The Syndrome of Dextroversion of the Heart

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On the basis of 3 personal cases and 119 additional cases collected from the literature the difference between dextroversion and mirror-image dextrocardia is outlined. It is concluded that dextroversion is a part of an exceedingly primitive arrest in development that frequently includes other thoracic and abdominal organs as well as intracardiac structures. The defect is altogether different in embryogenesis and anatomic findings from mirror-image dextrocardia.

There are 3 conditions that are characterized by dextrocardia, or right-lying heart. The commonest and most familiar is mirror-image dextrocardia, in which the anterior-posterior relationships of the various parts of the heart are normal but their right-to-left orientation is reversed. It is usually easily recognized because it is practically always associated with some degree of abdominal situs inversus, rarely are there any other cardiac abnormalities, and the electrocardiographic changes it produces are diagnostic, with inverted P, QRS, and T waves in lead I. The second cause of dextrocardia is dextroposition, in which an otherwise normal heart is shifted to the right by some extracardiac factor such as evagination of the left diaphragm, fibrosis of the right lung, etc., and therefore is also usually easily recognized. The third type is dextroversion. It is the least familiar, but perhaps the most important of the 3 from the clinical point of view because it is frequently accompanied by other intracardiac abnormalities, often of a serious nature. Furthermore, the embryogenesis of dextroversion is closely related to the embryogenesis of the cono-truncal region of the heart, and understanding the mechanism of dextroversion sheds light on other congenital malformations.

Dextroversion has been recognized under several different names in the past: "isolated" dextrocardia, incomplete rotation of the heart, dextrotorsion, etc. However, the recent comprehensive studies by Korth and Schmidt have made it clear that it is a distinctive syndrome with broader clinical and embryologic implications than these names imply, justifying the special term dextroversion. In general, dextroversion consists of a rotation of the ventricular part of the heart to the right, as in turning the page of a book, with the atria remaining in normal position. Usually there are transposition of the great vessels and a ventricular septal defect. Since the atria are in normal position, the direction of spread of atrial depolarization is the same as in the normal subject and therefore the P wave in lead I is upright, differentiating it from mirror-image dextrocardia. The QRS and T waves in dextroversion depend upon the type and degree of associated intracardiac malformation. If there is no significant associated abnormality, the mean T vector is often rightward, producing a negative T and the QRS loop is counterclockwise in the frontal plane, producing a Q. In the past the negative T has often been attributed to ischemia; but, as shown in figure 1, it is likelier that it is due to rotation of the ventricular electric field on its long axis, for the QRS-T angle is usually a narrow one, as in the normal heart. A more detailed description of the certain aspects of the syndrome will follow the presentation of 3 selected cases, 2 of which were studied by biplane angiocardiology and 1 at postmortem examination.

Case 1. D.D. (Clinical Center #016355): a 2-year-old girl had been cyanotic with episodes of labored breathing since birth. There were no signs of congestive heart failure. The heart was not...
enlarged, lay to the right, and a high pitched grade III systolic murmur was noted over the entire precordium, maximal in the second interspace to the right of the sternum. The abdominal organs appeared to be normally placed. The hematocrit value was 83 per cent (Wintrobe). In the electrocardiogram the P waves were upright in all 3 limb leads, the mean QRS vector was directed superiority and to the right with QS deflections in the 3 standard limb leads and conventional V₃ to V₅. The mean T vector was directed inferiorly. On cardiac catheterization the catheter was passed through the venous right-lying atrium into both an arterial and a venous ventricle. The pressure in the venous ventricle was slightly higher than in the arterial ventricle; neither the aorta nor pulmonary artery could be entered. On selective angiocardiography with the catheter in one of the ventricles (fig. 2) there was immediate filling of both ventricles, with a small jet of radiopaque medium extending into the pulmonary artery (first film). At ¾ second the pulmonary artery was well filled, showing narrowing proximally, and the contrast material could be seen regurgitating slightly into both atria. At 1½ seconds ventricular diastole had occurred, with marked reduction in the degree of opacification of the right ventricle and a central area of translucency in the left ventricular shadow due to the entrance of nonopacified blood from the atria. The difference in opacification between the right and left ventricles at this stage demarcated the ventricular septum, which was more or less parallel with the horizontal or transverse plane of the body. At 1¾ seconds the ventricles had contracted and the aorta was opacified for the first time, showing it to lie anteriorly to the pulmonary artery. In later films (not shown) the pulmonary veins were opacified and could be seen draining to the lower left region of the cardiac silhouette, indicating that the left atrium was normally placed. The gastric bubble appeared normally placed. The diagnosis was dextroversion, with atrial and ventricular septal defects and transposition of the great vessels.

