DEXTROCARDIA AND ISOLATED LÆVOCARDIA
I: ISOLATED LÆVOCARDIA

BY

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Isolated levocardia, with the heart on the left but with the abdominal viscera inverted, is less well known than its counterpart, isolated dextrocardia, but is not very rare. In this paper we are reporting our findings in 20 patients, and Campbell (1963) has given reasons for thinking that its incidence may be about 1 in 22,000 of the general population.

In any series of patients with cyanotic heart disease, there will be some with isolated levocardia and others with dextrocardia. Among our first 670 cyanotic patients, seen in 1947–53, there were nearly 2 per cent with isolated levocardia, nearly 3 per cent with isolated dextrocardia, and nearly 1 per cent with situs inversus and other malformations of the heart (Table I). This means that nearly 6 per cent have complete or partial inversion of the viscera, compared with rather less than 1 per cent among those with acyanotic heart disease. Berri (1958), among 2695 patients with cyanotic or acyanotic congenital heart disease at the Hospital de Niños de Buenos Aires, and Heim de Balsac et al. (1954) among 1100 similar patients in Paris, found about the same proportion with dextrocardia (2-2% and 2-1% against our 2-5%) and a rather smaller proportion with isolated levocardia (0-4% and 0-6% respectively against our 1-2%).

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<td><strong>Incidence of Dextrocardia and Levocardia among Patients with Other Congenital Heart Disease (C.H.D.), and Age Distribution of those with Levocardia</strong></td>
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* Figures in parentheses are percentages.
Although we think isolated levocardia and isolated dextrocardia must be considered together because of their mirror-image relationship, this first paper is confined to a description of our cases of levocardia with a short preliminary discussion.

Our first 14 patients with isolated levocardia, all with other malformations of the heart, were reported by Campbell and Forgacs (1953) who divided them into two groups. There were 6 with venae cavae and the venous atrium on the left side, and 8 where they were on the right, as normally: 5 of each group had been fully investigated with necropsies on 4 of them, and a fifth necropsy was reported later by Campbell (1960). Since then, we have seen 6 more patients, and the venous atrium was on the left in 5 and on the right in only 1, making our total figures 11 with the venous atrium on the left and 9 with it on the right side. Where there is a common atrium the patients are grouped according to the position of the venae cavea. Generally the P wave in lead I is inverted when the venous atrium is on the left, and upright when it is on the right, as normally.

The sex and age incidence of the 20 patients are shown in Table I. There were about equal numbers of each sex. Most of them (80%) were under 10 years and only one was over 20 years of age.

We know that in all these cases the heart was in its normal position pointing to the left and that the stomach and generally the liver (and probably all the abdominal viscera) were inverted.* Our classification depends on the side on which the venae caveae and the venous atrium are placed, regardless of any theoretical question about the morphological identity of the right and left atrium though we think this reasonably certain. The embryological development of the atria supports the view that the atrium receiving the venae caveae is the morphological right atrium. When the abdominal viscera are inverted, and the superior venae caveae (S.V.C. and I.V.C.) and the venous atrium are all found on the left side, the venae caveae have probably been inverted with the abdominal viscera, and the left-sided venous atrium is probably a morphological right atrium that has been inverted. When, on the other hand, the venous atrium and the venae caveae have remained on the right, it is almost certain that it is a morphological right atrium, in spite of the inversion of the abdominal viscera: this is supported further by the fact that PI is generally upright.

When the abdominal viscera are inverted, one might expect the venae caveae and, therefore, the venous atrium also to be inverted and on the left side. Keith, Rowe, and Vlad (1958) found this was so in about two-thirds of their reported cases, but we have found the proportion only a little over one-half (11 of 20 cases). Campbell and Forgacs (1953) found it rather less than half in the earlier part of this series and much less than half in the reported cases they collected. Probably, therefore, there is no great difference in the frequency of these two forms of levocardia, those with the venae caveae and venous atrium on the left and those with them on the right.

I. LEFT-SIDED VENÆ CAVÆ AND VENOUS ATRIUM (CASES 1–6 AND 16–20: GROUP L.III)

This group consists of 5 of the 6 new cases, shown in Table II, and 6 of the 14 earlier ones: all 11 had full investigations and 4 of them had necropsies (Cases 1, 3, 18, and 20). In all 11, the S.V.C., and the I.V.C. also in all 8 where its position was known, were on the left, i.e. inverted in the same

* Sometimes, however, the liver was not completely inverted but was almost symmetrical. Thus, it occupied both sides of the upper abdominal cavity about equally in Cases 7, 8, and 9 of our group L.IV (see discussion). The peritoneal attachments suggested that the liver was inverted in Case 7, and the gall-bladder was on the left side in Case 8, but on the right side in Case 9.

