COMPLETE AND INVERTED TRANSPOSITION OF THE GREAT VESSELS

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The embryology, pathology, clinical picture, and surgical treatment of transposition of the great vessels have been extensively reviewed. Less attention, however, has been given to the anatomy of the chambers of the heart in this condition. One name has been used to describe more than one anatomical type of transposition, or several names (mostly non-descriptive) have been used to describe one anatomical type. Corrected transposition (Rokitansky, 1875; Anderson, Lillehei, and Lester, 1957; and others), reversed torsion of the human heart (Lewis and Abbott, 1915), mirror image transposition of the ventricles, aorta, and pulmonary artery (Beattie, 1922), transposition of the ventricles and the arterial stems (Walmsley, 1931), mixed *lævocardia* with transposed ventricles with complete pseudotransposition (Lev, 1954), and inverted transposition (Spitzer, 1923): these are all different terms used to describe one type of corrected transposition of the great vessels. Dextro-rotation and 'some type of *lævocardia*', were two terms used by de La Cruz, Polansky, and Navarro-López (1962) to describe one type of corrected transposition of the great vessels with isolated dextrocardia and isolated *lævocardia* respectively. Whereas the term dextroversion has been used by Grant (1958) to describe one variety of isolated dextrocardia, it has been used by Lichtman (1931) to indicate displacement of the heart to the right by acquired or congenital causes, by Paltauf (1901) and Mönckeberg (1924) to signify an extrinsic congenital dextrocardia, and by Harris and Faber (1939) to describe their Case 16 of corrected transposition with *lævocardia*. Mixed *lævocardia* and mixed dextrocardia (Lev, 1954) are two terms that have been used when each atrium is not connected to its corresponding ventricle, in *lævocardia* and in dextrocardia respectively.

The object of this paper is to discuss the basic anatomical types of transposition of the great vessels and to advocate the use of their actual anatomical names. Inversion means a reversal in the frontal plane of the lateral relations between structures, and transposition means a reversal of relations in the antero-posterior plane. The chambers of the heart are best named according to the morphology, and morphology is best recognized by the architecture of the septum (Lev and Rowlatt, 1961). Thus the right atrium presents a limbus and a fossa ovalis on its septal surface; and the left atrium has on its septal surface the irregular configuration of the septum primum as it is adherent to the septum secundum. The right ventricle presents the septal and parietal bands forming the crista supraventricularis, and a trabeculated septum in the inlet. The left ventricle presents a relatively non-trabeculated septal surface in its basilar portion, and a mildly trabeculated apical portion, with the fibres streaming parallel to the outflow of blood.

In 1923 Spitzer recognized four grades of transposition of the great vessels. In his type 1, the pulmonary trunk arises from the right ventricle, and the aorta overrides the ventricular septum. In type 2, the aorta and the pulmonary trunk both arise from the right ventricle. In type 3 (crossed transposition) the aorta arises from the right ventricle anteriorly and to the right of the pulmonary trunk, which arises from the left ventricle, posteriorly and to the left. Type 4 is similar to type 3, but in addition the tricuspid valve opens into the left ventricle, i.e. a single ventricle. Spitzer also claimed that these four types might also have inverted forms. Thus in his type 1, the aorta

overrides the septum, whereas the pulmonary trunk arises from the left ventricle. In his type 2, the two great vessels originate from the left ventricle. In his type 3, the aorta arises anteriorly and to the left from the left ventricle, whereas the pulmonary trunk arises posteriorly and to the right from the right ventricle. His type 4 is similar to type 3, but in addition the left-sided atrio-ventricular (A-V) valve opens into the right ventricle, the left ventricle functioning as a pure aortic ventricle.

Since the object of this paper is not to discuss partial forms of transposition of the great vessels of which Spitzer's types 1 and 2 are examples, my discussion will be confined to his type 3, i.e. crossed or complete transposition and its inverted form. As the anatomy of the great vessels is the same in Spitzer's types 3 and 4, I have considered type 4 to be a variant of type 3.

Basically, there are two forms of transposition where the aorta and the pulmonary trunk arise from two separate ventricles. These are complete and inverted transposition of the great vessels. In complete transposition with *lævocardia* (Fig. 1), the aorta originates from the right-sided ventricle (whether morphologically right or left) and is situated anteriorly and to the right, while the pulmonary trunk originates from the left-sided ventricle (whether morphologically left or right) and is situated posteriorly and to the left. In inverted transposition with *lævocardia* (Fig. 2), the aorta originates from the left-sided ventricle (whether morphologically left or right) and is situated anteriorly and to the left, while the pulmonary trunk originates from the right-sided ventricle (whether morphologically right or left) and is situated posteriorly and to the right.