Case 2. J.W. (Clinical Center #017046), a 5-month-old girl, had been cyanotic and slightly short of breath since birth. There were no murmurs or signs of decompensation. Hemoglobin was 15 Gm. per cent, with many Howell-Jolly bodies and a few spherocytes in the peripheral blood smear. Her electrocardiogram showed upright P waves in the 3 standard limb leads; the mean QRS axis was leftward and inferior in direction with qR deflections on leads I and II and principally upright QRS complexes of QR type from V₃ to V₅. During her episodes of dyspnea the mean QRS vector gradually but transiently rotated to marked left axis deviation without pro-

Fig. 1. A, the normal heart, B, classical mirror-image dextrocardia, C, classical dextroversion. The position of the 4 cardiac chambers, the great vessels, and the gastric gas bubble are shown for each. Below, standard limb leads and the frontal plane projections of mean cardiac vectors in the uncomplicated case are shown. Note the difference in cardiac silhouette between mirror-image dextrocardia and dextroversion. In the latter, the apex is blunter, there is a prominent shadow high on the right side due to the right atrium, and the transposed aorta forms a prominent slow curve on the upper left side of the silhouette in the typical case.
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Fig. 2. Simultaneous lateral (top) and anterior-posterior (bottom) biplane angiocardiogram in case 1. The opacification for each film is diagrammed below it, and the time of the film after injection of the contrast material is indicated at the bottom of the figure. The position of the catheter through which the contrast material was injected is shown. Both ventricles fill in the first film, demonstrating the presence of a ventricular septal defect. In the second and third anterior-posterior films, the plane separating the right and left ventricles is well defined, indicating that the ventricular septum lay more or less perpendicular to the frontal plane of the body, with the right ventricle superior to the left ventricle. In the fourth lateral film it can be seen that the aorta is anterior to the pulmonary artery, demonstrating the presence of transposition of the great vessels; usually the aorta has a more leftward position than in this case. In still later films (not shown) the passage of contrast material from the pulmonary veins into a chamber to the left of the left ventricle could be seen, indicating that the left atrium was normally located.
The heart in case 2, viewed frontally as it lay in the chest. Right, schema of the internal structure of the heart from the same view. The interventricular septum, represented by only a shelf, is perpendicular to the frontal plane of the body; the right ventricular portion of the heart lies superiorly to the left ventricle. The muscular ridge at the proximal end of the tissues separating aorta and pulmonary artery is interpreted to be the crista supraventricularis; it is more or less parallel with the frontal plane of the body, which is its normal anterior-posterior position, and in this heart it is at right angles to the position of the ventricular septum.

