location of the dextral and sinistral atria.

"Transposition," for purposes of this classification, indicates that the aorta arises anteriorly from the dextral ventricle and the pulmonary artery arises posteriorly from the sinistral ventricle. Transposition, as a term, has been applied to almost any abnormality of origin of the great vessels. "Inverted transposition" has been used previously by Lev in the sense that it is employed in the classification. It should be noted that "dextral" and "sinistral" when used in describing the ventricles, as in describing the atria, refer to morphology and not location.

In a normal individual with a left-sided heart and concordant atria, the aorta arises slightly to the right and posterior to the pulmonary artery. In noninverted transposition (complete transposition of the great vessels) the aorta arises anterior and slightly to the right of the pulmonary artery; whereas in inverted transposition (so-called corrected transposition) the aorta arises on the same level or slightly anterior and to the left of the pulmonary artery. It might be expected that in isolated ventricular inversion the aorta would arise posterior and slightly to the left of the pulmonary artery. Thus, the aorta

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**Figure 1** Possible courses of blood flow from a dextral atrium.
would make a complete circle around the pulmonary artery in presenting these four anatomic situations (table 3). Unfortunately, isolated ventricular inversion is extremely rare, and the few cases (cases 9 and 10) discovered are reported not to demonstrate this aortic-pulmonary artery relationship.

Normal ventriculotrunical alignment is said to result from a normally directed bulboventricular loop with the truncocoanal septum eventually evolving into a normal spiral arrangement of the great vessels, although the underlying mechanisms are debated and not fully understood. Although the embryologic aberrations that lead to noninverted transposition (complete transposition of the great vessels) are not completely known and in some measure disputed, many investigators believe that the basic disturbance is failure of the truncocoanal septum to develop in a spiral fashion. It may well be that some malpositions of the great vessels at the base of the heart represent an additional or different patho-embryogenic mechanism. According to de la Cruz, inverted transposition (corrected transposition) results from the bulboventricular loop developing in a direction opposite to that expected in association with a straight truncocoanal septum. Isolated ventricular inversion possibly might be expected as the end product of the bulboventricular loop developing in a direction opposite to that expected but with the truncocoanal septum eventuating into a normal spiral arrangement (table 3).

From an embryologic point of view the dextral ventricle should lie anterior to the sinistral ventricle because of the developmental continuity of the future dextral ventricle and truncus at the cephalic end of the primitive heart tube. The portion of this tube that eventually will become the sinistral ventricle is caudal and continuous with the atria. Since the atria are anchored posteriorly and the great vessels emerge anteriorly, the sinistral ventricle usually is disposed relatively posterior to the dextral ventricle. This anteroposterior relationship is much more constant than definite laterality. Unfortunately, so much variation in ventricular size and rotation may exist that this anteroposterior relationship of the bodies of the two ventricles cannot be used with assurance. If only the anteroposterior relationship of the outflow valves of the ventricles is considered, however, this differentiation is more reliable, since the outflow point of the dextral ventricle in most cases tends to be slightly anterior to the outflow point of the sinistral ventricle.

Although most case reports in the literature do not describe the internal anatomy of the heart in detail, differentiation of the dextral from the sinistral ventricle is quite easy at the autopsy table if two distinct ventricles are present. As with the atria, a septum must be present for a definitive identification (table 1). The dextral ventricle is characterized by a tricuspid atroventricular valve although exceptions have been reported. Its septal surface is trabeculated, and a definite crista supraven-

| Table 3 |
|-----------------|-----------------|-----------------|-----------------|
| **Ventriculotrunical alignment** | **Ventricular origin of great vessels** | **Relations of origins of great vessels** | **Possible embryologic explanation** |
| Normal | Aorta from sinistral ventricle; PA from dextral ventricle | Aorta posterior to the right of PA | Normally directed BV loop; spiral truncocoanal septum |
| Noninverted transposition | Aorta from dextral ventricle; PA from sinistral ventricle | Aorta anterior to the right of PA; PA from sinistral ventricle | Normally directed BV loop; straight truncocoanal septum |
| Inverted transposition | Aorta from dextral ventricle; PA from sinistral ventricle | Aorta anterior to the left of PA | Reversal of BV loop; straight truncocoanal septum |
| Isolated ventricular inversion | Aorta from sinistral ventricle; PA from dextral ventricle | ??Aorta posterior and to the left of PA | ??Reversal of BV loop; ??spiral truncocoanal septum |

