

CORRECTED TRANSPOSITION OF THE GREAT VESSELS

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The purpose of this paper is to report an example of the rare developmental anomaly corrected transposition of the great vessels and to discuss the concept of corrected transposition.

NOMENCLATURE

It is essential to define certain terms used in this report as there is much variation in usage.

Transposition. In transposition of the great vessels with the cardiac chambers in the normal position, the arterial trunks are altered in their relationship to each other at their origin from the ventricles so that the aorta is moved to a varying degree to the right and front. The general result is that the aorta, being thus moved to the right and anteriorly, is now more closely related to the right ventricle, and with the exception of the mildest degree of transposition the aorta comes to lie in the path of unaerated blood from the right ventricle. Transposition is generally ascribed to abnormal growth or absorption of the bulbar region of the primitive heart tube which interferes with the movement backwards and to the left (clockwise torsion) that normally takes place in this region. The embryological basis of transposition and the classification of various types have been extensively discussed (Spitzer, 1923; Harris and Farber, 1939; Bremer, 1942; Lev and Saphir, 1945; Lev, 1953).

Inversion. When there is inversion (*situs inversus*) as opposed to the normal position (*situs solitus*), an organ (or part of an organ) is formed as a mirror image of the normal with alteration of relation in the transverse plane but not in the antero-posterior plane. Inversion probably occurs at a very early stage of development but the embryological basis is unknown.

Inversion and transposition are thus distinct anomalies, one essential distinguishing feature being that inversion preserves the antero-posterior relations and transposition changes them. The two anomalies may co-exist, however, and the combination of inversion and transposition is much more frequent than would be expected by chance. Such combinations are best designated inverse transposition, and inverse counterparts of most types of transposition have been recorded (Spitzer, 1923 and 1929; Pernkopf, 1926).

Identification of Cardiac Chambers. In cases of inverse transposition individual cardiac chambers cannot be identified by their position, attached vessel, valve or by the type of blood conveyed. The only satisfactory routine method is to designate a chamber according to the following structural features as outlined by Lev (1954).

- (1) The right atrium presents a limbus and fossa ovalis on its septal surface.
- (2) The left atrium presents on its septal surface the irregular configuration of the septum primum as it is adherent to the septum secundum.
- (3) The right ventricle presents a sinus and a conus divided by the crista supraventricularis. Its posterior septal surface is trabeculated and contains a variable number of papillary muscles.
- (4) The left ventricle is without a conus. Its septal surface is only slightly trabeculated and contains no papillary muscles.

Corrected Transposition. Corrected transposition is a term which has been used in several senses. Its meaning can best be elucidated by examining the several types of inverse complete transposition.

In complete transposition (crossed or type 3 of Spitzer) in situs solitus the aorta arises from the right ventricle in an anterior position and receives unaerated blood: the pulmonary artery arises from the left ventricle posteriorly and receives aerated blood. Theoretically inversion may occur in any part of the cardiac tube (bulbar, ventricular, or sino-atrial region) so that eight types of complete transposition result and these are represented diagrammatically in Fig. 1. A1 shows complete transposition in situs solitus and A2 complete transposition in situs inversus; it will be seen that in each case the aorta arises from the right ventricle and receives unaerated blood. In A3 and A4 the aorta arises from the morphological left ventricle but receives unaerated blood; hence it can be regarded as "corrected" anatomically but not functionally. B1 and B2 show both functional and anatomical correction, the aorta arising from the morphological left ventricle and receiving aerated blood. In B3 and B4 the aorta arises from the morphological right ventricle but receives aerated blood; hence it is corrected functionally but not anatomically.

