

Anatomically Corrected Transposition of the Great Arteries*

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It has long been doubted whether or not it is possible for a transposed aorta to arise from a morphologically *left* ventricle, and for a transposed pulmonary artery to originate from a morphologically *right* ventricle, this having been designated *anatomically corrected transposition* by Harris and Farber in 1939. Such cases have been regarded as errors in observation (Lochte, 1898), as inexplicable variations of nature (Geipel, 1903), as embryologically impossible and hence non-existent (Van Mierop and Wiglesworth, 1963), and as very probably non-existent (Van Praagh *et al.*, 1964b).

Hence, the principal purposes of this paper are (1) to provide convincing evidence that anatomically corrected transpositions do indeed exist, and (2) to consider some of the developmental and diagnostic implications of this fact.

CASE REPORTS

Case 1. This girl, 2 years and 20 days of age, died during a surgical attempt to relieve her pulmonary out-flow tract stenosis. The salient post-mortem findings were: dextrocardia (Fig. 1A); situs solitus of the viscera and atria, with a normal spleen (Fig. 2); left juxtaposition of the atrial appendages (both to the left of the great arteries) (Fig. 1A); tricuspid atresia (Fig. 1E); large secundum type of atrial septal defect (Fig. 1B); ventricular *d*-loop (morphologically right ventricle to the right of the morphologically left ventricle) (Fig. 1 and 2); small ventricular septal defect (Fig. 1C, D); crista supraventricularis, largely above the morphologically left ventricle (Fig. 1C) but also above the morphologically right ventricle (Fig. 1D); a combined conus (subaortic

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and subpulmonary) (Fig. 2) preventing fibrous continuity between the mitral valve and either semilunar valve (Fig. 1C, D); infundibular and valvar pulmonary stenosis with a thickened bicuspid pulmonary valve (Fig. 1D, E); *l*-transposition of the great arteries (transposed aortic valve to the left of the transposed pulmonary valve) (Fig. 1E and 2); transposed aorta arising entirely above the morphologically left ventricle (Fig. 1C, E); transposed pulmonary artery originating completely above the morphologically right ventricle (Fig. 1D, E); right aortic arch (Fig. 1A); and probe-patent ductus arteriosus (Fig. 1D).

Examination, using normal controls and the measurements of Lev, Rowlatt, and Rimoldi (1961), revealed cardiomegaly, combined atrial hypertrophy and enlargement, a thick-walled but small chambered right ventricle, left ventricular hypertrophy and enlargement, tricuspid atresia, mitral annulus larger than normal, pulmonary annulus smaller than normal, and aortic annulus within normal limits.

Cardiac geometry (Van Praagh, Ongley, and Swan, 1964a) showed the following horizontal plane projections relative to the Z axis (the antero-posterior line) in this case (Fig. 1E), compared with the normal (Fig. 2).

Atrial septal angle $\approx 60^\circ$ left (normal $\approx 30^\circ$ left); ventricular septal angle $\approx 60^\circ$ right (normal $\approx 35-40^\circ$ left); atrio-ventricular septal angle (difference between atrial and ventricular septal angles) $\approx 120^\circ$ (normal $\approx 5-10^\circ$); conal septal angle $\approx 65^\circ$ right (normal \approx same); cono-ventricular septal angle (difference between conal and ventricular septal angles) $\approx 5^\circ$ (normal $\approx 100^\circ$); and rotation at the semilunar valves $\approx -20^\circ$ (normal $\approx +150^\circ$).

The abnormally leftward atrial septal angle appears related to the left juxtaposition of the atrial appendages. The exceedingly abnormal atrio-ventricular septal angle accurately expresses the marked malalignment of the ventricular loop and the ventricular septum relative to the atria and the septum of the atrio-ventricular canal. This malalignment resulted in the expected site of the atretic tricuspid orifice being directly over the