Location of the heart determined by direction of BV loop and occurrence or lack of terminal apical shift. Atrial positions theorized to reflect general visceral laterality: PA, pulmonary artery; BV, bulboventricular. * Origin of the great vessels as expected in a left-sided heart with atrial concordance.
tricularis and infundibulum are evident. On the other hand, the sinistral ventricle usually shows a bicuspid atrioventricular valve, although again exceptions have been reported. The septal surface is relatively smooth, and no crista supraventricularis or infundibulum is present. The muscular fibers of the sinistral ventricle usually stream parallel to the direction of the blood outflow.

A fibroplastic continuum runs from the atrioventricular valve guarding the sinistral atrium to a semilunar valve. Continuity with the aortic valve indicates absence of transposition of the great vessels. A similar structure running from the atrioventricular valve guarding the sinistral atrium to the pulmonic valve is an anatomic hallmark of transposition.

Clinical differentiation of the ventricles and the great vessels is quite difficult and usually requires angiocardiography (table 2). Prolongation of the PR interval is common in inverted transposition, since the sinus node is located in the dextral atrium, and the conduction fibers therefore must pursue a more lengthy course than usual to reach the ventricular musculature. Although this electrocardiographic pattern as well as certain patterns of QRS progression in the chest leads may be helpful, they are not diagnostic. Aspects of the physical examination are often suggestive but not conclusive. A film of the chest in inverted transposition often shows an unusual prominence above the expected location of the pulmonary artery, which represents the displaced aorta, and there is rarely a normal aortic impression on the barium-filled esophagus.

Difficulty in passing a cardiac catheter through a ventricle suggests its being a sinistral ventricle. This difficulty arises from the relative contiguity of the atrioventricular and semilunar valves guarding the sinistral ventricle. Since these valves are placed near each other, the catheter must inscribe a rather sharp curve in traversing this chamber. The valves of the dextral ventricle are more widely separated, allowing easier passage of a cardiac catheter. Location of the semilunar valves and separation of atrioventricular and semilunar valves may be appreciated at cardiac catheterization and can be helpful in differentiating noninverted and inverted transposition as well as the other entities. De la Cruz et al. have remarked upon the parallel courses and lack of crossing of catheters placed in the two sides of the circulation in inverted transposition.

Greatest aid comes from angiocardiography. Locations and proximity of the atrioventricular valves and the semilunar valves are seen. In the majority of cases a well-defined crista supraventricularis can be seen in the dextral ventricle, and the more trabeculated septal wall of the dextral ventricle is usually appreciated when adequate opacification is obtained. The dextral ventricle is a more elongated structure than the sinistral ventricle and quite often its inflow and outflow tracts can be identified. As mentioned previously, the ventricle whose outflow valve lies distinctly posterior to its mate is the sinistral ventricle, if no outstanding factors of rotation, dilatation, or underdevelopment exist.

Angiocardiography permits ready identification of the aorta and pulmonary artery. Determination of the atrium (dextral or sinistral) from which a ventricle fills and the great vessel (aorta or pulmonary artery) into which it discharges, when considered in the light of other evidence, usually allows its anatomic identification (dextral or sinistral) and classifies the spatial alignment (fig. 2).

In classical examples, when the aorta arises distinctly posterior to the pulmonary artery, it is expected to be arising from a sinistral ventricle, and the circulation will be "normal ventriculo-truncal alignment" or possibly "isolated ventricular inversion." When the aorta arises distinctly anterior to the pulmonary artery it is expected to be arising from the dextral ventricle and represents either "non-inverted transposition" or "inverted transposition." With a left-sided heart and concordant atria, origin of the aorta to the right and posterior (from the sinistral ventricle) to the origin of the pulmonary artery occurs in "normal ventriculo-truncal alignment." In an otherwise similar situation aortic origin to the
A simplified basic classification of spatial alignment of the heart, its chambers, and great vessels.