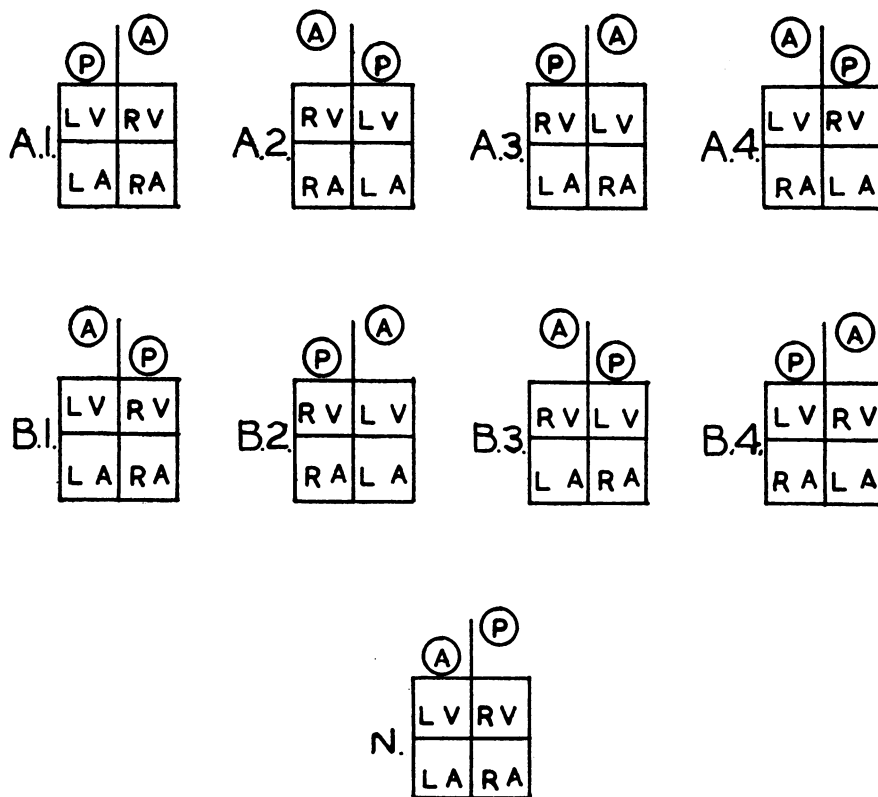


FIG. 1.—Diagram illustrating the various types of complete transposition. A similar diagram of a normal heart, N, is included below for the purposes of orientation. A indicates the aorta; P, the pulmonary artery; LA, LV, RA, and RV, indicate the *morphological* left or right, atrium or ventricle. A1 shows complete transposition in situs solitus; A2 shows the same in situs inversus; A3 shows ventricular inversion; A4 shows sino-atrial and bulbus inversion; B1 shows bulbus inversion; B2 shows sino-atrial and ventricular inversion; B3 shows bulbo-ventricular inversion; and B4 shows sino-atrial inversion (after Harris and Farber).

Thus, in cases of complete transposition of the great vessels, the term corrected transposition has been used in a functional sense to indicate that the aorta receives aerated blood, and in an anatomical sense to indicate that the aorta arises from the morphological left ventricle. In inverse forms of other types of transposition similar concepts of partial correction have been applied.

It would seem best, however, to restrict the use of the term corrected transposition to cases of functionally corrected complete transposition, i.e. cases of types B1-4 where the aorta receives aerated blood whether it arises from the morphological left ventricle or from the right. This is the most common use of the term and is of value from a clinical standpoint, as this small group of cases has a considerably better prognosis than other types of complete transposition.

ILLUSTRATIVE CASE REPORT

Clinical Features. A girl, aged 3 1/2 months was admitted to hospital moribund, with a "slow pulse rate." Death occurred twenty minutes later. There had been two cyanotic attacks earlier in the day. Delivery had occurred normally at full term but during pregnancy the fetal heart rate had been abnormally slow. Examination after birth revealed slight cyanosis, moderate cardiac enlargement, and an apical systolic murmur. The pulse rate was 48 and an electrocardiogram showed 2:1 A-V block. Feeding had been difficult and occasional attacks of cyanosis and dyspnoea had occurred.

Post-mortem Examination. A complete necropsy was performed 20 hours after death. The body was that of a poorly developed girl of 61 cm. length, with moderate cyanosis but no oedema. The only significant findings outside the cardiovascular system were generalized visceral congestion, bilateral pleural effusions of 15 ml., and a peritoneal effusion of 50 ml.