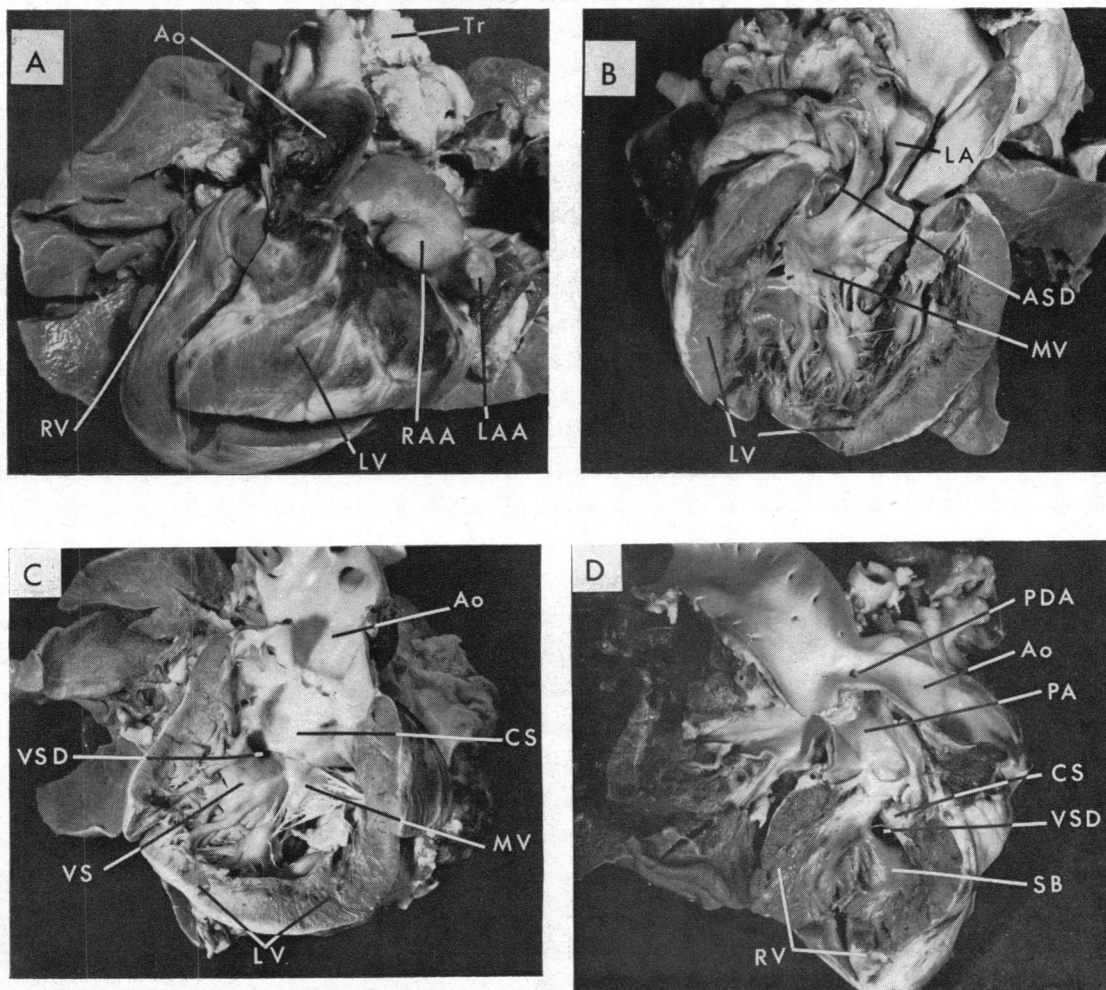
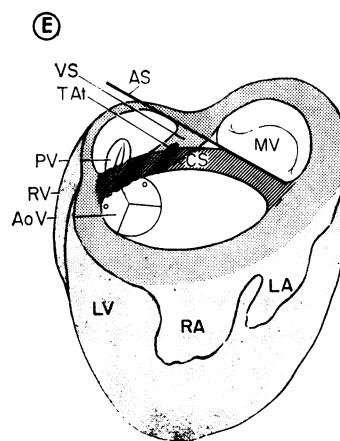