At autopsy (performed by Dr. Louis Thomas) abnormalities of thoracic and abdominal organs were found. The heart lay principally rightward with the posterior sulcus (which normally faces the diaphragm and contains the descending branch of the right coronary artery) facing anteriorly, indicating that the right ventricle was superior to the left (fig. 3). The heart was somewhat enlarged, measuring 5.5 cm. in its long axis, with right and left ventricular walls 5 mm. thick. There were 4 venae cavae. The 2 superior venae cavae lay on each side of the mediastinum, one entering the right, the other the left side of a single large atrial chamber, which had only a few strands of atrial septal tissue. These strands indicated that the septum was more or less parallel with the frontal plane of the body, its normal position. The identification of right versus left atrial structures was not possible, but in view of the normally directed P vector in the electrocardiogram, the sinus node lay rightward, indicating that the atria were normally placed. One inferior vena cava drained solely the left lobe of the liver and entered the left side of the atrial chamber. The other inferior vena cava entered into the right side of the common atrium, draining the right lobe of the liver, the kidneys, adrenal glands, pancreas, and lower extremities with only minor variations from the normal in its architecture. The ventricular portion of the heart consisted of essentially a single chamber, with a rudimentary ledge at its lateral and inferior border suggesting a ventricular septum. A single A-V orifice connected the common atrium and common ventricle. It had 3 leaflets that were incompletely developed and separated. There was a prominent muscular ridge in the outflow region of the common ventricle that separated the aortic and pulmonary orifices. Both orifices had 3 well-formed cusps. The coronary artery orifices were normally placed, and the coronary arteries had a normal distribution over the ventricular myocardium. The pulmonary artery was anterior and to the left of the root of the aorta, and was 14 cm. in diameter while the aorta was 1.0 cm. A completely obliterated ligamentum arteriosum connected the 2 vessels. The branchings and distributions of the pulmonary artery and aorta were normal. The pulmonary veins from both lungs joined to form a common pulmonary vein that entered the right superior vena cava. The right lung had 3 lobes, and a prominent denervation of the lingular part of the left lung gave it 3 lobes also. In the abdomen, the left lobe of the liver was larger than the right lobe and the gallbladder was attached to the left-
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lying lobe. The spleen was absent. The stomach lay in the left upper quadrant, its antral region deviated by a torsion in the position of the duodenum that was folded by its attachment to the left-lying gallbladder. The tail of the pancreas extended leftward but was not retroperitoneal and lay free in the abdomen. Both the lesser and greater omenta were incompletely developed, as were also the mesenteries of the small and large bowels. Counterclockwise rotation of the bowel had failed to take place, and the ileocecal valve, cecum and vermiform appendix lay in the left upper quadrant. The large bowel from the sigmoid to the rectum was normal in location. The portal venous system was normal. The diagnoses were dextroversion of the heart with atrial and ventricular septal defects, common pulmonary vein, single atrioventricular orifice, persistent right superior and inferior venae cavae, asplenia, partial situs inversus of the liver and gallbladder and incomplete development of other abdominal and thoracic organs.

Case 3. S.C. (Clinical Center #019515), an 8-year-old girl, was first noted to have a murmur at 3 months of age. Cyanosis had not been noted, but she complained of fatigability on exertion. On examination the arterial blood pressure was 90/60; there was no cyanosis, clubbing, or evidence of congestive heart failure. The maximal cardiac impulse was noted to be in the right fifth interspace, but abdominal organs appeared to be normally placed. An extremely loud, high-pitched diastolic murmur was noted along the left sternal border. Hemoglobin was 16 Gm. per cent. The electrocardiogram disclosed upright P waves in the standard limb leads, the mean QRS axis was directed rightward and inferiorly, with a qrS contour in lead I; the mean T vector was directed horizontally rightward, and the conventional pre-

FIG. 4. Angiocardiogram of case 3. The anterior-posterior view prior to injection of contrast material is shown on the left, illustrating the rightward position of the heart. Middle, a film selected from the angiocardiographic series to show the positions of the right ventricle and right atrium. Right, diagram of the opacification in this film. Later films outlined the aorta and pulmonary artery, shown by dotted lines in the diagram. The plane separating right from left ventricle has a more sagittal position in this case than in case 1. There was no transposition of the great vessels. Note the gastric gas bubble under the left leaf of the diaphragm. The catheter through which the contrast material was injected can also be seen.

FIG. 5. Topologic schemata of the surfaces separating arterial from venous parts of the heart; A, the normal heart, B, transposition of the great vessels in a normally placed heart, C, dextroversion without transposition (drawn from case 2; the atrial and ventricular septa were not completely developed in this heart, the undeveloped portions are represented by dotted lines), D, dextroversion with transposition of the great vessels. T, the plane tangential to both the pulmonary artery and the aorta; V, ventricular septum; A, atrial septum; C.S., crista supraventricularis.
cordial leads all showed RS contours with inverted T waves as far as V6. On right heart catheterization the pulmonary artery wedge pressure was normal, pulmonary artery pressure was 110/60, right ventricular pressure was 110/5, and right atrial pressure was normal. The arterial blood was 81 per cent saturated and both right-to-left and left-to-right shunts were detected at the level of the pulmonary artery. On angiocardiography (fig. 4) the pulmonary artery and aorta filled simultaneously with the proximal part of the aorta poorly visualized and the pulmonary artery extremely large in size. There was no evidence of transposition of the great vessels, and the aortic arch was left-sided. The atria were normally placed, but the right ventricle lay to the right of the left ventricle with the interventricular septum parallel with the sagittal plane of the body (fig. 5). The gastric bubble was normal in location. The diagnosis was mesosversion of the heart with patent ductus arteriosus. In view of the pulmonary hypertension and evidence of reversed flow through the ductus, surgery was postponed.