According to the anatomy of the atria and of the ventricles, therefore, each of these two syndromes of transposition with *lævocardia*, may be further subdivided into four types.

A. Complete transposition of the great vessels (Fig. 1).

- (1) With normal position of the atria and of the ventricles (Fig. 1a).
- (2) With inversion of the ventricles and normal position of the atria (Fig. 1b).
- (3) With inversion of the atria and normal position of the ventricles (Fig. 1c).
- (4) With inversion of both the atria and of the ventricles (Fig. 1d).

B. Inverted transposition of the great vessels (Fig. 2).

- (1) With normal position of the atria and of the ventricles (Fig. 2a).
- (2) With inversion of the ventricles and normal position of the atria (Fig. 2b).
- (3) With inversion of the atria and normal position of the ventricles (Fig. 2c).
- (4) With inversion of both the atria and of the ventricles (Fig. 2d).

Two types of the normal heart exist, each being a mirror image of the other. First, the normal heart situated on the left with its venous chambers on the patient's right and *situs solitus* of the viscera. Secondly, the normal heart situated on the right with its venous chambers on the patient's left and *situs inversus* of the viscera. When describing anomalies of the heart in terms of transposition or inversion, naturally we have to refer to the normal heart as the basis of reference. With abnormal hearts situated on the left our frame of reference was the normal *lævoposed* heart with its venous chambers on the patient's right. Logically when describing inversion and transposition in a right-sided heart, we should refer to the normal *dextroposed* heart with its venous chambers on the patient's left. Most authors (Keith and MacDonnell, 1921; Mandelstamm and Reinberg, 1928; Burchell and Pugh, 1952; Keith, Rowe, and Vlad, 1958; Grant, 1958; de La Cruz *et al.*, 1959; Portillo *et al.*, 1959; Espino-Vela *et al.*, 1959; and others) compared anomalies like inversion and transposition in a right-sided heart with the normal heart situated on the left. According to their definition a heart situated on the right with its venous chambers on the patient's right has normal relation of the cardiac chambers to each other. If we were to accept this definition, mirror image lesions occurring with *lævocardia* and with *dextrocardia* would have to be given two different embryological explanations and two different anatomical names. To avoid this confusing situation, I shall use the normal *dextroposed* heart as my frame of reference when describing inversion or transposition in a right-sided heart. For example if in *dextrocardia* the venous atrium is on the patient's right and the arterial on the left, they are by my definition inverted. In complete trans-

FIG. 1.—The four types of complete transposition of the great vessels. SVC, superior vena cava; IVC, inferior vena cava; RA, morphological right atrium; RV, morphological right ventricle; LV, morphological left ventricle; LA, morphological left atrium; Ao, aorta; PA, pulmonary artery.

position with dextrocardia, the aorta originates from the left-sided ventricle (whether morphologically right or left) and is situated anteriorly and to the left of the pulmonary trunk which originates from the right-sided ventricle (whether morphologically left or right). In inverted transposition with dextrocardia, the aorta originates from the right-sided ventricle (whether morphologically left or right) and is situated anteriorly and to the right of the pulmonary trunk which originates from the left-sided ventricle (whether morphologically right or left). According to the anatomy of the atria and of the ventricles, each of these two types of transposition with dextrocardia

FIG. 2.—The four types of inverted transposition of the great vessels. Abbreviations as in Fig. 1.

may be similarly further subdivided into four types. The problem of nomenclature with dextrocardia is illustrated by Fig. 3.

SUBJECTS

I studied 97 patients with transposition of the great vessels, 83 from Guy's Hospital and 14 from Southampton Chest Hospital. There were 34 female and 63 male subjects, and their ages ranged from 12 hours to 32 years. The diagnosis has been confirmed by necropsy in 53, cardiac catheterization in 1, angiocardiology in 19, catheterization and angiocardiology in 12, operation in 5, catheterization and operation in 2, angiocardiology and operation in 2, and all three in 3 patients. Of the whole group of 97 patients, 63 had complete transposition and 34 had inverted transposition. In the 63 with complete transposition 12 had a single ventricle, i.e. 20 per cent, while in the 34 with inverted transposition, 20 had a single ventricle, i.e.