FIG. 1.—Case 1. Dextrocardia with situs solitus of the viscera and atria, a *d*-loop and *l*-transposition. This results in anatomically corrected transposition, and in physiologically corrected transposition without ventricular inversion. (A) Exterior antero-posterior view. (B) Opened left atrium and left ventricular inflow tract. (C) Left ventricular outflow tract to *l*-transposed aorta. (D) Right ventricle and transposed pulmonary artery. (E) Diagram of heart, viewed from above (as in Fig. 2). Tr = trachea; Ao = aorta; RAA = right atrial appendage; LAA = left atrial appendage; RV = morphologically right ventricle; LV = morphologically left ventricle; LA = left atrium; ASD = atrial septal defect; MV = mitral valve; CS = conal septum (crista supraventricularis); VSD = ventricular septal defect; VS = ventricular septum; PDA = patent ductus arteriosus; PA = pulmonary artery; SB = septal band; AS = atrial septum; TAt = tricuspid atresia; PV = pulmonary valve; AoV = aortic valve; RA = right atrium; and LA = left atrium.



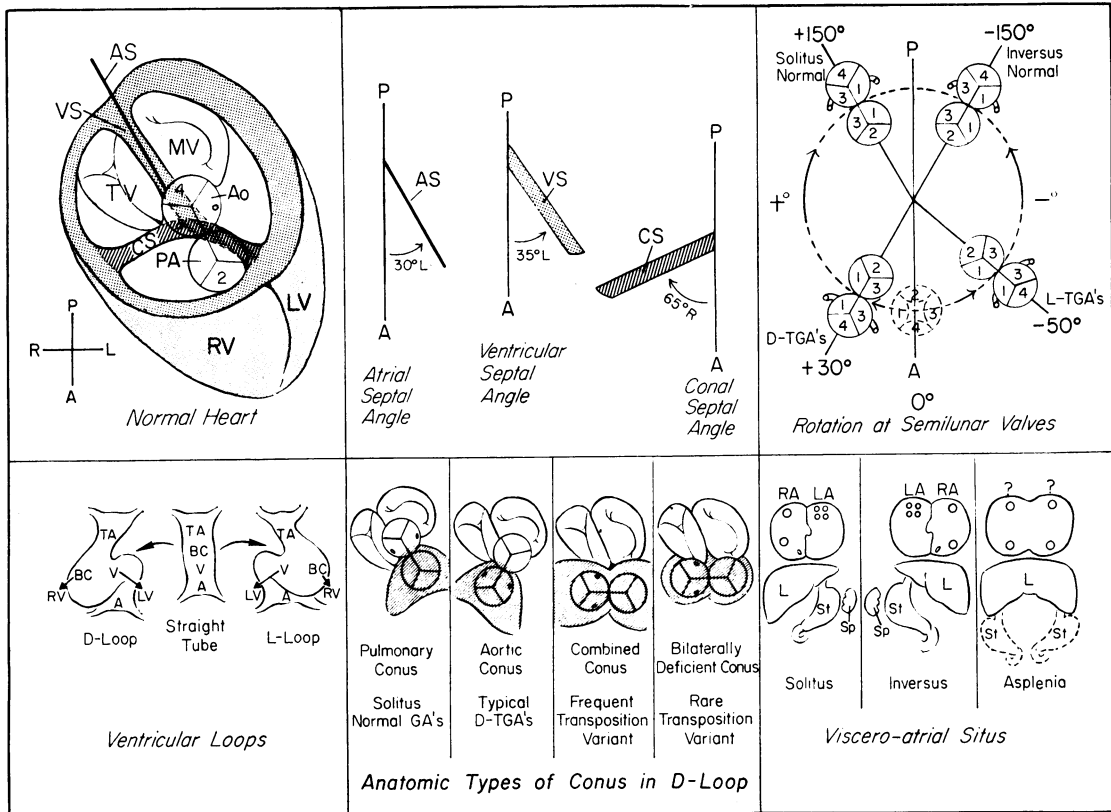


FIG. 2.—Basic concepts. See text. A=anterior; P=posterior; R=right; L=left; TV=tricuspid valve; TA=truncus arteriosus; BC=bulbus cordis; V=ventricle (of bulbo-ventricular loop); A=atrium (primitive, undivided) ?=morphology of atrium uncertain (with asplenia); L=liver; St=stomach; and Sp=spleen. Other abbreviations as in Fig. 1. (This diagram is published through the courtesy of the *Amer. J. Cardiol.*)