The four basic alignments of the atria, ventricles, and great vessels in this classification have been designated normal ventriculotrunical alignment, noninverted transposition, inverted transposition, and isolated ventricular inversion. These four alignments may be seen in four different situations depending upon the location of the heart and concordancy of discordance of the atria (table 4). Consequently, use of this system permits cardiac position and chamber alignment to be

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### Figure 2

A simplified basic classification of spatial alignment of the heart, its chambers, and great vessels.

<table>
<thead>
<tr>
<th>LOCATION OF THE HEART</th>
<th>ATRIA</th>
<th>VENTRICLES</th>
<th>GREAT VESSELS</th>
<th>COURSE OF BLOOD FLOW</th>
<th>VENTRICULOTRUNICAL ALIGNMENT</th>
<th>CF WITH NUMBERS</th>
<th>DIAGRAMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>LEFT-SIDED</td>
<td></td>
<td></td>
<td>NONINVERTED</td>
<td>DA-DV-PA</td>
<td>NORMAL</td>
<td>5,9,13</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>INVERTED</td>
<td>DA-DV-AORTA</td>
<td>NONINVERTED TRANSPOSITION</td>
<td>6,10,14</td>
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<tr>
<td></td>
<td></td>
<td>DEXTRAL ATRIUM ON RIGHT</td>
<td></td>
<td>DA-SV-PA</td>
<td>INVERTED TRANSPOSITION</td>
<td>7,11,15</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>DEXTRAL ATRIUM ON LEFT</td>
<td>NONINVERTED</td>
<td>DA-SV-AORTA</td>
<td>ISOLATED VENTRICULAR INVERSION</td>
<td>8,12,16</td>
<td>4</td>
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<tr>
<td>RIGHT-SIDED</td>
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<td>DA-DV-PA</td>
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<td>DA-DV-AORTA</td>
<td>NONINVERTED TRANSPOSITION</td>
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<td>2,10,14</td>
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<td>DA-SV-PA</td>
<td>INVERTED TRANSPOSITION</td>
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<td>ISOLATED VENTRICULAR INVERSION</td>
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<td>4,8,12</td>
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left and posterior to the pulmonary artery should represent “isolated ventricular inversion,” although no such case has been uncovered. Likewise, origination of the aorta anterior (from a dextral ventricle) and to the right of the pulmonary artery indicates “noninverted transposition,” and aortic origin parallel or anterior and to the left of the pulmonary artery is seen in “inverted transposition.” Reversed lateral positions of distal chambers are expected with atrial discordance in the presence of a left-sided heart. A similar list of alignments can be enumerated for a rightsided heart (fig. 2). Obviously, biplane angiography is required for this differentiation. Confirmatory differential information can be obtained from a study of the coronary artery distribution as seen on properly executed angiography of these vessels.

**Comment**

The four basic alignments of the atria, ventricles, and great vessels in this classification have been designated normal ventriculotrunical alignment, noninverted transposition, inverted transposition, and isolated ventricular inversion. These four alignments may be seen in four different situations depending upon the location of the heart and concordancy of discordance of the atria (table 4). Consequently, use of this system permits cardiac position and chamber alignment to be
described as one of the four patterns with either a left-sided or right-sided heart and atrial concordance or discordance. By this classification basic spatial alignments are reduced to their simplest components, and confusing and differently defined terms are avoided. An effort has been exerted to avoid ambiguous terminology and to offer as few new terms as possible to maintain clarity of meaning. Each term used is carefully defined (table 5).