FIG. 3.—Illustration of the problem of nomenclature with dextrocardia. Abbreviations as in Fig. 1.

60 per cent. Only the external anatomy of the aorta and the pulmonary trunk and the position of the atria have been considered in determining the type of transposition with single ventricle.

The following description of the syndromes assumes *laevocardia*, but will also apply to *dextrocardia* if the sides are reversed. It is reiterated that the normal heart has been used as a basis of reference, whether it be on the left or on the right. The nomenclature of the syndromes applies equally to transpositions with *laevocardia* or to their mirror images with *dextrocardia*. Since absolute identification of the ventricles depends on the topography of the ventricular septum, the 32 patients with single ventricle (12 with complete transposition and 20 with inverted transposition) have been excluded from the following classification.

COMPLETE TRANSPOSITION OF THE GREAT VESSELS, 51 PATIENTS

(1) *Complete Transposition with Normal Position of Atria and Ventricles* (Fig. 1a); 47 patients. This type of transposition, generally known as complete transposition, has been extensively reviewed. Its embryology has been discussed by Pye-Smith (1872), Rokitsansky (1875), Keith (1909), Robertson (1913), Spitzer (1923), Pernkopf and Wirtinger (1935), Lev and Saphir (1937, 1945), Bremer (1942), and de La Cruz and da Rocha (1956). Its pathology has been discussed by Kato (1930), Lev (1953), Lev, Alcalde and Baffes (1961), and Elliott *et al.* (1963). Its coronary circulation has been discussed by Keith *et al.* (1953), Bernreiter (1958), Rowlatt (1962), and Shafer (1963c). Its clinical picture has been discussed by Taussig (1938), Campbell and Suzman (1951), Rushmer *et al.* (1953), Cleland *et al.* (1957), and Noonan *et al.* (1960). Its haemodynamics have been discussed by Becker and Brill (1948), Campbell *et al.* (1949), Fowler and Ordway (1952), Lenkei, Swan, and DuShane (1959), Noonan *et al.* (1960), and Shafer (1964). Its angiography has been discussed by Castellanos, Pereiras, and Garcia (1938 and 1950), Keith and Munn (1950), Campbell and Hills (1950), and others. Its surgical treatment has been discussed by Blalock and Hanlon (1950), Lillehei and Varco (1953), Cross, Kay, and Jones (1954), Baffes (1956), Kay and Cross (1955), Senning (1959), Idriss *et al.* (1961), Ochsner *et al.* (1961), Shafer (1963a), and others.

(2) *Complete Transposition with Inversion of the Ventricles and Normal Position of the Atria* (Fig. 1b). As long ago as the earlier part of the nineteenth century, writers were discussing the problem of transposition of the great vessels with ventricular 'transposition'. Thus Farre (1814), Walshe (1842), and Stoltz (1851) described examples of complete transposition with 'transposition of the ventricles'. These cases had a right-sided bicuspid A-V valve and a left-sided tricuspid A-V

valve, which suggested to these authors that the ventricles were inverted. That such criteria for diagnosing ventricular inversion were not acceptable to all workers even at that time was shown by the statement of Peacock (1866) that "In most instances the arteries only are transposed but in some cases, the ventricles also, as indicated by their relative size and by the form of the auriculo-ventricular valves, are transposed", and by the statement of King (1844) that "Dr. Walshe thought the ventricles and their valves were transposed, but some of his statement induce me to distrust this supposition".

There is no example of this type of transposition in the present series, but Cases 9 and 10 of Lev and Rowlatt (1961) are good examples and form the basis of the following description. Right-sided venæ cavæ drain into the right-sided morphological right atrium which communicates through a bicuspid A-V valve with the right-sided but morphological left ventricle. The morphological left atrium on the left communicates through a tricuspid A-V valve with the morphological right ventricle, now situated on the left. The aorta arises anteriorly and to the right of the pulmonary trunk, from the right-sided ventricle. The pulmonary trunk arises posteriorly and to the left from the left-sided ventricle. The coronary arteries are usually transposed and inverted. The left coronary artery arises from an anterior sinus and courses to the right, dividing into an anterior descending and a right circumflex branch. The right coronary artery arises from a posterior sinus and courses to the left dividing into a posterior descending and a left marginal branch. Since the atria are not connected to their corresponding ventricles, e.g. a morphological left atrium connected to morphological right ventricle, complete heart block may occur in this type of transposition, e.g. Case 10 of Lev and Rowlatt (1961).