posterior end of the muscular ventricular septum, which appears highly relevant to the finding of tricuspid atresia. Moreover, the abnormally rightward ventricular septal angle correlates with the right-sided ventricular apex, resulting in dextrocardia, and with the unusual finding of a morphologically left ventricle lying anteriorly to a morphologically right ventricle (Fig. 1E). The exceedingly small cono-ventricular septal angle indicates that these two septa are nearly parallel. In view of this septal parallelism, and because the transposed aorta and the morphologically left ventricle are both anterior, while the transposed pulmonary artery and the morphologically right ventricle are both posterior, the great arterial-ventricular relationships of anatomically corrected transposition result.

Comment. It is noteworthy that this case of anatomically corrected transposition may also be regarded as corrected physiologically—in view of the veno-arterial relationships (Schiebler *et al.*, 1961). In fact, however, this potential physiological correction was vitiated by the presence of tricuspid atresia. With this reservation, the present case may

be viewed as physiologically corrected transposition without ventricular inversion. This viewpoint may be expressed somewhat more accurately as isolated inverted transposition (the viscera, atria, and ventricles not being inverted).

Relative to dextrocardia, it is of interest that this case documents the real dextroversion with transposition of the great vessels of Schmidt and Korth (1954), and the classical dextroversion of Grant (1958), the existence of which was questioned by Van Praagh *et al.* (1964b).

Case 2. This baby girl died at 4 weeks and 3 days of age during an attempt to perform a right subclavian-pulmonary anastomosis. Necropsy revealed: a left-sided heart (Fig. 3A); situs solitus of the viscera and atria; left juxtaposition of the atrial appendages (Fig. 3A); large secundum atrial septal defect (Fig. 3B); ventricular *d*-loop (Fig. 2 and 3); high large ventricular septal defect (Fig. 3B, C); combined conus (Fig. 2, 3C, D); *L*-transposition of the great arteries (Fig. 2, 3A, D); pulmonary outflow tract atresia (Fig. 3B, D); transposed

