Dextrorotation of the Heart

An Angiocardiographic Study of Forty-one Cases

By Stephen M. Ayres, M.D., and Israel Steinberg, M.D.

Misalignment of the heart in the right hemithorax has interested physicians ever since Severinus of Rome in 1643 first observed dextrocardia in complete situs inversus.1 Unfortunately the literature has been confused by the inclusion of all cases of right-sided hearts under the term dextrocardia with further grouping based on the presence or absence of situs inversus or associated cardiac malformations. Thus hearts with completely different morphologic and developmental attributes have been lumped together merely because of location in the right hemithorax. Although many classifications have been devised, the analysis of congenital dextrocardia is greatly simplified by differentiation into two main groups: Mirror image dextrocardia—the classical dextrocardia—is almost always associated with complete situs inversus and is rarely complicated by additional cardiac anomalies.2 The cardiac silhouette is a perfect mirror image; if the chest roentgenogram be reversed or the patient fluoroscoped from the rear, completely normal relationships are observed. Dextrorotation or dextroversion, a completely different entity both morphologically and embryologically, is not associated with situs inversus and frequently is complicated by additional cardiac malformations. The confusing term “isolated dextrocardia” has often been applied to this type of right-sided heart.

Angiocardiography, which permits the precise identification of cardiac chambers and analysis of their in-vivo position in the thorax, is the method of choice in the study of right-sided hearts. This paper is an angiocardiographic study of 41 patients fulfilling the morphologic criteria for the diagnosis of dextrorotation of the heart.

Methods and Materials

Venous angiocardiography was performed by technics previously described.3 Forty-one patients, 19 female and 22 male, ranging in age from 3 weeks to 51 years, were found to have dextrorotation of the heart. All had complete clinical and laboratory evaluations, which included conventional roentgenographic studies. Electrocardiograms were available for analysis in 33. Cardiac catheterization was performed in 12 cases to evaluate associated congenital malformations.

Results

The morphologic features of dextrorotation uncomplicated by associated malformations are depicted in figures 1 and 2. The longitudinal plane of the heart forms an angle of 30 to 45° with the midsagittal plane of the thorax in the transverse plane of the body. This may be simulated by rotating a normal heart into the left anterior oblique position. In addition the entire heart is displaced to the right, so that the longitudinal axis of the heart forms an angle of 0 to 50° in the frontal plane. This displacement may be simulated by fixing the great vessels and swinging the heart to the right in the frontal plane (like a pendulum). As a result of this rotational anomaly the two ventricles lie side by side; the right ventricle has actually become the “right” ventricle instead of the anterior ventricle, the left ventricle is now the “left” ventricle instead of the posterior ventricle. The position of the atria is almost normal; the right atrium is posterior to the right ventricle, the superior vena cava is more medial than usual, and the left atrium is slightly anterior to the right atrium (fig. 3). The great vessels arise normally but the rotation to the right

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opens the loop between the ascending and descending aorta. The main pulmonary artery is displaced medially.

The degree of frontal plane displacement varies, but is usually more marked in patients with associated congenital cardiac defects and right ventricular hypertrophy. Figure 3 depicts marked rightward displacement in a patient with pulmonic stenosis and right ventricular hypertrophy. The right-to-left atrial shunt in this patient produces simultaneous filling of both atria and thus fortuitously demonstrates their relative positions. In contrast, many of the patients with normal hearts have little rightward displacement in the frontal plane although the ventricles lie side by side in the transverse plane. This midline location of the heart within the thorax has been called mesoversion but is actually a lesser degree of dextrorotation.

Three quarters of the patients studied had additional congenital cardiac anomalies (table 1). Anomalous drainage of pulmonary veins was observed in 10, atrial septal defects in seven, hypoplasia of a main pulmonary artery in six. In eight patients the aorta arose anteriorly and lateral to the posterior and medial pulmonary artery; the ascending aorta produced a prominent shadow along the left upper cardiac border and then turned medially and descended normally. This orientation of the great vessels is usually associated with ventricular inversion (the venous ventricle is a morphologic left ventricle and bears a bicuspid atriocvalvalve, the arterial ventricle is a morphologic right ventricle with tricuspid valve) and has been termed corrected transposition of the great vessels. This produces a narrow aortic loop which contrasts with the wide aortic loop usually seen in dextrorotation. Figure 4 demonstrates this anomaly of the great vessels; postmortem examination demonstrated ventricular inversion.