(3) *Complete Transposition with Inversion of the Atria and Normal Position of the Ventricles (Fig. 1c), 2 patients.* The two patients (D.P. 3 months and P.B. 8 years) in the present series with this type of transposition, both have isolated dextrocardia. The diagnosis has been confirmed by necropsy in D.P., and by cine-angiography in P.B. (Case 13 of Shaher, 1963b). The following description is based on these two patients, on cases with lævocardia reported by Griffith (1899), Yater (1929), Platzer (1955) Case 2, Lev and Rowlatt (1961) Case 11, Rosenbaum, Pellegrino, and Treciokas (1962) Case 1; and on Case 1 of Platzer (1955) with dextrocardia. The case described by Hickman (1869) with lævocardia is probably another example but with atresia of the pulmonary trunk.

As far as I can tell this type of complete transposition only occurs in isolated lævocardia or in isolated dextrocardia. The left-sided superior and inferior vena cava drain into a left-sided but morphological right atrium which communicates with the left-sided morphological left ventricle through a bicuspid A-V valve. This ventricle gives rise to the pulmonary trunk which is situated posteriorly and to the left. The pulmonary veins drain into a right-sided but morphological left atrium which communicates through a tricuspid A-V valve with the right-sided morphological right ventricle. This ventricle gives rise to the aorta which is situated anteriorly and to the right. In all the patients studied in this group, the aortic arch has always been on the side opposite to the cardiac apex. In patient D.P. both the right and the left coronary arteries arose by separate orifices from the anterior aortic sinus, while a very small coronary artery arose from the right posterior sinus: this passed to the right behind the pulmonary artery and in front of the venous atrium. In Case 2 of Platzer (1955) and in Case 11 of Lev and Rowlatt (1961), the right coronary artery arose from the right posterior sinus while the left arose from the left posterior sinus. In the Case of Yater (1929) the left coronary artery arose from the left anterior sinus, while the right arose from the posterior sinus. Since the atria are not connected with their corresponding ventricles, these cases are liable to develop complete heart block. Owing to the presence of atrial inversion, the aorta will be supplied with oxygenated blood, whereas the pulmonary trunk will be supplied with venous blood. Thus this type of transposition is physiologically corrected. Its pathogenesis has been discussed by Shaher and Johnson (1963) and its clinical features by Shaher (1963b).

(4) *Complete Transposition with Inversion of both the Atria and the Ventricles (Fig. 1d), 2 patients.* There are two patients with this type of transposition in the present series (B.T. and E.P.). Patient B.T., aged 14 years, has isolated lævocardia and is Case 9 of Shaher and Johnson (1963). Patient

E.P., aged 8 years, has isolated dextrocardia and is Case 12 of Shaher (1963b). The diagnosis has been confirmed by angiocardiology in B.T. and by catheterization and angiocardiology in E.P. The following description is based on these two patients, on cases with *lævocardia* reported by Campbell and Forgacs (1953) Case 1, and by Campbell (1960), and on cases with dextrocardia reported by Grunmach (1890), and by Kelsey, Gilmore, and Edwards (1953). The case reported by McCrae (1905) is probably another example with pulmonary atresia as the drawings suggest a posteriorly situated pulmonary trunk.

This type of complete transposition probably only occurs in isolated *lævocardia* and in isolated dextrocardia. Left-sided superior and inferior venæ cavæ drain into the morphological right atrium situated on the left. Through a left-sided tricuspid A-V valve, blood enters a left-sided but morphological right ventricle which gives rise to a pulmonary trunk situated posteriorly and to the left. Pulmonary venous blood returns to the morphological left atrium situated on the right. Through a bicuspid A-V valve, blood enters the right-sided but morphological left ventricle. The aorta arises from this ventricle anteriorly and to the right. In all the patients studied in this group, the aortic arch has been on the side opposite to the cardiac apex. A report of the anatomy of the coronary arteries was not available in any of the patients studied in this group. Since the atria and the ventricles are inverted together, complete heart block does not usually occur in this type of transposition. As a result of atrial inversion, oxygenated blood will enter the aorta and deoxygenated blood will enter the pulmonary trunk. This type of transposition is, therefore, physiologically corrected. Its pathogenesis has been discussed by Shaher and Johnson (1963) and its clinical features by Shaher (1963b).