The heart (90 g.) was much enlarged and lay in the left thorax with its long axis directed downwards and to the left. It was roughly triangular in outline with a prominence at the left upper portion of the anterior surface from which the large aorta arose. The pulmonary artery lay posterior to, and to the right of, the aorta. Both vessels coursed directly upwards. The right atrium was large and projected to some extent to the left in front of the great vessels. The small elongated left atrium lay behind the great vessels and its tip just extended to the left of the aorta.

The right atrium received the superior and inferior venæ cavæ and the coronary sinus but the venous valves were rudimentary. The fossa ovalis measured 1.3 × 1.0 cm. and showed a valvular defect measuring 0.7 × 0.3 cm.

The ventricle that lay on the right side (Fig. 2a) resembled a mirror image of a morphological left ventricle. The maximum internal diameters were 4.5 cm. vertically and 2.8 cm. transversely. The wall was 0.8 cm. thick. The atrio-ventricular orifice measured 4.9 cm. in circumference and bore two cusps situated anteromedially and posterolaterally. The chordæ tendineæ of the cusps arose from two large papillary muscles, one anterior and one posterior. The interior of the ventricle showed a fine trabeculation. The upper part of the septum was smooth and showed three defects. One rounded defect A, 0.8 cm. in diameter, lay in the midpart beneath the septal cusp of the pulmonary valve; its walls were muscular and rounded save posteriorly where it was membranous and the medial portions of the atrio-ventricular valves were fused together: the upper part of the septum posterior to this defect was membranous. Two defects B and C, of 0.9 and 0.2 cm. diameter respectively, with rounded muscular walls were present in the most anterior part of the septum.

The pulmonary artery arose from the midpoint of the ventricle that lay on the right side. At its origin it measured 2.2 cm. in circumference. It lay over and behind the septal defect A and anterior to the anterior leaf of the right atrio-ventricular valve. The pulmonary valve possessed two cusps, left and right, of about equal size. The right cusp bore the same intimate relation to the anterior cusp of the right atrio-ventricular valve as the left posterior cusp of a normal aorta does to the anterior cusp of the mitral valve.

The left atrium was slightly smaller than the right atrium. It received four pulmonary veins in a normal manner and showed the small opening of the foramen ovale.

The ventricle that lay on the left side (Fig. 2b) resembled a mirror image of a morphological right ventricle. Its maximum internal measurements were 3.8 cm. vertically and 2.2 cm. transversely. Its wall was 0.7 cm. thick. The atrio-ventricular orifice was 3.7 cm. in circumference and possessed medial, posterior, and antero-lateral cusps. The anterior papillary muscle was slender, bifid, and inserted half way up the ventricular wall. The posterior papillary muscle arose somewhat laterally from the inferior part of the ventricle. The medial cusp was attached by numerous chordæ to the upper portion of a muscular