A confusing triad of mirror-image dextrocardia and situs inversus, corrected transposition of the great vessels, and dextrorotation was observed in two patients. The basic pattern of mirror-image dextrocardia—venous atrium and venae cavae on the left, apex on

### Table 1

<table>
<thead>
<tr>
<th>Associated Congenital Anomalies in 41 Cases of Dextrorotation</th>
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<tbody>
<tr>
<td>No cardiac anomalies</td>
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<tr>
<td>Associated cardiac anomalies</td>
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<tr>
<td>Corrected transposition of great vessels</td>
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<tr>
<td>Anomalous drainage of pulmonary veins</td>
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<td>Hypoplasia or atresia of pulmonary artery</td>
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<tr>
<td>Atrial septal defect</td>
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<td>Ventricular septal defect</td>
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<td>Left superior vena cava</td>
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<td>Pulmonic stenosis</td>
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<tr>
<td>Tetralogy of Fallot</td>
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<tr>
<td>Mirror-image dextrocardia</td>
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<tr>
<td>Associated extracardiac anomalies</td>
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<tr>
<td>Eventration of right or left diaphragm</td>
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<tr>
<td>Pectus excavatum</td>
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<tr>
<td>Coarctation of aorta</td>
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<tr>
<td>Aberrant aortic branch to lung</td>
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<tr>
<td>Bronchial abnormalities</td>
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<tr>
<td>Absent pulmonary lobulation</td>
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<tr>
<td>Absent manubrium sterni</td>
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<tr>
<td>Hamartoma of lung</td>
</tr>
<tr>
<td>Scoliosis</td>
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<tr>
<td>Undescended testicle</td>
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<tr>
<td>Fetal lobulation of kidney</td>
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</tbody>
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* Circulation, Volume XXVII, February 1963
the right—was complicated by rotation in the frontal and transverse plane typical of dextrorotation. Since the right-sided heart of mirror-image dextrocardia was actually rotated to the left (the opposite of dextrorotation in a normally situated heart) this anomaly should correctly be termed mirror-image dextrocardia with levorotation or "levocardia." Visualization of this anomaly is enhanced if the chest roentgenogram is reversed; the mirror-image dextrocardia is eliminated and the cardiac silhouette resembles dextrorotation with corrected transposition in a normally situated heart.

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In all but one case the P wave in the electrocardiogram was upright in lead I, providing a clear differentiation from mirror-image dextrocardia. In the latter anomaly, the P wave in lead I is always inverted, since the electrical forces are directed away from the left arm. The mean P axis varied from 135° to −75°; the QRS axis varied from 150° to −165° and was more related to the associated cardiac anomalies than to the dextroversion (fig. 5). The height of the R wave in lead V1 varied from 5 to 15 mm. in the nine patients with normal hearts, thus giving rise to the erroneous impression of right ventricular hypertrophy. In most of the cases a large R wave and a deep S wave were present in V1 regardless of the state of the underlying myocardium. In none of the cases in which a diastolic overload pattern of the right ventricle might be predicted was an rSR' pattern observed.

Discussion

In the nineteenth century European morphologists described many examples of right-sided hearts with and without situs inversus. Mandelstamm and Reinberg divided the dextrocardias into four main groups: (1) mirror-image dextrocardia in association with situs inversus, (2) mirror-image dextrocardia without situs inversus, (3) dextrocardia with normal arrangement of chambers, (4) acquired dextrocardia. The term "isolated" dextrocardia was used by subsequent authors to indicate dextrocardia without situs inversus but could also designate dextrocardia without associated cardiac anomalies. Paltauf, in 1901, differentiated dextrocardia with inversion of chambers (mirror-image dextrocardia) from dextrocardia with normal position of chambers and applied the term dextroversion to the latter group. In 1931, Lichtman presented two new cases of isolated dextrocardia and exhaustively reviewed the literature. He accurately described the morphology of isolated dextrocardia, pointed out that the defect was a rotational arrest occurring in the sixth week of gestation, and demonstrated the common association of transposition of the great vessels. It seems clear that Paltauf's dextroversion, Lichtman's isolated dextrocardia, and Mandelstamm and Reinberg's type 3 dextrocardia are morphologically the same anomaly.

Interest was rekindled in this anomaly by the extensive studies of Korth and Schmidt in 1953. These authors reviewed 1,000 cases of mirror-image dextrocardia, observed situs inversus in all, and congenital heart anomalies in only 1. In contrast, 53 of 59 cases of dextroversion were associated with congenital cardiac defects. While situs inversus was not present in this group, skeletal anomalies, mal-

Figure 3

Marked displacement to the right in a patient with dextroversion, pulmonic stenosis, and right ventricular hypertrophy (left). A right-to-left atrial shunt demonstrates the relative position of the atria and the medial displacement of the superior vena cava (center). Left heart filling shows the position of the ventricular septum and the characteristic side-by-side arrangement of the ventricles (right).
rotation of the gut, and asplenia were common. Mirror-image dextrocardia without situs inversus was not found. Numerous case reports of complex congenital cardiac anomalies associated with right-sided hearts have appeared in the last decade.\textsuperscript{8}-\textsuperscript{12} Some authors\textsuperscript{13, 14} have further subdivided the "isolated" dextrocardias into dextroversion, dextroposition, and extrinsic dextrocardia. The presence of associated chest wall or pulmonary anomalies has frequently been considered sufficient evidence for classification as extrinsic dextrocardia regardless of the actual cardiac anatomy. This paper attempts to simplify the classification of right-sided hearts; dextrorotation is considered a morphologic entity and all hearts fulfilling the morphologic criteria are so classified regardless of associated thoracic anomalies.