INVERTED TRANSPOSITION OF THE GREAT VESSELS, 14 PATIENTS

(1) *Inverted Transposition with Normal Position of the Atria and Ventricles (Fig. 2a), 1 patient.* There is only one patient (P.H.) in this series with dextrocardia and situs inversus of the viscera who probably has this type of transposition. For the sake of comparison the following description assumes *lævocardia* although it is based on this one patient.

Right-sided superior and inferior venæ cavæ drain into a morphological right atrium situated on the right. This atrium communicates through a tricuspid A-V valve with the morphological right ventricle situated on the right, which in turn gives rise to the pulmonary trunk which is situated posteriorly and to the right. The pulmonary veins drain into a morphological left atrium on the left, which communicates through a bicuspid A-V valve with the morphological left ventricle situated on the left. The aorta arises from this ventricle anteriorly and to the left and thus is occupying the normal position of the pulmonary trunk at the left cardiac border. Since each atrium is connected with its corresponding ventricle, complete heart block would not be a common feature of this type of transposition. Since the atria are normally situated, oxygenated blood will enter the aorta and deoxygenated blood will enter the pulmonary trunk, and the transposition is physiologically corrected.

In 1956 Cardell gave reference to the following six cases which he thought were examples of this type of transposition: Grunmach (1890), Théremin (1895) Case 47, Lewis and Abbott (1915), Doerr (1938), Carns, Ritchie, and Musser (1941), and Brown (1950). These six cases have been discussed elsewhere (Shaher, 1963b) and it has been shown that the Case of Grunmach (1890) is an example of complete transposition with inversion of both atria and ventricles, as already discussed, but with isolated dextrocardia. Théremin (1895) described the left-sided ventricle of his Case 47 as having an outflow tract which gave rise to the anteriorly situated aorta. No detailed anatomy of the topography of the ventricles was given in Brown's case (1950) to allow any definite conclusions to be made. The remaining three cases of Doerr (1938), Lewis and Abbott (1915), and Carns *et al.* (1941) are all examples of inverted transposition of the great vessels with single ventricle. All six cases, however, shared a common feature in having a right-sided tricuspid A-V valve and a left-sided bicuspid A-V valve. It appears that Cardell accepted a right-sided tricuspid A-V valve as indicating a right-sided morphological right ventricle, and a left-sided bicuspid A-V valve as indicat-

ing a left-sided morphological left ventricle. Since we know that absolute identification of the ventricles depends on the topography of the ventricular septum (Lev, 1954), Cardell's conclusions are probably unjustified (Shaher 1963b). Since this last paper was written, the above-mentioned patient (P.H.), who is probably an example of inverted transposition of the great vessels with normally situated atria and ventricles, in dextrocardia with situs inversus, has been seen at Guy's Hospital.

FIG. 4.—Electrocardiogram of patient P.H. with dextrocardia and inverted transposition of the great vessels: right-sided chest leads without right and left arm leads being reversed.

TABLE I
DATA OBTAINED AT CARDIAC CATHETERIZATION OF PATIENT P.H.

Site	Saturation (%)	Pressure (mm. Hg)
Left superior vena cava ..	67	—
Inferior vena cava ..	61	—
Venous atrium	66	11, 9, 10, 8
Venous ventricle	—	90/0
Arterial atrium	88	12, 2, 11, 4
Arterial ventricle	87	90/5
Left brachial artery ..	87	90/65

Case History. A boy (P.H.) aged 11 years, whose mother was stated to have been ill during pregnancy, was late developing. On examination there was a moderate degree of cyanosis and clubbing of the fingers. The pulse had a good volume and was regular at a rate of 88 a minute. The jugular venous pressure was not raised. There was a systolic thrill to the right of the sternum. The first heart sound was normal and the second was single. There was a systolic ejection murmur. Chest radiograph showed the heart and the stomach air bubble to be situated on the right. The cardiogram (Fig. 4) showed nodal rhythm with an R wave in V6R and a deep S in V2. Cardiac catheterization was performed in 1962 (Table I) at which all four cardiac chambers of the heart were intubated. The pulmonary trunk was not entered. Atrial and ventricular septal defects were demonstrated by direct passage of the catheter from the venous atrium on the