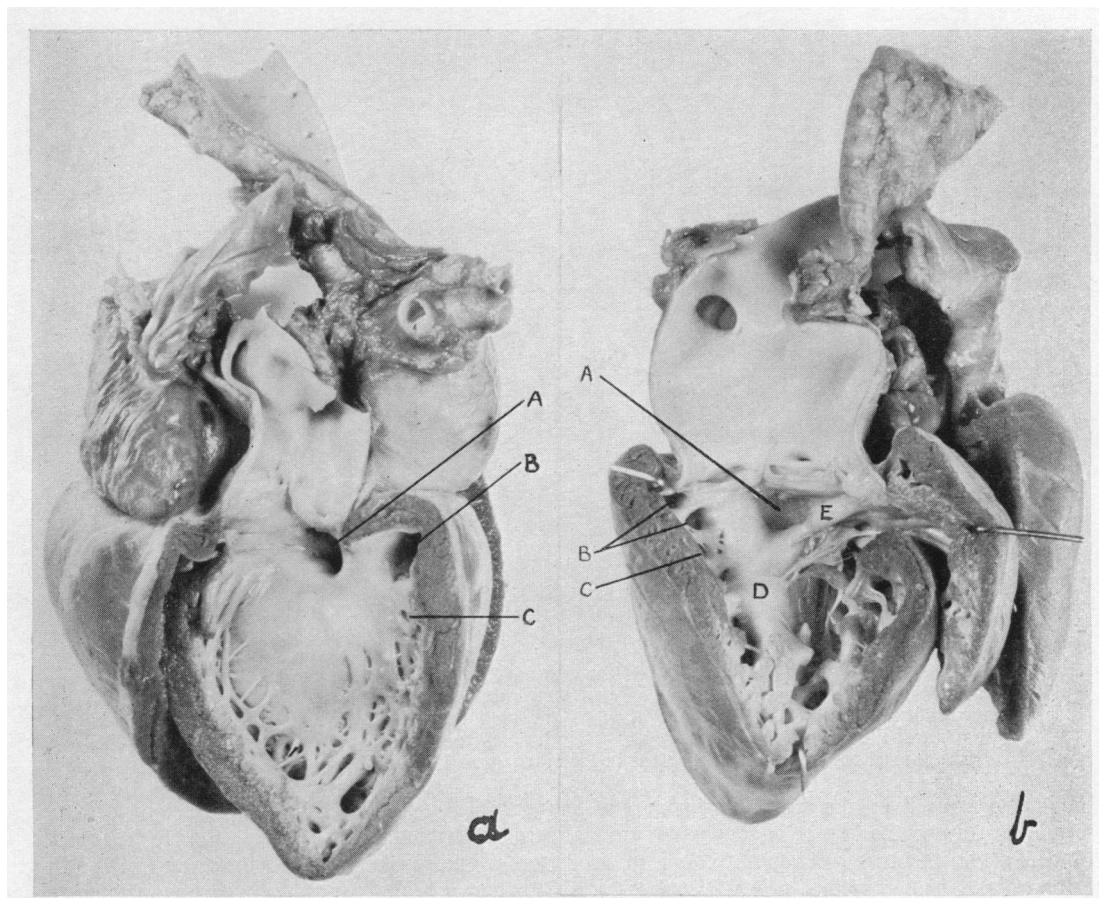


FIG. 2.—Septal aspect of ventricle which lay on (a) the right side (morphological left ventricle), and (b) the left side (morphological right ventricle). A, B, and C=ventricular septal defects. D=crista supraventricularis. E=bulbo-atrial ledge.

band, D (described later), and adjacent ventricle. The posterior cusp was attached by chordæ to the posterior papillary muscle and adjacent ventricle. The antero-lateral cusp was attached by chordæ to the two papillary muscles and the muscular bands D and E. The surface of the ventricular septum was irregular and heavily trabeculated and in its upper portion there were the three defects corresponding to those seen on the right side of the septum. The defect A was centrally situated beneath the posterior cusp of the aortic valve. The large anterior defect B was subdivided by a smooth muscular band and just below it was the smaller defect C. A firm muscle band D arose on the anterior wall of the ventricle close to the apex below the anterior papillary muscle. It coursed to and became incorporated with the septum, and minor bands from the ventricular wall joined it including one from the base of the posterior papillary muscle. The band had a slightly concave anterior margin. It ascended on the left side of the septum and attained a maximum width of 1.0 cm. just below the septal defect A where it divided. The larger part then ascended to the posterior half of the right anterior aortic cusp and its junction with the posterior cusp. The smaller part formed the inferior border of the septal defect A. Anterior to the main band there was a deep recess into which septal defects B and C opened. This band D was interpreted as the crista supraventricularis and it divided the ventricle into a sinus portion (inflow tract) and a conus portion (outflow tract). There was another small muscle band E which arose from the inferior border of the septal defect A, where it was attached by a fibrous septum to the posterior membranous part of the ventricular septum. It arched over the roof of the ventricle between the aorta and the anterior tricuspid leaflet to become incorporated in the upper part of the antero-lateral wall of the ventricle. This band E was interpreted as the bulbo-atrial ledge.

