Dextrocardia—value of segmental analysis in its categorisation

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SUMMARY Dextrocardia can be defined as a heart in the right chest with the major axis to the right. This definition, however, conveys no information regarding the chamber arrangements and internal anatomy of the heart.

Of 40 patients satisfying this definition in the files of the Brompton Hospital, 33 had angiocardio- graphic data adequate for complete analysis in terms of connections, relations, and morphology of cardiac segments. They form the subject of this report.

There were 16 (48%) patients with situs solitus, 11 (33%) with situs inversus, and six (18%) with situs ambiguus. Of the cases of situs ambiguous, four exhibited laevoisomerism and two dextroisomerism. Of the 16 patients with situs solitus, six had two ventricles and 10 had univentricular hearts; two patients had concordant and three discordant ventriculoarterial connections, seven had double outlet ventricle, and four a single outlet heart. Of the 11 patients with situs inversus, nine had two ventricles and two a univentricular heart of right ventricular type; the arterial connection was concordant in two, discordant in two, double outlet in six, and single outlet in one. Of the six patients with situs ambiguous and laevo or dextroisomerism, four had two ventricles, and two univentricular hearts; the arterial connection was concordant in one, double outlet in three, and single outlet in two.

Segmental analysis and the use of basic descriptive terms are essential to define the complex anatomy of such hearts.

The term dextrocardia is used by some authors (Van Praagh et al., 1964; Van Praagh and Vlad, 1978) to describe the situation where on plain chest x-ray the heart is predominantly in the right hemithorax. Others use the term to describe the situation where the apex or major axis of the heart (Lev et al., 1968; Squarcia et al., 1973) points to the right. As a right-sided position of the heart has no implications for the internal anatomy, controversy about the precise definition or use of the term dextrocardia, or such terms as dextroposition, dextroversion, dextrorotation, or dextrosis, is unhelpful. As Van Praagh and colleagues (1964) and Squarcia et al. (1973) have shown, segmental analysis is essential for complete description of such hearts. In this paper we describe an analysis of cases from the Brompton Hospital, fulfilling these definitions for dextro-

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Fig. 1 Chest radiographs illustrating dextrocardia with situs solitus (a), situs inversus (b), situs ambiguus with dextroisomerism (c), and with laevoisomerism (d). Situs was judged from bronchial anatomy which was easily visible on radiographs. It is less obvious on prints and position of bronchi is indicated by dotted lines.

this report. Each of these patients was analysed in terms of connections, relations, and morphology using the system described elsewhere in detail (Tynan et al., 1979).

Findings

The anatomy of the hearts in the patients studied is shown in the Table. There were 16 patients with situs solitus (48%), 11 (33%) with situs inversus, and six (18%) with situs ambiguus (Fig. 2). Four of the cases of situs ambiguous exhibited laevoisomerism, and two dextroisomerism.

Situs solitus
In all patients we found visceral, thoracic, and atrial situs to correspond.

Atrioventricular junction (Fig. 3)
Of 16 patients with situs solitus, two had concordant and four discordant atrioventricular connections. The remaining 10 patients had
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Solitus - 16 cases

Inversus - 11 cases

Ambiguous with dextro isomerism - 2 cases

Ambiguous with laevo isomerism - 4 cases

Fig. 2. Diagram illustrating bronchial anatomy. RAA, right atrial appendage; LAA, left atrial appendage.

ventricles (concordant atrioventricular connection), to the left in four (discordant atrioventricular connections), and anterosuperior to the left ventricle in one (concordant atrioventricular connections).

Arterial junction

One of the two patients with concordant atrioventricular connections had concordant ventriculo-arterial connections, the aorta being anterior and to the right of the pulmonary artery. The other had a truncus arteriosus with a right-sided origin of the pulmonary trunk.

Of the four patients with discordant atrioventricular connections, the arterial connections were discordant in one, the aorta being anterior and to the left, and double outlet right ventricle in three. In the latter cases the aorta was anterior and to the left in two and directly anterior in one; all had bilateral infundibula.