FIG. 5.—Dye curves of patient P.H. obtained with injections of 2 per cent Coomassie Blue into venous atrium (RA), venous ventricle (RV), arterial atrium (LA), and arterial ventricle (LV).

left to the arterial atrium on the right, and from the venous ventricle on the left to the arterial ventricle on the right. Dye curves recorded with an ear-piece type photocell were obtained with injections of Coomassie Blue from each of the cardiac chambers (Fig. 5). The curves obtained from the left-sided venous atrium and ventricle showed an early hump, followed by a larger curve representing the dye that had circulated in the lungs before reaching the ear. The two curves obtained from the right-sided arterial atrium and ventricle showed an early appearance and early disappearance. These curves show that the pulmonary trunk is arising from the venous ventricle, and that the aorta is arising from the arterial ventricle. Angiocardiography was performed by Dr. J. Dow with a selective dye injection into the arterial ventricle: although the origin of the great vessels was not clearly demonstrated, the aorta was shown to be anterior and to the right of the pulmonary trunk, i.e. inverted transposition for dextrocardia. The two ventricles lay side by side, and the arterial ventricle on the right was shown to have a smooth outline, suggesting that it was a morphological left ventricle (Fig. 6 and 7).

(2) *Inverted Transposition with Inversion of the Ventricles and Normal Position of the Atria* (Fig. 2b), 11 patients. Under the title of corrected transposition of the great vessels, this type has been adequately described by Walmsley (1931), Harris and Farber (1939), Liebow and McFarland (1941), Anderson *et al.* (1957), Gasul, Graettinger, and Bucheleres (1959), de La Cruz *et al.* (1959, 1962), Malers *et al.* (1960), Beck *et al.* (1961), Schiebler *et al.* (1961), and Honey (1962, 1963).

FIG. 6.—Antero-posterior angiogram of patient P.H. showing aorta (Ao) occupying the normal position of the pulmonary artery (PA) in dextrocardia. The right-sided arterial ventricle (LV) has a smooth outline, suggesting a morphological left ventricle. The venous ventricle (RV) is on the left.

FIG. 7.—Lateral angiogram of patient P.H. showing anterior position of aorta (Ao) and posterior position of pulmonary artery (PA).

The morphological right atrium is situated on the right. It communicates through a bicuspid A-V valve with the right-sided but morphological left ventricle. This ventricle gives rise to the pulmonary trunk which is situated posteriorly and to the right. The morphological left atrium is situated on the left, communicates through a tricuspid left A-V valve with the left-sided but morphological right ventricle, which gives rise to the aorta situated anteriorly and to the left. Consequently the ascending aorta takes the position that is normally occupied by the pulmonary trunk at the left cardiac border. The coronary arteries are transposed and inverted: the left arises from an anterior sinus and courses to the right, dividing into an anterior descending and a right circumflex branch; and the right arises from a posterior sinus and courses to the left, dividing into a posterior descending and a left marginal branch. Since each atrium is not connected with its corresponding ventricle, heart block may occur in this type of transposition. Since the atria are normally situated, the aorta will be supplied with oxygenated blood, and the pulmonary trunk with venous blood. This type of transposition is therefore corrected physiologically.

(3) *Inverted Transposition with Inversion of the Atria and Normal Position of the Ventricles* (Fig. 2c). There is no example of this type of transposition in the present series, but Case T.N. 551105 (Fig. 707) of Kjellberg *et al.* (1959), with isolated dextrocardia, is probably an example. The following description, which assumes *lævocardia*, is based on this case. As far as I can tell, this type of transposition probably occurs only with isolated dextrocardia or isolated *lævocardia*. Left-sided *venæ cavæ* drain into the morphological right atrium situated on the left, which communicates with the left-sided morphological left ventricle. This gives rise to the anteriorly situated aorta. The pulmonary veins drain into the morphological left atrium situated on the right which communicates with the morphological right ventricle situated on the right. This gives rise to the

posteriorly situated pulmonary trunk. In the one case studied in this group, the aortic arch was on the side opposite to the cardiac apex. Owing to the presence of atrial inversion, oxygenated blood enters the pulmonary trunk, while deoxygenated blood enters the aorta, and the transposition is not corrected physiologically. Its pathogenesis has been discussed by Shaher and Johnson (1963).