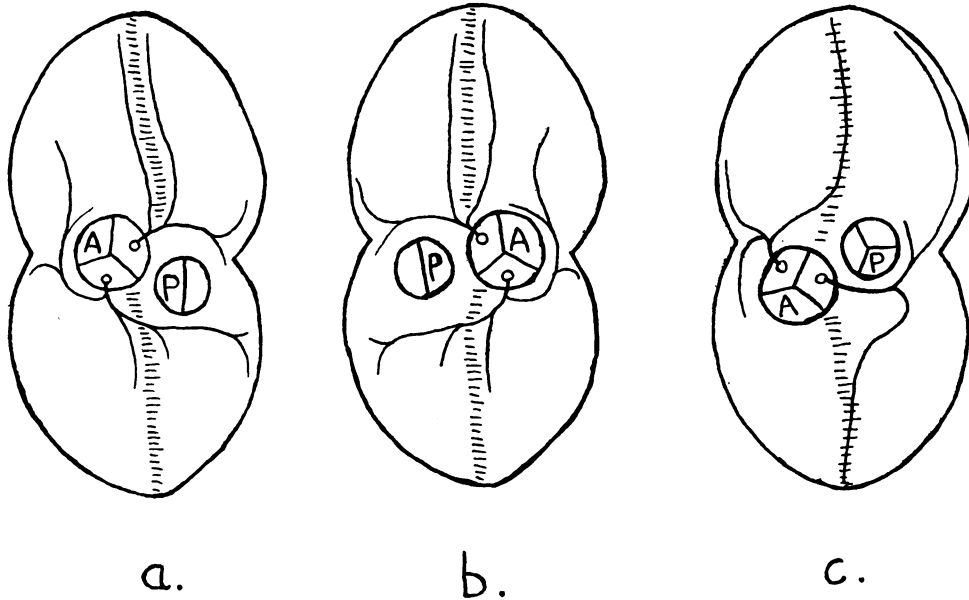


FIG. 3.—Diagram illustrating the course of the coronary arteries in this case (a), its mirror image (b), and the normal distribution (c). The heart is depicted, viewed from above, as though it were sectioned coronally from the apex to the base and opened with the anterior surface above and the posterior surface below. A indicates the aorta; P, the pulmonary artery. The shaded areas denote the septa present (after Harris and Farber).

The aorta arose from the most anterior part of the ventricle on the left side. It did not have the usual close relationship to the left atrio-ventricular orifice, being separated from it by the muscle bands D and E. At its origin the aorta measured 3.9 cm. in circumference and possessed a posterior and left and right anterior cusps. The coronary artery distribution is shown in Fig. 3a. The right coronary artery arose from the right anterior sinus and immediately gave off two large anterior descending branches which coursed on either side of the septum. It continued in the right atrio-ventricular groove giving off several small branches. The left artery arose from the posterior sinus and immediately gave off a large branch which coursed round the left side of the base of the aorta to supply branches to the lateral and anterior wall of the left-sided ventricle. The main stem coursed to the right giving off branches to the posterior septum and the posterior and lateral walls of the right-sided ventricle.

The ductus arteriosus was patent with a lumen of 0.2 cm. diameter. The remaining branches of both aorta and pulmonary artery were normal.

DISCUSSION

This case shows situs solitus of the sino-atrial portion of the heart, which is of normal structure, and of all other viscera. The ventricles, however, are inverted and they form a mirror image not of normal ventricles but of those found in complete transposition of the great vessels (crossed transposition or type 3 of Spitzer). The great vessels show complete transposition as evidenced by their almost parallel upward course, their origin one from each ventricle, the anterior position of the aorta, and the internal structure of the ventricles. In addition the bulbus is inverted as shown by the left anterior position of the aorta and the right posterior position of the pulmonary artery. Furthermore, the distribution of the coronary arteries shows transposition and inversion. This is illustrated diagrammatically in Fig. 3 which shows (a) the coronary distribution in this case, (b) its mirror image, and (c) the normal distribution. Comparing the mirror image of this case with the normal distribution, the left circumflex and anterior descending arteries take origin from the left anterior aortic sinus which was originally in the right anterior position, and the right

circumflex and posterior descending arteries take origin from the posterior aortic sinus which was originally in the left anterior position.