Of the 10 patients with univentricular heart, only one, with a main chamber of left ventricular type, had concordant arterial connections. Two univentricular hearts of left ventricular type exhi-
Table Distribution and anatomy of patients studied; every heart was analysed in terms of connections, relations, and morphology (see text)

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Atrial situs</th>
<th>AV junction connection</th>
<th>Mode of connection</th>
<th>Ventricular morphology</th>
<th>Relation of ventricles and of rudimentary to main chamber</th>
<th>Arterial junction connection</th>
<th>Relation of arteries</th>
<th>Infundibular morphology</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Solitus</td>
<td>Concordant</td>
<td>2 AV valves</td>
<td>2 ventricles</td>
<td>RV superior to LV RV to right of LV and anterior</td>
<td>Concordant</td>
<td>Single outlet truncus</td>
<td>AR</td>
</tr>
<tr>
<td>2</td>
<td>Solitus</td>
<td>Concordant</td>
<td>2 AV valves</td>
<td>2 ventricles</td>
<td>RV superior to LV RV to right of LV and anterior</td>
<td>Concordant</td>
<td>Single outlet truncus</td>
<td>AR</td>
</tr>
<tr>
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<td>Solitus</td>
<td>Discordant</td>
<td>2 AV valves</td>
<td>2 ventricles</td>
<td>RV to left of LV and anterior</td>
<td>Discordant</td>
<td>AL</td>
<td>Subaortic</td>
</tr>
<tr>
<td>4</td>
<td>Solitus</td>
<td>Discordant</td>
<td>2 AV valves</td>
<td>2 ventricles</td>
<td>RV to left of LV and anterior</td>
<td>Discordant</td>
<td>AL</td>
<td>Subaortic</td>
</tr>
<tr>
<td>5</td>
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<td>Discordant</td>
<td>2 AV valves</td>
<td>2 ventricles</td>
<td>RV to left of LV and anterior</td>
<td>Discordant</td>
<td>AL</td>
<td>Subaortic</td>
</tr>
<tr>
<td>6</td>
<td>Solitus</td>
<td>Discordant</td>
<td>2 AV valves</td>
<td>2 ventricles</td>
<td>RV to left of LV and anterior</td>
<td>Discordant</td>
<td>AL</td>
<td>Subaortic</td>
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<tr>
<td>7</td>
<td>Solitus</td>
<td>Double inlet</td>
<td>R AV valve straddling</td>
<td>UV heart</td>
<td>OC to right of MC and anterior</td>
<td>Concordant</td>
<td>RP</td>
<td>Subaortic</td>
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<tr>
<td>8</td>
<td>Solitus</td>
<td>Double inlet</td>
<td>Common AV valve</td>
<td>UV heart</td>
<td>Pouch to right of MC</td>
<td>Double outlet</td>
<td>A</td>
<td>Subaortic</td>
</tr>
<tr>
<td>9</td>
<td>Solitus</td>
<td>Double inlet</td>
<td>L AV valve</td>
<td>UV heart</td>
<td>Pouch posterior to MC</td>
<td>Single outlet-sorta; pulmonary atresia</td>
<td>A</td>
<td>Subaortic</td>
</tr>
</tbody>
</table>

Abbreviations: AV, atrioventricular; A, anterior; R, right; P, posterior; L, left; UV, univentricular; RV, right ventricle; LV, left ventricle; DORV, double outlet right ventricle; PS, pulmonary stenosis; VSD, ventricular septal defect; ASD, atrial septal defect; IVC, inferior vena cava; SVC, superior vena cava; MC, main chamber; OC, outlet chamber
Dextrocardia