DISCUSSION

One hundred and nineteen cases of dextroversion have been reported in the literature, proved either by electrocardiogram or autopsy.* The electrocardiogram is diagnostic of this syndrome, for in the presence of a congenitally right-lying heart an upright P wave in lead I is, with rare exceptions,15,19,22 indicative of dextroversion. An analysis of the 119 cases discloses a number of features of this syndrome that have not been widely appreciated in the past.

It is often believed that a basic difference between mirror-image dextrocardia and dextroversion is that the former is associated with situs inversus while the latter is not. In fact, it is because of this notion that dextroversion has frequently been called "isolated dextrocardia" in the past. However, while the first part of this statement is relatively accurate (among more than a thousand cases of mirror-image dextrocardia in the literature, Korth and Schmidt could find only 12 that did not have situs inversus, and only 1 of these was studied at autopsy,1,23-25) the second part is not. A number of cases of well-authenticated dextroversion have been reported in which other thoracic and abdominal organs were abnormal either in structure or location, often taking a form that superficially resembled situs inversus. Among the 69 cases of dextroversion in which autopsy data are adequate, at least 16 had some degree of abdominal heterotaxy, including case 2 of the present report. However, there tends to be a basic difference between situs inversus and the heterotaxy seen with dextroversion. In situs inversus the viscera are inverted but, with few exceptions,24,26-28 they are otherwise normal and fully developed. On the other hand in dextroversion with abdominal heterotaxy, the involved abdominal organs are nearly always abnormal in form or structure. The abnormality takes a form that is perhaps best described as an embryologic arrest prior to the development of body symmetry, for the viscera tend to be abnormally symmetric in form and primitive in structure. For example, in the typical case of dextroversion with advanced abdominal heterotaxy, the liver is symmetric with both lobes equal in size, the gallbladder is often central, the duodenum may be either central or nonrotated, the stomach may be central and embedded in the substance of the liver or it may have primitive mesenteric attachments permitting it to lie anywhere in the abdomen, the mesentery of the bowel is often underdeveloped with many different bowel arrangements possible, the pancreas may be a central, unorganized tissue mass, both lungs will be tri-lobed, etc. The heart in dextroversion often best illustrates this arrest at a stage when the body has a simple, symmetric form, for it may show many of the features of the "single heart tube." For example, of the 16 cases of dextroversion with abdominal heterotaxy reported in the literature, 8 had cor biloculare and 5 of these had the combination of a single pulmonary vein, single atrium, single atrio-ventricular ring, single ventricle, and single outflow organ (truncus arteriosus).29-35

A curious anomaly often associated with dextroversion, and one which further empha-
sizes the difference between dextroversion and mirror-image dextrocardia with situs inversus, is congenital absence of the spleen, a subject that has recently been reviewed by Ivemark and Putsche and Manion. Ten of the 69 autopsied cases of dextroversion (including case 2 of the present report) had asplenia, while among hundreds of cases of situs inversus totalis reported in the literature, only 1 had asplenia and in this case the diagnosis was made during surgical exploration and not at autopsy. Nearly half of the 107 cases of asplenia reported in the literature had right-lying hearts, and although the data are incomplete in many of them, most if not all were instances of dextroversion. Furthermore, among all cases of asplenia 90 per cent had associated intracardiac abnormalities, usually cono-truncal abnormalities; this is the same incidence of cono-truncal deformity seen in dextroversion, while in mirror-image dextrocardia additional intracardiac abnormalities are exceedingly uncommon. Finally, the abdominal heterotaxy that usually accompanies asplenia more closely resembles that seen with dextroversion (i.e., symmetric heterotaxy) than it does situs inversus. Ivemark explained the association of asplenia, abdominal heterotaxy, and cono-truncal malformation by ascribing it to an arrest of development at the stage when the body first searches out its normal asymmetry, and he points out that the splenic bud appears and cono-truncal differentiation takes place at the same fetal date. Putsche and Manion suggested that asplenia and heterotaxy may be due to suppression or inhibition of structures that are asymmetric by their left-sidedness; however this would not account for the high incidence of cono-truncal abnormalities in these cases.