(4) *Inverted Transposition with Inversion of both the Atria and Ventricles* (Fig. 2d), 2 patients. The two patients with this type of transposition in the present series both have isolated dextrocardia. In one Cn.L., 3 years, who is also Case 12 of Shaher and Johnson (1963), the diagnosis has been confirmed by cardiac catheterization, angiocardiography, and necropsy, and in the second patient Cy.L., 10 years, by catheterization and angiocardiography. These two patients, as well as Specimen 2 of Lev (1954), which is also a case of isolated dextrocardia, form the basis of the following description which assumes lœvocardia.

This type of transposition probably occurs only in isolated dextrocardia and isolated lœvocardia. Left-sided superior and inferior venæ cavæ drain into the morphological right atrium on the left. This atrium communicates through a tricuspid A-V valve with the left-sided but morphological right ventricle which gives rise to the anteriorly situated aorta, usually occupying the normal position of the pulmonary trunk. The pulmonary veins drain into the right-sided but morphological left atrium, which communicates through a bicuspid A-V valve with the right-sided but morphological left ventricle: this gives rise to the posteriorly situated pulmonary trunk. In patient Cn.L. as well as in Specimen 2 of Lev (1954) the right and left coronary arteries arose from the right and left posterior aortic sinuses respectively. In the three patients studied, the aortic arch has been on the side opposite to the cardiac apex. Since the atria are connected with their corresponding ventricles, complete heart block does not usually occur. Owing to the presence of atrial inversion, oxygenated blood enters the pulmonary trunk, and deoxygenated blood enters the aorta, and the transposition is not corrected physiologically. Its pathogenesis has been discussed by Shaher and Johnson (1963).

Case History. A 10-year-old girl (Cy.L.) had been cyanosed and breathless since birth. On clinical examination, there was moderate central cyanosis and clubbing of the fingers. The brachial pulse was regular and the jugular venous pressure was normal. On palpation the apex was found to be on the right side of the chest, and a systolic thrill was felt over the præcordium. On auscultation there was a loud systolic murmur and the second sound was single. The cardiogram with normal lead connexions (Fig. 8) showed upright P

FIG. 8.—Electrocardiogram of Cy.L. with dextrocardia and inverted transposition of the great vessels: right-sided chest leads without right and left arm leads being reversed.

FIG. 9.—Antero-posterior angiogram of patient Cy.L. showing the aorta (Ao) occupying the normal position of the pulmonary artery (PA) in dextrocardia. The right-sided ventricle (RV) has a trabeculated outline and an outflow tract suggesting a morphological right ventricle.

FIG. 10.—Antero-posterior angiogram of patient Cy.L. showing the pulmonary artery (PA) centrally situated and arising from a left-sided ventricle (LV) with a smooth outline suggesting a morphological left ventricle. SVC, superior vena cava.

FIG. 11.—Lateral angiogram of patient Cy.L. showing anterior position of the aorta (Ao) and posterior position of the pulmonary artery (PA).

waves in lead I indicating a right-sided venous atrium. The right-sided chest leads showed deep S waves from V2–V6R. Chest radiograph showed the heart to be situated mainly on the right and the stomach air bubble on the left. Cardiac catheterization and venous angiography in 1957 demonstrated the presence of a right-sided superior vena cava draining into a right-sided venous atrium. Two pulmonary veins were intubated at the left cardiac border through an atrial septal defect. There was almost complete mixing of the blood-streams at atrial level. Angiocardiology was repeated by Dr. J. Dow in 1961. A right-sided inferior vena cava was demonstrated, and the dye was selectively injected into each of the two ventricles separately. A right-sided ventricle with heavy trabeculation and an outflow tract gave rise to the aorta (Fig. 9). A left-sided ventricle with a smooth outline gave rise to the pulmonary trunk (Fig. 10). The great vessels were transposed (Fig. 11), the aorta lying anteriorly and to the right of the pulmonary trunk, i.e. inverted transposition for dextrocardia.