<table>
<thead>
<tr>
<th>Aortic arch</th>
<th>Associated anomalies</th>
<th>Visceral situs</th>
<th>Thoracic situs</th>
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<tr>
<td>Right</td>
<td>Ventricular septal defect</td>
<td>Solitus</td>
<td>Solitus</td>
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<tr>
<td>Left</td>
<td>Ascending aorta gives R-L pulmonary arteries; ductus—descending aorta</td>
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<td>Solitus</td>
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<tr>
<td>Left</td>
<td>Ventricular septal defect; pulmonary stenosis</td>
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<tr>
<td>Right</td>
<td>Persistent ductus arteriosus</td>
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<td>Left</td>
<td>Pulmonary stenosis</td>
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<tr>
<td>Right</td>
<td>Atrial septal defect; pulmonary stenosis</td>
<td>Solitus</td>
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<tr>
<td>Left</td>
<td>Left juxtaposition of atrial appendages; pulmonary stenosis</td>
<td>Solitus</td>
<td>Solitus</td>
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<tr>
<td>Left</td>
<td>Patent foramen ovale; persistent ductus arteriosus</td>
<td>Solitus</td>
<td>Solitus</td>
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<tr>
<td>Right</td>
<td>Severa pulmonary stenosis</td>
<td>Solitus</td>
<td>Solitus</td>
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<tr>
<td>Left</td>
<td>Persistent ductus arteriosus</td>
<td>Solitus</td>
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<tr>
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<td>Aneurysm of membranous septum</td>
<td>Inversus</td>
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<tr>
<td>Left</td>
<td>Severe pulmonary stenosis</td>
<td>Inversus</td>
<td>Inversus</td>
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<tr>
<td>Right</td>
<td>Supravalvular stenosis; persistent ductus arteriosus</td>
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<td>Inversus</td>
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<td>Left</td>
<td>PDA; VSD; left SVC</td>
<td>Inversus</td>
<td>Inversus</td>
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<tr>
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<td>ASD; right juxtaposition of atrial appendages; left IVC-azygos-left SVC</td>
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<tr>
<td>Right</td>
<td>Ventricular septal defect</td>
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<td>Right</td>
<td>Pulmonary stenosis</td>
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<td>Right</td>
<td>Right SVC; ASD</td>
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<tr>
<td>Right</td>
<td>Severe pulmonary stenosis</td>
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<td>Pulmonary stenosis</td>
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<td>Right</td>
<td>VSD; ASD; severe PS; left IVC—azygos—left SVC</td>
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<td>Right liver, left stomach</td>
<td>L ambiguus</td>
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<td>Left</td>
<td>Midline liver, left stomach</td>
<td>L ambiguus</td>
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<td>Left</td>
<td>Right liver, left stomach</td>
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<td>Left</td>
<td>Midline liver, left stomach</td>
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<td>Left</td>
<td>Asplenia, midline liver, left stomach</td>
<td>R ambiguus</td>
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<tr>
<td>Left</td>
<td>Asplenia, left-midline liver, left stomach</td>
<td>R ambiguus</td>
<td>R ambiguus</td>
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<tr>
<td>Left</td>
<td>Persistent ductus arteriosus</td>
<td>Solitus</td>
<td>Solitus</td>
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Bitted discordant arterial connections and the other of left ventricular type had double outlet main chamber. One of the univentricular hearts of right ventricular type had double outlet main chamber, the other two having single outlet with pulmonary atresia. Of the univentricular hearts of indeterminate type, two had double outlet and the other single outlet with pulmonary atresia.

**Aortic arch**
The aortic arch was left sided in 11 patients (70%) and right sided in five (30%).

**Associated anomalies**
Persistent ductus arteriosus, ventricular septal defect, and pulmonary stenosis were the commonest anomalies. Two patients had left juxtaposition of the atrial appendages.

**Situs Inversus**
In all cases we found visceral, thoracic, and atrial situs to correspond.

**Atrioventricular junction (Fig. 4)**
Eleven patients had situs inversus, of whom five had concordant and four discordant atrioventricular connections. The remaining two patients had double inlet ventricles, in both of which the ventricular morphology was of a univentricular heart of right ventricular type with a posterior right-sided trabecular pouch of left ventricular type.

The right ventricle, in the nine cases with two ventricular chambers, was to the left of the left ventricle in five (all with concordant atrioventricular connections) and to the right in four (with discordant atrioventricular connections).

**Arterial junction**
The ventriculoarterial connection in the five patients with concordant atrioventricular connections was concordant in one, double outlet right ventricle in three, and single outlet in the other (aorta with pulmonary atresia). Of the four patients who had discordant atrioventricular connections, one had concordant ventriculoarterial connections (Fig. 5). This chamber arrangement has been termed 'isolated atrial inversion' by other workers (Clarkson et al., 1972), and was confirmed at operation in our case. Of the remaining patients, two had discordant ventriculoarterial connections and one double outlet right ventricle. In both the patients with univentricular hearts the arterial connection was double outlet from the main chamber.

In the group as a whole the aorta was anterior to the pulmonary artery in two cases, anterior and to
the right in four cases, anterior and to the left in three, posterior and to the left in one, and posterior in one.

**Aortic arch**
The aortic arch was right sided in eight patients (73%) and left sided in three (27%).

**Associated anomalies**
Pulmonary stenosis and ventricular septal defect were the commonest lesions. The patient with discordant atrioventricular connections and discordant ventriculoarterial connections (isolated atrial inversion) had right juxtaposition of the atrial appendages with azygos continuation of the inferior vena cava to the left superior vena cava (Fig. 5).

**Situs ambiguus, laevoisomerism**
In four patients atrial laevoisomerism was suggested by the bronchial anatomy on the chest x-ray film, confirmed by necropsy in one (Fig. 6). In two of these patients the liver was central (visceral situs ambiguus) but in two the liver was right sided (visceral situs solitus).

**Atrioventricular junction (Fig. 7)**
Three patients had two ventricles and one a univentricular heart of indeterminate type (without rudimentary chamber). In the biventricular hearts the right ventricle was to the right in one case and to the left in two cases.