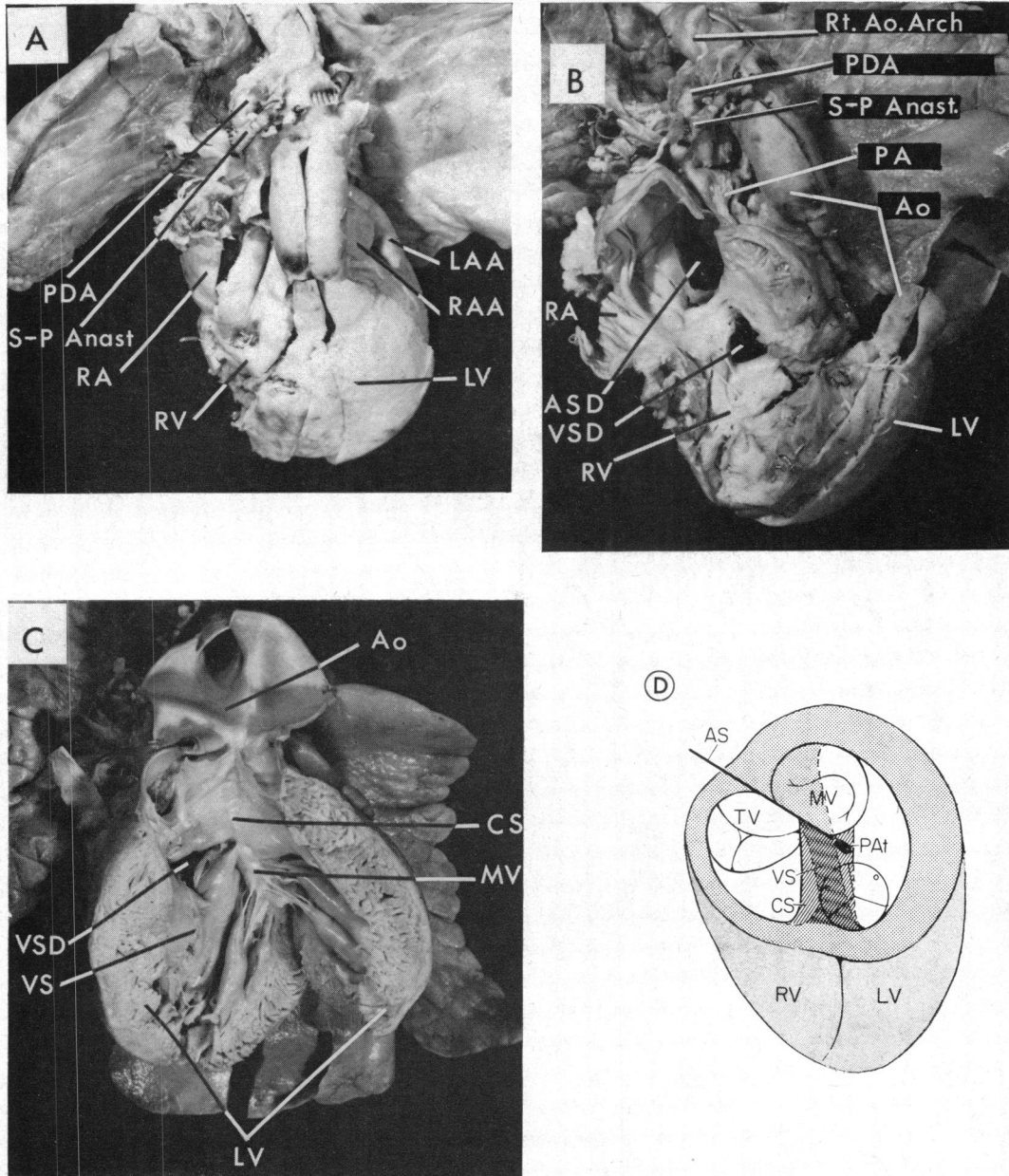


FIG. 3.—Case 2. Left-sided heart with situs solitus of the viscera and atria, *d*-loop and *l*-transposition. This results in anatomically corrected transposition, and in physiologically corrected transposition without ventricular inversion. (A) Exterior antero-posterior view. (B) Opened right atrium and right ventricle. (C) Left ventricle and *l*-transposed aorta. (D) Heart diagram. S-P Anast.=subclavian-pulmonary anastomosis; Rt. Ao. Arch=right aortic arch; and Pat=pulmonary atresia. Aortic valve is indicated by coronary ostia. Other abbreviations as before.

aorta arising above the morphologically left ventricle (Fig. 3C); right aortic arch (Fig. 3A, B); right patent ductus arteriosus (Fig. 3A, B); and right subclavian-pulmonary anastomosis (Fig. 3A, B).

Examination indicated cardiomegaly, combined atrial hypertrophy and enlargement, right ventricular hypoplasia, left ventricular hypertrophy and enlargement, tricuspid annulus smaller than normal, mitral annulus