DISCUSSION

Rokitasky (1875), Harris and Farber (1939), Liebow and McFarland (1941), Becu *et al.* (1955), Anderson *et al.* (1957), Schaefer and Rudolph (1957), Fink *et al.* (1958), Keith *et al.* (1958), Helmholtz, Daugherty, and Edwards (1956), Kernen (1958), Badawi *et al.* (1961), Kahn *et al.* (1960), de La Cruz *et al.* (1959, 1962), Espino-Vela *et al.* (1959), Honey, Rushton, and Taylor (1960), Schiebler *et al.* (1961), Beck *et al.* (1961), Goodman and Kuzman (1961), El Sayed *et al.* (1962), and others, have confined corrected transposition of the great vessels to one anatomical type, namely that of inverted transposition with ventricular inversion. Spitzer (1923) admitted that he had never seen an example of corrected transposition of the great vessels, and he thought that this term should not be used and the term inverted transposition substituted.

It had been pointed out by Rokitansky (1875), Lev (1953), Schiebler *et al.* (1961), and Honey

(1962) that "correction" is achieved in inverted transposition with ventricular inversion by means of the ventricular inversion. Only the atria and venæ cavæ can 'correct' each of the syndromes of complete and inverted transposition: it would not make any difference to the hæmodynamics of the circulation, if the ventricle from which the aorta originates in complete or inverted transposition is morphologically left or right. Only the position of the atria will determine whether the aorta will be perfused with venous or arterial blood. In inverted transposition, with or without ventricular inversion, arterial blood will enter the aorta so long as the left atrium is situated on the left, i.e. normal position of the atria. Similarly in complete transposition, with or without ventricular inversion, arterial blood will enter the aorta only if the arterial atrium is situated on the right, i.e. atrial inversion.

It seems that any syndrome of transposition, whether complete or inverted, that is associated with atrial inversion either alone or together with ventricular inversion, will occur only in isolated lævocardia or isolated dextrocardia. Four syndromes of transposition are in this category.

The normal atrio-ventricular relation is disrupted if the atria alone are inverted, or if the ventricles alone are inverted. Each of the two syndromes of complete and inverted transposition has two anatomical situations where the atria are not connected to their corresponding ventricles. In three of these situations complete heart block tends to occur. In the fourth type, inverted transposition with atrial inversion, of which there is only one example, the cardiogram was not published, but it is reasonable to assume that complete heart block would also tend to occur. It seems, therefore, that whenever the atria are not connected to their corresponding ventricles heart block may develop. Why heart block should occur under such circumstances is far from clear. In Yater's case (1929) of complete transposition with atrial inversion, there was actual interruption of the bundle of His. In Walmsley's case (1931) of inverted transposition with ventricular inversion, dense fibrosis surrounded and infiltrated the bundle. The occurrence of complete heart block in transposition of the great vessels suggests, therefore, that the atria are not connected to their corresponding ventricles.

It has been shown by Lev and Rowlatt (1961) that the A-V valves usually follow their respective ventricles. Thus in each of the two syndromes of transposition two anatomical situations will exist where the A-V valves are usually inverted together with ventricular inversion. Fig. 1b, 1d, 2b, and 2d illustrate the anatomical situations where the morphological right ventricle and probably also the tricuspid A-V valve are situated on the left.

The anatomy of the coronary arteries in transposition of the great vessels has been discussed by Spitzer (1923), Keith *et al.* (1953), Rowlatt (1962), and Shaher (1963c). In the majority of cases of transposition, the left coronary artery arises from an anterior sinus, and the right coronary artery from a posterior sinus. It has been shown by Harris and Farber (1939), Anderson *et al.* (1957), and Lev and Rowlatt (1961) that inversion of the coronary arterial pattern usually occurs with ventricular inversion. The course of the left coronary artery to the left or to the right usually depends on whether the morphological left ventricle is situated on the left or on the right. Similarly the course of the right coronary artery to the right or to the left usually depends on whether the morphological right ventricle is situated on the right or on the left.

In the group of 34 patients with inverted transposition 20 (60%) had a single ventricle, while in the group of 63 patients with complete transposition only 12 (20%) had a single ventricle. These figures suggest that a diagnosis of single ventricle should be considered as the first possibility when inverted transposition is present.

SUMMARY

Patients with transposition of the great vessels are divided into two groups: Group 1 with complete transposition and Group 2 with inverted transposition. Each of the two groups is further divided, according to the relative positions of the atria and of the ventricles, into four types; (1) with normal position of the atria and of the ventricles, (2) with inversion of the ventricles and normal position of the atria, (3) with inversion of the atria and normal position of the ventricles, and

(4) with inversion of both the atria and of the ventricles. It is suggested that each type of transposition should be given its actual anatomical name.

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