The degree of developmental arrest in the dextroversion syndrome varies from case to case and from organ to organ in individual cases. In the heart, for example, certain cardiac metameres may be arrested while adjacent ones develop normally. This is illustrated by case 2 of this report where, with most of the heart arrested at the "single heart tube" stage (bilateral superior and inferior venae cavae, single atrium, single atrioventricular ring, single ventricle, single common pulmonary vein), the great vessels were completely differentiated without even transposition. And there are cases at the other end of the spectrum, with only a single feature of the dextroversion syndrome present. For example, Korth and Schmidt found in the literature 6 cases of dextroversion in which the rightward rotation of the ventricular part of the heart was the only abnormality found at autopsy, with no septal defects, transposition, or abdominal heterotaxy. When the abnormality is limited in its extensiveness, as in these cases, it is clinically benign. Of the 119 cases of proved dextrocardia in the literature, 50 were adults and living at the time they were reported. Although other abnormalities were identified in many of these cases by clinical or x-ray examination, the abnormalities were not marked enough to interfere seriously with cardiac function.

One of the commonest associated intracardiac abnormalities in dextroversion is transposition of the great vessels. Of 59 cases in which the position of the great vessels was described either from autopsy or angiography, 5 had truncus arteriosus, and 44 had transposition of the great vessels. Thus 80 per cent had some form of outflow abnormality. The transposition is somewhat different from that seen when the heart is normally placed, for in dextroversion the aorta lies to the left of the pulmonary artery and only slightly anterior to it, while in transposition with a normally placed heart the aorta is usually directly anterior to the pulmonary artery. Thus, the roentgenogram of the heart in dextroversion shows a wide base if transposition is present, with the aorta often forming a prominent, long left margin of the cardiac silhouette as shown in figure 1. Since the great vessels lie side by side in this form of transposition, one might wonder if they are not simply "uncoiled" by the abnormal rightward position of the heart. However, turning the normally placed heart to the right would tighten the coil of the great vessels, not uncoil them. To uncoil the great vessels, the
ventricular part of the heart must rotate markedly leftward, a phenomenon that Shriner has reported in the pig embryo and possibly in 1 human fetus. The transposition of dextroversion is nearly always accompanied by a ventricular septal defect. Of 55 cases of dextroversion with adequate autopsy data, 44 had ventricular septal defects. There is only a single case of dextroversion in the literature with transposition but no ventricular septal defect.

This means that a high percentage of cases of dextroversion have one or more associated intracardiac anomalies, (90 per cent of autopsied cases). This is in striking contrast with mirror-image dextrocardia. Among more than a thousand cases of mirror-image dextrocardia with situs inversus in the literature only 5 cases have been found in which additional intracardiac anomalies were or may have been present, an incidence not greatly different from that of the population at large. This difference between the 2 syndromes emphasizes the fact that they must be due to quite different embryologic defects. It also leads to the useful clinical aphorism that if, in a patient with a right-lying heart, there is also cyanosis or an abnormal cardiac murmur, the diagnosis is probably dextroversion and not mirror-image dextrocardia regardless of the position of the abdominal organs. The variety of intracardiac and extracardiac abnormalities that may be associated with dextroversion of the heart warrant treating it as a "syndrome" of which the right-lying heart is only a part and perhaps not always present. For example, among Ivemark's 69 cases of asplenia, abdominal heterotaxy, and conotruncal deformity, 46 had no dextroversion. Although precise data are not available, clinical experience also indicates that dextroversion is uncommon among all cases of conotruncal deformity.

The most striking positional anomaly in dextroversion is the rightward location of the ventricular part of the heart. There are 2 reasons for believing that this is due to an arrest at the stage of cardiac development when the ventricular loop normally has a rightward position and is not due to either abnormal inversion or torsion of the heart. In the first place the ventricles are the only part of the heart to have the abnormal position. If it were due to inversion one would expect the atria or the great vessels would also occasionally be inverted. However, there are no cases of dextroversion so far reported in whom the atrial bodies were also abnormal in position; and the arch of the aorta is left-sided in the majority of cases of dextroversion, whether or not transposition is present. (Most students do not consider transposition of the great vessels to be due to inversion, if by inversion one means a translocation resulting from a disturbance of factors that determine body laterality, as in situs inversus; generally, transposition is considered due to a local disturbance in either the rate or magnitude of the spiralling of the truncal septum as it descends.) In the second place, there are in the literature 13 cases in which both atrial appendages lay to the left of the aorta but with normally placed atrial bodies, and at least 8 of these had some degree of dextroversion. Clearly, in these cases the anomalous location of the right atrial appendage must have been due to delay or failure of the bulb to migrate to the left, and in the cases with dextroversion this failure evidently included the entire ventricular loop.