The mode of connection was through a common atrioventricular valve in three cases and via two perforate atrioventricular valves in one case.

**Arterial junction**
Of three patients with two ventricles, one had concordant ventriculoarterial connection, one double outlet right ventricle, and one single outlet (truncus arteriosus). The remaining patient had double outlet from the sole indeterminate ventricular chamber.

The aorta was anterior and to the right in one case, anterior and to the left in one case, and to the left in one case. The remaining case had a truncus with the aortic component posterior to the pulmonary component.

**Aortic arch**
The aortic arch was left sided in three patients and right sided in one.
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Fig. 6  Posterior view of specimen with laevoisomerism dissected to show bilateral hyparterial (morphologically left) bronchi. Both lungs were bilobed and atrial appendages were of left morphology. The bronchi are hyparterial as both upper lobe bronchi (ULB) pass below lower lobe pulmonary arteries (LLPA).

Associated lesions
Three patients had azygos continuation of the inferior vena cava. One patient had left juxtaposition of the atrial appendages (Fig. 8).

Situs inversus, dextroisomerism
In two patients atrial dextroisomerism was present, as suggested from the chest x-ray film and confirmed at necropsy in both. Both also had asplenia. In one the liver was central (visceral situs ambiguus) but in the other the liver was left sided (visceral situs inversus).

Atrioventricular junction (Fig. 9)
Two ventricles were present in one patient. The other had a univentricular heart of left ventricular type with outlet chamber of right ventricular type which was anterior and to the left of the main chamber.

Arterial junction
The patient with two ventricles had double outlet right ventricle with the aorta directly anterior, and the patient with a univentricular heart had single outlet (pulmonary atresia), the aorta arising from the outlet chamber.

Aortic arch
Both patients had a left-sided aortic arch.

Associated anomalies
Partial anomalous pulmonary venous return was found in one patient.

Discussion
Our study endorses the findings of others (Van Praagh et al., 1964; Squarcia et al., 1973) that a variety of chamber combinations can be found in hearts situated in the right chest with their long axis orientated to the right. We concur, therefore, with the opinion that the term dextrocardia conveys no information regarding chamber organisation and internal anatomy of the heart but should be used only for description of this cardiac position, a feature easily discernible from the plain chest radiograph. In this investigation our cases not only had the greater part of the heart within the right hemithorax but also had the long axis and apex directed to the right. It is possible that cases were not included that others might have categorised
as dextrocardia. However, as we have indicated, arguments concerning the precise definition of dextrocardia are unproductive, the important feature being the segmental arrangement in a heart. In this respect only two of our cases had 'normal' chamber arrangements, compared with 30 per cent of those analysed by Van Praagh et al. (1964), though only 7 per cent of those studied by Lev et al. (1968) had normal chamber connections. It is possible that these differences reflect the mode of selection of cases. Thus, as shown by the earlier investigations, it is essential to utilise a segmental approach in order adequately to diagnose and classify the cases. We have not, however, found previous approaches (Van Praagh et al., 1964; Squarcia et al., 1973; Van Praagh and Vlad, 1978) to be universally applicable to the anomalies encountered. In contrast, utilisation of the system advocated by Tynan et al. (1979) has enabled all hearts to be described simply and unambiguously.
The hearts investigated here highlight the deficiencies in diagnostic approaches which do not distinguish clearly between anatomical connections and spatial relations.

In the hearts we examined, atrial situs was determined from bronchial anatomy on plain chest radiography. In this way not only could situs solitus and inversus be identified but the laevoisomeric and dextroisomeric forms of situs ambiguous could be distinguished. Atrial situs was confirmed in the cases coming to necropsy. We believe this method of categorising situs to be preferable to those relating atrial morphology to the state of the spleen. Furthermore, if there is doubt from the plain film, bronchial morphology can easily be confirmed by tomography (Partridge et al., 1975) and there is better correlation between atrial and bronchial anatomy than between atrial morphology and presence or absence of splenic tissue or multiple spleens (Macartney et al., 1978).

When describing the ventricular segment of the heart, we found it necessary to be able to describe connections of the atria to the ventricles independently both of spatial relations of chambers within the ventricular mass and of the morphology of the ventricular chambers. Thus, in our cases we found all five types of atrioventricular connection considered possible by Tynan et al. (1979), namely concordant, discordant, ambiguous, double inlet, and absence of one atrioventricular connection. In the last category absent right but not absent left atrioventricular connection was seen. The provision of the category for ambiguous connection was necessary to describe the connections in the hearts with situs ambiguous and two ventricles, as the terms concordance and discordance are inappropriate for this arrangement (Tynan et al., 1979). Our cases also illustrated the usefulness of describing type of connection separately from mode of connection, since the ambiguous connection was effected through a common valve in five cases, but through separate right and left valves in one case.