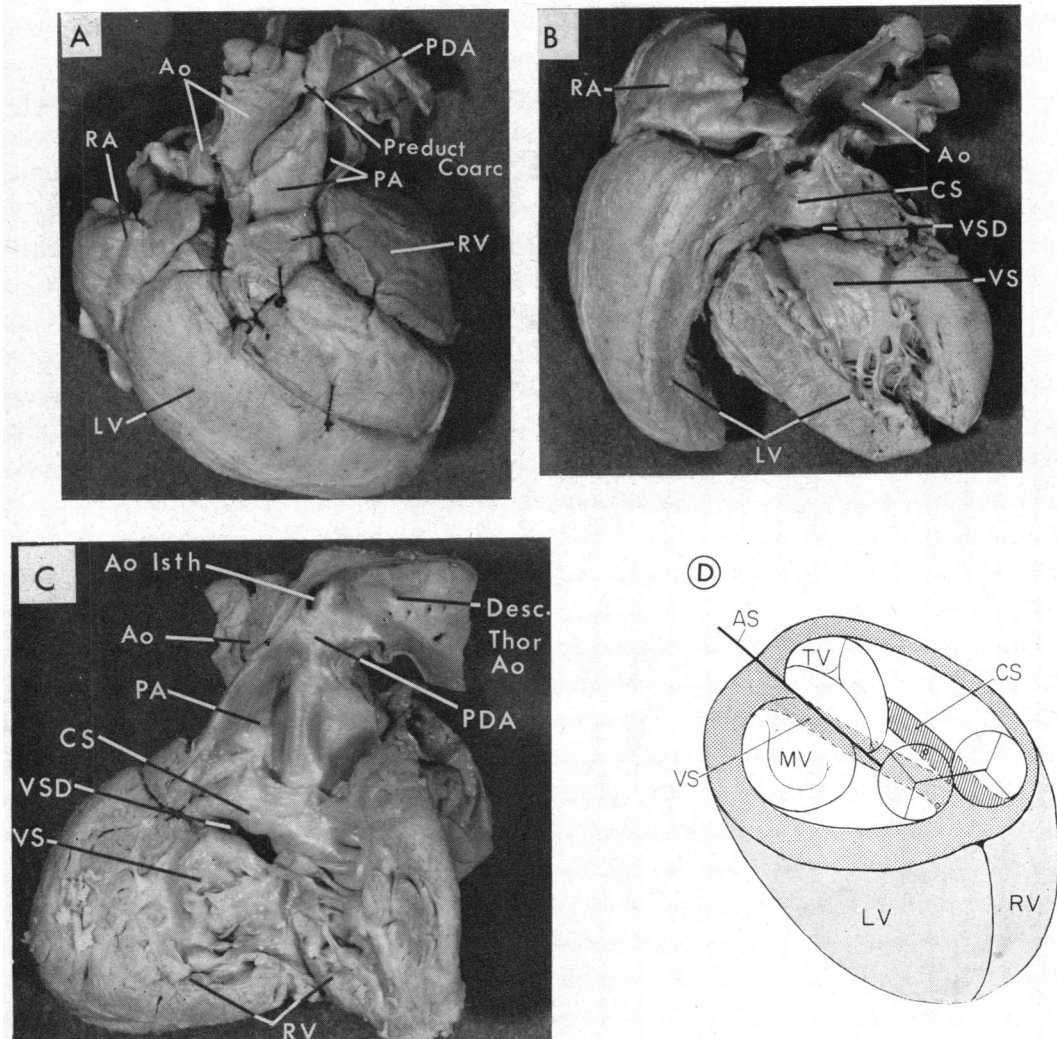


FIG. 4.—Case 3. Left-sided heart with situs solitus of the viscera and atria, *l*-loop and *d*-transposition. This results in anatomically corrected transposition, and in complete transposition with ventricular inversion. (A) Exterior antero-posterior view. (B) Morphologically left ventricle (right-sided) leading to *d*-transposed aorta. (C) Morphologically right ventricle (left-sided) leading to transposed pulmonary artery. (D) Heart diagram. Product. Coarc=productile coarctation; Ao Isth.=aortic isthmus; Desc. Thor Ao=descending thoracic aorta. Aortic valve is indicated by coronary ostia. Other abbreviations as before.

normal, pulmonary atresia, and the aortic annulus larger than normal.

Geometry showed (Fig. 3D) an atrial septal angle abnormally to the left (60° left), a ventricular septal angle abnormally to the right (5° left), an abnormally large atrio-ventricular septal angle (55°), a conal septal angle too far to the right (0°), an abnormally small cono-ventricular septal angle (5°), and a very abnormal rotation at the semilunar valves (-10°).

Comment. Case 2 is very similar to Case 1, except for the position of the heart within the thorax

(left-sided instead of right-sided), tricuspid hypoplasia (instead of atresia), and pulmonary atresia (instead of stenosis).