The particular position of the ventricles in dextroversion requires comment. While it may be graphic to describe the ventricles as "turned to the right as in turning the page of a book," this is not completely accurate. As can be seen from the schema in figure 5, the anatomic drawing in figure 3, and the angiocardiogram in figure 2, the right ventricle lies superiorly to the left ventricle, and the interventricular septum is at right angles to the frontal plane of the body while normally it is parallel with the frontal plane. Thus the ventricular part of the heart is not only rotated to the right but is also rotated on its long axis. For a normally placed heart to take this position it may be swung rightward in either the frontal plane (like a pen-
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Dulum) or in the horizontal plane (like turning the page of a book). The difference is of some importance because it is also necessary to rotate the ventricular heart on its long axis to simulate dextroversion completely, and for the former the supplemental rotation of the ventricles is 90 degrees in a counterclockwise direction while for the latter it is 90 degrees in a clockwise direction as viewed from the apex. This is, of course, an artificial way to look at dextroversion. Rather one should ask what is the embryologic positional shift that fails to take place, is it a leftward swing in the frontal plane (pendular) or a leftward swing in the horizontal plane (page turning)? Perhaps this is answered by the angiocardio-gram of case 3 (fig. 4), for this is an example of mesoversion, with the heart less markedly rightward than in dextroversion. It can be seen that the ventricular septum lies in the sagittal plane of the body, with the right ventricle to the right and the left ventricle to the left. If this heart is representative of the mid-position between dextroversion and the normally placed heart, then the ventricular loop comes to lie in the left chest by swinging about 120 degrees as a pendulum in the frontal plane, and the long axis rotation that brings the right ventricle to its normal position anterior to the left ventricle is a clockwise rotation of 90 degrees. Evidently this latter type of rotation occurs late in the course of the leftward swing of the ventricular heart for, in case 3 at least, it has not yet taken place.

It was pointed out earlier that when the aorta and pulmonary artery are transposed in dextroversion they have a somewhat different location than when they are transposed in the normally placed heart. Doerr explained this difference by suggesting that in dextroversion there is an inversion of the bulbus cordis in addition to transposition. However, when the heart of dextroversion is studied topologically it is found not necessary to postulate the additional anomaly. In figure 5 are shown the positional relationships between the various parts of the normal heart and the dextroverted heart, with and without transposition. The positions have been schematized by plotting 3-dimensionally the surfaces that separate the right and left sides of the heart. The surface between the atria is defined by the atrial septum, that between the ventricles by the ventricular septum, and that between the aorta and the pulmonary artery by the plane tangential to both. It can be seen from the diagrams that the peculiarity of the transposition seen in dextroversion is due to the fact that the rightward position of the ventricles is simply communicated to the transposed vessels. For example, if the truncal surface in D is swung as a pendulum to the left it will have the same spatial position as in transposition in the normally placed heart, B, and there is no evidence of inversion of the bulbar region. In contrast with this, in C is shown the schema of the cardiac surfaces in case 2. When the truncal surface in this case is swung leftward as a pendulum, it will have the same spatial position as in the normal heart, A. In other words there is no transposition in this case, and the normally related great vessels are simply deviated to the right by the rightward position of the ventricles. Note that in C while the crista supraventricularis has a normal spatial location, as a result of the ventricular dextroversion it is at right angles to the ventricular septum. This resulted in a much severer degree of deformity from a functional point of view than if transposition had taken place. Perhaps, then, the high incidence of transposition in cases of dextroversion is simply due to the fact that normal descent of the truncal septum in a heart with dextroversion often results in a deformity that is incompatible with survival beyond earliest fetal stages and therefore rarely comes to the attention of either the clinician or the pathologist. In any case, it is likely that case 2 survived as long as she did only because of the presence of large ventricular and atrial septal defects and a single atrio-ventricular orifice, permitting oxygenated blood to reach the systemic circuit.