These cases also serve to illustrate what we believe to be a further refinement (Shinebourne et al., 1978) of our initial approach to this connection (Shinebourne et al., 1976). Following Van Praagh's lead, we initially described ventricular relations with ambiguous connection in terms of the loop, as was recently commented upon by Freedom et al. (1978). However, because of the additional connotations of 'd' and l-loop', we now prefer to describe the ventricular relations simply in terms of right/left and antero/posterior orientation. One reason for this is the possibility of a 'criss-cross' heart (Anderson et al., 1974) with an ambiguous connection. Previously in a criss-cross heart, we had used 'd-loop' to describe the relation where the morphologically right ventricle was right sided. However, if a criss-cross situation were found with an ambiguous connection, would the relation be described as 'd-loop' or 'l-loop'? As Freedom et al. (1978) indicate, the current concept of the loop (Van Praagh and Vlad, 1978) makes it independent of situs. As such, in situs ambiguous it is simply being used to describe ventricular relations. Thus, in the criss-cross situation the left-sided right ventricle, having achieved its position as a consequence of cardiac rotation, can still be recognised as belonging to a d-loop. Is it not, however, simpler to describe the ventricular position in terms of right/left and antero/posterior position rather than asking the observer to interpret the presumed embryological looping of the heart tube? We also submit that simple usage of the right/left and antero/posterior co-ordinates is more useful and more accurate than the terms 'inversion' and 'non-inversion'. The 'inverted' ventricles of classical corrected transposition in situs solitus are rarely, if ever, the mirror image of the normal ventricular arrangement: in the presence of atrioventricular discordance the ventricles tend to be more side by side with horizontal inclination. Furthermore, there is no consensus as to what 'ventricular inversion' describes. For some authors, 'ventricular inversion' in situs inversus describes ventricles occupying the normal position for the solitus individual with a normal heart, with the right ventricle anterior and to the right (Espina-Vela, 1978). Such different interpretations are avoided by the use of simple adjectives such as right, left, anterior, and posterior.

The value of describing ventricular morphology independently of the connection is well illustrated by our cases with absence of the right atrioventricular connection ('tricuspid atresia'). These hearts illustrate the wide variability found in hearts having the atrial morphology of classical tricuspid atresia, a possibility predicted in our previous studies (Anderson et al., 1977). Thus, univentricular hearts of right ventricular type, left ventricular type, and indeterminate type were found with absent right atrioventricular connection. We find it difficult to describe these hearts using the loop concept. The finding of such variability in ventricular morphology in hearts with 'tricuspid atresia' has considerable surgical significance. Rudimentary chambers may be posterior or anterior, right sided or left sided, or absent. Most posterior chambers are not suitable for incorporation into the circulation in 'corrective' procedures. We also do not know the effect that ventricular morphology will have on the possible outcome of a Fontan procedure, and clearly this is another factor which must be considered when...
assessing these cases for possible operation (Fontan et al., 1978).

It is of note that several hearts in our series had the morphology of a main chamber of right ventricular type with a rudimentary chamber of left ventricular type. Five cases were identified (16.5%), three with double inlet connection, and two with absence of the right atrioventricular connection. We believe that in part this relatively high prevalence is the result of an awareness of the anomaly, since we are now finding this malformation with increasing frequency in hearts both with and without dextrocardia (Keeton et al., 1979). Again, identification of this type of ventricular morphology in the univentricular heart is important since it affords a good guide to the likely disposition of the conducting system (Anderson et al., 1978).

Juxtaposition of the atrial appendages was not infrequent in our series. Otero Coto et al. (1978) have also pointed to an association between right-sided apex and juxtaposition with a left-sided aorta. However, the aorta was left sided in only one of our cases. All the cases were identified during life, a possibility discussed by Deutsch et al. (1974).

At the arterial junction we again found categorisation to be facilitated by describing separately the connections, the arterial relations, and infundibular morphology. It seems to us that if this is done there can be little room for controversy concerning such matters as ‘transposition’ or the ‘conus’.

In conclusion, we have studied a series of patients whose hearts were within the right hemithorax with their long axes orientated to the right, this being the only information to be drawn from the term ‘dextrocardia’. The identification and categorisation of the extremely varied chamber arrangement within these hearts was greatly facilitated by use of a descriptive system accounting separately for connections, relations, and morphology.

References


Dextrocardia


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