Case 3. This baby boy died at 4 weeks and 5 days of age from congestive heart failure. Necropsy revealed: left-sided heart (Fig. 4A); situs solitus of the viscera and atria; ventricular *l*-loop (morphologically right ventricle left-sided) (Fig. 2, 4); slit-like high ventricular septal defect between the conal septum (crista supraventricularis) above and the crest of the muscular ventricular septum below (Fig. 4B, C); combined conus, precluding

atrio-ventricular semilunar fibrous continuity (Fig. 2, 4B, C); *d*-transposition of the great arteries (transposed aortic valve to the right of the transposed pulmonary valve) (Fig. 2, 4A, D); transposed aorta arising above the morphologically left ventricle (right-sided) (Fig. 4A, B, D); transposed pulmonary artery originating above the morphologically right ventricle (left-sided) (Fig. 4A, C, D); mild subaortic infundibular stenosis (Fig. 4B), with mild hypoplasia of the ascending aorta, left aortic arch, and preductile coarctation (Fig. 4A, C); dilated main pulmonary artery, with absence of the left pulmonary artery; and a widely patent ductus arteriosus (Fig. 4A, C).

Examination revealed cardiomegaly, combined atrial hypertrophy and enlargement, mild left ventricular hypertrophy and enlargement (right-sided), thick-walled but small-chambered right ventricle (left-sided), and all valve circumferences within normal limits.

Geometry revealed (Fig. 4D) an approximately normal atrial septal angle (50° left), a ventricular septal angle $\approx 60^\circ$ left (approximately normal for non-inverted ventricles (Fig. 2) but very abnormal for inverted ventricles, which should have a ventricular septal angle $\approx 35^\circ$ right, *i.e.* a mirror-image of the normal *d*-loop ventricular septal angle), a normal atrio-ventricular septal angle (10°), a very abnormal conal septal angle (50° left), an abnormally small cono-ventricular septal angle (10°), and abnormal rotation at the semilunar valves ($+80^\circ$).

Comment. Thus, Case 3 displays complete transposition with ventricular inversion.

DISCUSSION

These three cases document the existence of anatomically corrected transposition: they indicate that it is possible for a transposed aorta to arise above the morphologically *left* ventricle, and for a transposed pulmonary artery to originate above the morphologically *right* ventricle; hence, the anatomical "correction" of the transposition (Harris and Farber, 1939).

Transposition of the great arteries is considered to be present, despite the origin of the great arteries above the morphologically appropriate ventricles, and despite the absence of mitral-pulmonary fibrous continuity (Grant, 1962), for the following reasons, which are based on two previous studies (Van Praagh and Van Praagh, 1966; Van Praagh, Vlad, and Keith, in the press).

(1) An abnormal type of conus is present, a combined (subaortic and subpulmonary) conus (Fig. 2). Non-transposed great arteries almost always have a pulmonary conus, without an aortic conus (Fig. 2).

(2) The relation between the aorta and the atrio-ventricular canal is abnormal. Mitral-aortic fibrous continuity is not present, apparently due to the interposition of abnormal subaortic conal myocardium (Fig. 1C, 3C, 4B). In our experience,

the presence of abnormal subaortic conal myocardium is a constant finding in transposition of the great arteries, but exceedingly rare with non-transposed great arteries which, therefore, almost always display mitral-aortic fibrous continuity (Fig. 2).

(3) The relation between the great arteries at the semilunar valves is abnormal, the aortic valve being anterior to the pulmonary valve in all (Fig. 1E, 3D, 4D).

(4) Although the great arteries arise above the morphologically appropriate ventricles, the great arteries are not *normally* related to these ventricles; e.g. in Cases 1 and 2 (Fig. 1, 3), the aorta arises far too anteriorly and superiorly above the left ventricle, while the pulmonary artery is located much too posteriorly and inferiorly above the right ventricle.

Hence, all three cases have a transposition type of conotruncal malformation, with abnormal relationships.

Types of Anatomically Corrected Transposition.

There are two types: with a ventricular *d*-loop (morphologically right ventricle right-sided) (Type I); and with a ventricular *l*-loop (morphologically right ventricle left-sided) (Type II).