Summary and Conclusions

Three cases of dextroversion and an anal-
ysis of an additional 116 cases collected from the literature are presented. There are 3 major differences between dextroversion and mirror-image dextrocardia. 1. In dextroversion the atria have a normal position but the ventricular heart lies rightward, as if swung like a pendulum through an arc of 120 degrees in the frontal plane from its normal position; there is also a 90 degree counterclockwise rotation of the ventricular heart on its long axis, with the right ventricle lying superiorly to the left ventricle. In contrast with this, in mirror-image dextrocardia all cardiac structures are rightward mirror-images of the normal heart. 2. The vast majority of cases of dextroversion (90 per cent of autopsied cases) have additional intracardiac malformations, usually of the cono-truncal region, while in mirror-image dextrocardia additional intracardiac anomalies are probably no more frequent than in the population at large. 3. An abdominal heterotaxy is often present in dextroversion, which is different from the situs inversus that accompanies mirror-image dextrocardia: instead of inversion of abdominal organs there is a tendency for abdominal structures to be primitive and bilaterally symmetric, frequently with congenital absence of the spleen.

It is suggested that dextroversion is a part of a potentially multiorgan developmental defect that takes place at a very early fetal stage when the body first begins to search out its normal asymmetry, when cardiac septation and cono-truncal differentiation start, when the splenic bud first appears, and when the ventricular loop is normally predominantly rightward in location. Depending upon the intensity and distribution of the abnormality a wide variety of intracardiac and extracardiac anomalies and combination of anomalies may result. Dextroversion of the heart is perhaps only one part of this syndrome.

Transposition of the great vessels is present in over 80 per cent of cases of dextroversion. Indirect evidence is offered which suggests that the high incidence of transposition may partly be due to the fact that, in the presence of dextroversion, a normal descent of the truncal septum often produces a severer functional abnormality than does transposition and these cases do not usually survive earliest fetal stages.

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**SUMMARIO IN INTERLINGUA**

Es presentate 3 casos de dextroversion del corde e un analyse de 116 casos additional colligite ab le litteratura. Il ha 3 differentias principal inter dextroversion del corde e dextrocardia specular. (1) In dextroversion le atrios occupa un position normal, durante que le corde ventricular es displaciate verso le dextera, como si—in le maniera de un pendulo—illo habeva essite movite a transverso un arco de 120 grados in le plano frontal foras de su position normal. Il ha etiam un rotation sinistrose de 90 grados circa le axe longe del corde ventricular, e le ventriculo dextere occupa un sito superior al ventriculo sinistre. Per contrasto con isto, in dextrocardia specular, omne le structuras cardiac es dextrorse correspondentias specular del corde normal. (2) In le grande majoritate del casos de dextroversion del corde (90 pro cento secundo le necropsias reportate), il ha additionalmente malformaciones intracardiac, usualmente in le region cono-truncal, durante que anomalias intracardiac in subjectos con dextrocardia specular es probablemente non plus frequente que in le population general. (3) Un heterotaxia abdominal es frequentemente incontrate in casos de dextroversion del corde, que differe ab le sito inverse associate con dextrocardia specular. In loco de inversion del organos abdominal, il ha le tendentia que le structuras abdominal remane primitive e es bilateramente symmetric, frequentemente con absentia congenite del splen.
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Es presentate le these que dextroversión es posiblemente un parte de un potentialmente extense defecto disvelopplamental que ocurren mucho precocemente in le vita fetal, quando le organismo comencía su "tentativas de asimmetria," quando le septation cardiac e le differentiation cono-truncal se inicia, quando le button splenique se manifesta, e quando le ansa ventricular occupa normalmente un loco predominantemente dextrorse. In dependencia del intensitate e del distribution del defectos disvelopplamental, un grande varietate de anomalías intra- e extra-cardíace e de combinations de tal anomalías pote resultar. Dextroversión del corde es forsan solmente un parte de iste syndrome.

Transposition del vasos major es presente in plus que 80 pro cento del casos de dextroversión. Es presentate indicios indirecte que supporta le these que le alte incidentia de transposition del vasos mayores in parte causate per le facto que in le presentia de dextroversión un descendita normal del septo truncal produce frequentemente un anormalità tional que es plus sever que illo produceite per transposition, e in caso de un descendita normal del septo truncal le feto non supervive usualmente al prim stadios de su disvelopplamento.

REFERENCES
34. SHAPIRO, P.: Detorsion defects in congenital cardiac anomalies. Arch. Path. 9: 54, 1930.
35. IVENMARK, B. J.: Case #10, q.v.

For true Philosophers, who are perfectly in love with truth and wisdom, never find themselves so wise, or full of wisdom, or so abundantly satisfied in their own knowledge, but that they give place to truth whenssoever, or from whosoever it comes.—WILLIAM HARVEY. De Motu Cordis, 1628.
The Syndrome of Dextroversion of the Heart
ROBERT P. GRANT

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