Review of the literature suggests that an incomplete apical shift is more common in inverted transposition than in noninverted transposition and normal ventriculotruncal alignment. It might be reasonable to expect frequent failure of the terminal apical shift in isolated ventricular inversion. Since the basic embryologic defect in this condition, in common with inverted transposition, could well be a reversed bulboventricular loop, but without an alteration in definitive spiraling of the truncoconal septum that is said to be part of the embryologic substrate of inverted transposition. This assumption cannot be proved, since an insufficient number of cases of isolated ventricular inversion is available to test the hypothesis.

The relative incidence of the various spatial alignments is unknown, since there is an insufficient number of cases reported in adequate detail to allow classification. No attempt has been made to cover every single case report, particularly in the foreign, remote, or obscure journals. An initial effort in this direction was disappointing due to a general lack of details in the reports as well as frequent inability readily to appreciate the sense of the authors’ definitions. An increasing body of opinion is developing that many conclusions reached in older reports are not supported by anatomic data and appear, at least in many instances, to be incorrect. The major purpose of this manuscript is to present a logical system for future use rather than classify all previously reported cases. It is obvious, however, that normal ventriculotruncal alignment has been encountered in all positions of the heart and atrial concordance or discordance. The various heart and atrial locations associated with inverted and noninverted transposition can, in all likelihood, be documented. On the other hand, isolated ventricular inversion has been reported very infrequently. It is hoped a more detailed description of cases reported in the future will reveal more examples of this particular type of ventriculotruncal alignment.

It should be noted that the presence of a particular ventriculotruncal alignment in association with a specific heart location and atrial concordance or discordance may have distinct clinical and pathologic associations and significance, but a consideration of these matters is beyond the scope of this paper.

This classification is obviously an oversimplification of a very confused problem. Much remains to be learned about normal and abnormal cardiogenesis. There are, of course, complicated anomalies due to various embryogenic aberrations that defy classification within this system. Additional malformations can lead to chamber variations that make determination of the underlying spatial alignment difficult when only the exact observed locations of the chambers are considered.

<table>
<thead>
<tr>
<th>Location of the heart and the atria</th>
<th>Normal</th>
<th>Noninverted transposition</th>
<th>Inverted transposition</th>
<th>Isolated ventricular inversion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left-sided heart with atrial concordance</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Left-sided heart with atrial discordance</td>
<td>5</td>
<td>6</td>
<td>7</td>
<td>8</td>
</tr>
<tr>
<td>Right-sided heart with atrial concordance</td>
<td>9</td>
<td>10</td>
<td>11</td>
<td>12</td>
</tr>
<tr>
<td>Right-sided heart with atrial discordance</td>
<td>13</td>
<td>14</td>
<td>15</td>
<td>16</td>
</tr>
</tbody>
</table>

* Numbers listed refer to those shown in column immediately preceding diagrams of figure 2.
These factors, as well as the pivotal rotations described by Lev,13 cause absolute spatial locations of the chambers, especially the ventricles, to be of only relative value. Although anomalies exist that cannot be completely categorized into this classification, general acceptance of a terminology and a basic classification system provides a frame of reference for future refinements. The classification presented, although inadequate, appears to have merit and should permit a thoughtful and systematic consideration, discussion, and study of these conditions and consequently should be helpful in advancing knowledge and understanding of these anomalies.

Summary

The problems resulting from a lack of accepted terminology and a basic classification system for the various spatial alignments of the heart, its chambers and great vessels are reviewed. A basic simplified classification system founded on fundamental embryologic, clinical, and roentgenologic considerations is proposed. Terminology employed is precisely defined. The clinical studies needed to place patients in their category are enumerated and discussed. It is recognized that some forms of spatial derangement defy classification within this system. Nevertheless, this classification system with the suggested terminology, however inadequate, is offered as a means to promote clinical definition of these entities and to encourage clear discussion and a better understanding of these conditions. It is hoped that future study will add to our knowledge of the embryologic mechanisms involved and eventually lead to a more comprehensive classification embracing a larger number of congenital cardiac anomalies.

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References

A Simplified Basic Classification of Spatial Alignments of the Heart, Its Chambers, and the Great Vessels

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