Type I is exemplified by Cases 1 and 2 (Fig. 1 and 3), which have situs solitus of the viscera and atria with a *d*-loop and *l*-transposition, resulting in physiologically corrected transposition without ventricular inversion. Type II is illustrated by Case 3 (Fig. 4), which has situs solitus of the viscera and atria with an *l*-loop and *d*-transposition, resulting in complete transposition with ventricular inversion.

Anatomically corrected transposition has not as yet been adequately documented with situs inversus of the viscera and atria, nor with the asplenia syndrome, to our knowledge.

Developmental Considerations. The transposed great arteries and the ventricles appear to have twisted in opposite directions: *l*-transposition with a *d*-loop (Cases 1 and 2); and *d*-transposition with an *l*-loop (Case 3). This results in severe cono-ventricular malalignment, with a high ventricular septal defect at the cono-ventricular junction.

A combined (subaortic and subpulmonary) conus appears essential to the development of anatomically corrected transposition. Abnormal subaortic conal myocardium seems necessary to the development of any form of transposition (Fig. 2), thereby separating the aortic and mitral valves, and hence, disconnecting the aorta from the left ventricle. However, a subpulmonary conus must also develop, thereby separating the pulmonary and mitral valves. This makes it possible for the transposed pulmonary

artery to arise above the right ventricle, instead of above the left ventricle, as occurs in typical transpositions. None the less, the great majority of combined cono are not associated with the extreme cono-ventricular malalignment known as anatomically corrected transposition.

The infundibulum (conus) and the crista supraventricularis (parietal band) are not intrinsic, inseparable parts of the morphologically right ventricle, since in Cases 1 and 2 these structures are located predominantly above the morphologically left ventricle (Fig. 1C, 3C). The infundibulum and crista supraventricularis comprise the conus and are a part of the conotruncal segment, not an intrinsic part of either ventricle. Conversely, the septal band always appears to be an intrinsic inseparable part of the morphologically right ventricle (e.g. Fig. 1D), not part of the conus.

Diagnostic Considerations. Anatomically corrected transpositions constitute one type of exception to an angiocardigraphic method of localizing the morphological right and left ventricles by means of the type of relation between the great arteries, i.e. the loop rule (Van Praagh *et al.*, 1964b). Nevertheless, these correlations did prove accurate in 95 per cent of 149 cases of transposition of many types (Van Praagh, Vlad, and Keith, in the press) and in 92 per cent of 60 cases of single ventricle (Van Praagh *et al.*, 1964a). In view of the severe cono-ventricular malalignment which is the essence of anatomically corrected transposition, a segment-by-segment approach to the angiocardigram appears necessary, assisted by a segmentally deductive approach to the electrocardiogram (Portillo *et al.*, 1959; Sodi-Pallares *et al.*, 1959).

SUMMARY

Three cases of anatomically corrected transposition discovered at necropsy are presented, documenting a rare anomaly which has widely been regarded as embryologically impossible and, hence, non-existent.

There are two types: (1) with a ventricular *d*-loop (morphologically right ventricle right-sided), and (2) with a ventricular *l*-loop (morphologically right ventricle left-sided). In both types, the transposed aorta arises above the morphologically left ventricle, and the transposed pulmonary artery originates above the morphologically right ventricle; hence, anatomical "correction" of the transposition.

A transposition type of conotruncal malformation is present in all three cases, with a combined (sub-aortic and subpulmonary) conus, and with abnormal

relations between the aorta and the atrio-ventricular canal, between the great arteries and the ventricles, and between the great arteries themselves.

Developmentally, the great arteries and the ventricles appear to have twisted in opposite directions, resulting in severe cono-ventricular malalignment, with a high ventricular septal defect at the cono-ventricular junction. A combined conus appears essential to the development of this malalignment.

These cases illustrate that the infundibulum and crista supraventricularis are not intrinsic parts of the morphologically right ventricle, and that the septal band is not part of the conus, but rather of the morphologically right ventricle.

From the diagnostic standpoint, in view of the severe cono-ventricular malalignment which is the essence of anatomically corrected transposition, a segment-by-segment approach to the angiocardigram and electrocardiogram is necessary to permit accurate diagnosis of these rare anomalies.

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