This case therefore presents inversion of the bulbo-ventricular loop coupled with complete transposition of the great vessels and is an example of corrected transposition.

Twenty-four cases of corrected transposition have been recorded. Six are of type B1, complete transposition with bulbus inversion (Grunmach, 1890; Theremin, Case 47, 1895; Lewis and Abbott, 1915; Doerr, Case 1, 1938; Carns *et al.*, 1941; and Brown, 1950). Two cases are of type B2, complete transposition with sino-atrial and ventricular inversion (Gutwasser, 1870; and Fingerhuth, 1901). Fifteen cases are of type B3, complete transposition with bulbo-ventricular inversion (Rokitansky, p. 83, 2 cases, 1875; Tönnies, 1884; Mann, 1889; Lochte, 1894; Theremin, Case 46, 1895; Peters, 1901; Sato, 1914; Wurm, 1927; Stejfa, 1931; Walmsley, 1931; Roos, 1936; Harris and Farber, Case 16, 1939; Liebow and McFarland, Case 1, 1941; Lev, Case 3, 1954). One case is of type B4, complete transposition with sino-atrial inversion (Graanboom, 1891). Some other cases are unacceptable on account of associated lesions such as certain anomalies of the great veins or a common atrio-ventricular valve which impair the functional correction. (Geipel, 1903; Schmincke and Doerr, 1939; Moscovitz *et al.*, 1952).

Analysis of the recorded cases reveals several points of interest even though some of the reports are incomplete. The sexes are equally represented, 10 males and 11 females. The length of life is variable ranging from 9 days to 46 years with an average of 14 years. The outlook, therefore, is poor, though better than in ordinary complete transposition for which Abbott (1936) gives mean survival times of six months with an intact ventricular septum and three years with a defective ventricular septum. The main reason for this poor prognosis is the frequent existence of associated anomalies. Of 18 cases in which the information is given, only two were without other structural cardiac abnormalities and one of these had complete heart block. Septal defects (atrial or ventricular or both) were especially common, and in only four cases were both septa intact. This is not unexpected in view of the abnormal mechanism of septum formation in cases showing inversion and transposition. The septal defects varied considerably in size and some ventricular septal defects were large. Somewhat surprisingly most of the cases with large ventricular septal defects lived considerably longer than the mean for cases of corrected transposition. Of particular interest are the anterior septal defects in Wurm's case and the present case and the anterior septal niche in the case of Liebow and McFarland. According to Spitzer the posterior border of this defect or niche represents the forward directed course of the ventricular continuation of the crista aortico-pulmonalis, thus confirming that the anterior part of the ventricular septum in these cases is derived from the crista supraventricularis.

Another feature of these cases which is probably related to the abnormal septation of the heart is that five cases had heart block, complete in three, 2:1 A-V block in one, and right bundle-branch block in one.

Finally, in eight cases the coronary artery distribution showed inversion and transposition thus supporting the hypothesis that the cases are to be regarded as forms of inverse transposition. That other mechanisms may account for some cases is suggested by the finding that in two cases the coronary artery distribution showed only inversion.

SUMMARY

The concept of corrected transposition of the great vessels is discussed and it is suggested that the use of the term should be restricted to cases of functionally corrected complete transposition.

A case of corrected transposition is reported and compared with previously recorded cases.

Corrected transposition is a rare anomaly. It is commonly associated with other congenital heart lesions and although the prognosis is poor it is much better than in the usual forms of complete transposition. The genesis of the lesion remains obscure but it appears to involve both transposition and inversion of various parts of the cardiac tube.

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