Dextrorotation is a morphologic entity produced by a specific defect in embryogenesis. During the sixth week of fetal life the heart begins to attain its mature external configuration and descends from the cervical region to its permanent location in the thorax. At the same time it rotates into the left hemithorax. Important changes occur at the same time. The spiral conotruncal septum separates pulmonary artery from aorta and the primitive bulbus becomes absorbed into the right ventricle, the ventricular and atrial septa become differentiated, the sinus venosus and systemic veins become part of the right atrium, and the pulmonary veins unite with the left atrium. In addition, the splenic anlage is formed, the septum transversum reaches its definitive position and unites with the pleuropertitoneal membrane to form the diaphragm, the cartilaginous sternum develops, and the primitive lung buds subdivide into the bronchial tree. This cluster of important embryonic events between the sixth and eighth weeks of fetal life makes this period an especially vulnerable time for the inception of multiple congenital anomalies. A teratogenic agent acting at this time may prevent the primitive heart from reaching its definitive position in the left hemithorax and at the same time produce both cardiac and extracardiac malformations.

These developmental considerations suggest that the extracardiac anomalies frequently seen in dextrorotation may not be mechanical causes of the malrotation but rather are associated malformations probably resulting from the same teratogenic insult. If these extracardiac defects were mechanical causes of dextrorotation, that malrotation would be commonly observed with these extracardiac anomalies.
DEXTOROTATION OF THE HEART

defects. Dextrorotation, however, is rarely seen with pectus excaevatum and is not frequent in eversion of the diaphragm. Dextrorotation has been observed with eversion of either the left or right diaphragm. It is difficult to see how both defects could produce an identical malrotation of the heart if the cause were solely mechanical.

Spitzer pointed out early that transposition of the great vessels was common in dextrorotation.15 Grant16 confirmed this and demonstrated that this was not the passive result of the rotation, since this would open the aortic loop. Areilla and Gasul,17 however, considered that transposition was usually incorrectly diagnosed in this anomaly and suggested that the aorta always originated to the left and anterior to the pulmonary artery in dextrorotation. Eight patients in the present series and one illustrated by Grant had the conotruncal anomaly first described by Rokitansky and more recently by Anderson, Lester, and Lillehei4 as corrected transposition of the great vessels. The important differential point is evident on the frontal angiocardiogram: with normal aortic root the loop is wide and the ascending aorta arises convexly to the right (figs. 1 and 2), in corrected transposition the ascending aorta arises convexly to the left and forms a prominent portion of the left upper cardiac border (fig. 4). An explanation for the common association of corrected transposition and dextrorotation may be found in the theory of de la Cruz and associates,17 who postulated that corrected transposition in a normally situated heart results from complete inversion of the bulboventricular loop and failure of the loop to rotate into the right hemithorax. They suggest that if the inverted loop rotates into the right hemithorax, dextrorotation and corrected transposition result. If this explanation be correct, the combined defect of dextrorotation and corrected transposition results from a teratogenic agent acting on the embryonic heart before the straight cardiac tube develops its sigmoid twist (third week of fetal life).

Summary

Dextrorotation of the heart is a morphologic entity characterized by rotation to the right in both frontal and transverse planes of the body; the relative location of the cardiac chambers is normal. In contrast to mirror-image dextrocardia, which results from a defect in lateralization of the entire embryo occurring in the first 10 days of fetal life, dextrorotation results from a teratogenic agent acting in the sixth fetal week. More than 75 per cent of the patients with dextrorotation in this series had associated congenital cardiac anomalies. Corrected transposition of the great vessels was present in eight of 41 patients. A normal electrocardiogram provided a clear differentiation from mirror-image dextrocardia; angiocardiology permitted precise analysis of the in-vivo morphology.

Addendum

Since submission of this article for publication, Mirowski and associates18 have presented a case of dextrorotation of the heart with negative P waves in lead I. This was observed in one of our cases; negative P waves in lead aVL were present in six. Medial displacement of the superior vena cava and sinoatrial node may be the explanation (fig. 3).

References

6. MANDELSTAMM, M., AND REINBERG, S.: Quoted by Lichtman.1
7. PALTAUF, R.: Quoted by Lichtman.1
15. Spitzer, A.: Quoted by Lichtman.1

Professional Ethos

I hold every man a debtor to his profession; from which as men of course do seek to receive countenance and profit, so ought they of duty to endeavour themselves, by way of amends, to be a help and ornament thereunto. This is performed, in some degree, by the honest and liberal practice of a profession; where men shall carry a respect not to descend into any course that is corrupt and unworthy thereof, and preserve themselves free from the abuses wherewith the same profession is noted to be infected; but much more is this performed, if a man be able to visit and strengthen the roots and foundation of the science itself; thereby not only gracing it in reputation and dignity, but also amplifying it in profession and substance.—Francis Bacon (Essayes). The Quiet Art: A Doctor's Anthology. Compiled by Dr. Robert Coope. Edinburgh & London, E. & S. Livingstone Ltd., 1952, p. 204.
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