In these two (Cases 8 and 9) there was no spleen, but we did not find as much evidence of symmetry as Ivemark (1955) did in his series with splenic agenesis. Although 85 per cent of his cases had some degree of situs inversus, the degree of inversion of the abdominal viscera was often incomplete, especially for the liver, and the lungs were often symmetrical, each with three lobes. The lungs each had three lobes in our Case 9, but the right had 4 lobes and the left 3 in Case 8. In Case 9, where the right S.V.C. reached the right side of a common atrium, the left S.V.C. drained directly into its left side; but we did not find any example with bilateral S.V.C. where the left S.V.C. drained into a left atrium, instead of reaching the right atrium through the coronary sinus. Ivemark (1955), however, found this quite common in his series. Another case of isolated levocardia (our group L.IV) with partial inversion of the abdominal viscera has been reported by Gilber-Queraltó, Nolla Panadés, and Carrasco Azemar (1960).
way as the abdominal viscera (Fig. 1). In one of these (Case 16) there was a right S.V.C. also but it reached the left side through the coronary sinus.

The venous atrium was on the left in 10 of these 11 cases, and in the eleventh (Case 16) there was virtually a common atrium. With this qualification, the left-sided venous atrium was always thought to be a morphological right atrium that had been inverted. The pulmonary veins appeared to drain into the right-sided atrium (or the right side of a common atrium) in 10, but in Case 20 they joined the venous (left-sided) atrium, so there was anomalous pulmonary venous drainage in addition.

The first 6 cases were consistent in showing an inverted P wave in lead I, but in the 5 later ones its direction was less consistent and, therefore, less useful in diagnosis. In the 11 cases, PI was constantly inverted in 8, sometimes inverted and sometimes upright in 2, and small but upright in 1 (Case 18, an infant of 5 months with multiple malformations).

It is more difficult to know whether or not the ventricles are inverted in these cases, and Campbell and Forgacs (1953) did not try to answer this question. In 2 of the 4 with a necropsy (Cases 18 and 20) there was a common ventricle, and in the other 2 (Cases 1 and 3) the tricuspid A-V valve was on the left side and probably the ventricles were inverted, though in Case 1 the left-sided (presumed morphological right) ventricle was the more capacious and hypertrophied. Among those without necropsies the ventricles were thought to be inverted in Case 6 but not in Cases 16, 17, and 19. In the remaining 3 (Cases 2, 4, and 5) we have no good evidence as to whether the ventricles were inverted or not. In the group as a whole, therefore, both ventricles and atria were inverted in 3, the ventricles were not inverted though the atria were (mixed levocardia) in 3, there was a common ventricle in 2, and in the last 3, the condition of the ventricle was uncertain.

Transposition of the great trunks needs special consideration in dextrocardia and isolated levocardia, and has been discussed fully by de la Cruz, Polansky, and Navarro-López (1962), by Shafer and Johnson (1963), and by Shafer (1964). Complete transposition was shown at necropsy in Cases 18 and 20, but both these had a common ventricle so it was of little practical consequence. In Cases 1 and 3 (with necropsies) and in Case 6, both the aorta and left ventricle were inverted, so there was corrected transposition, probably complete in two but with the pulmonary trunk overriding the V.S.D. in Case 1. This state of affairs should have allowed a circulation that was substantially normal: this was so in Case 6, but in the other two the presence of pulmonary stenosis and V.S.D. made them cyanotic from an early age.

In Cases 16, 17, and 19, on the other hand, the aorta arose from a left-sided (morphological left) ventricle and the pulmonary trunk from a right ventricle, so there was no transposition of the great vessels. We thought, with less certain evidence, that the situation was the same in Cases 2, 4, and 5.

**Fig. 1.**—Diagrammatic coronal section through the heart of Case 1, showing inversion of the large veins and atria in isolated levocardia. The left-sided A-V valve was tricuspid and the right-sided one bicuspid, so probably the ventricles also were inverted. There was complete transposition of the main arterial trunks. His clinical and anatomical features were described and illustrated as Case 1 by Campbell and Forgacs (1953). Severe pulmonary stenosis and a V.S.D. prevented a normal circulation. S.V.C. and I.V.C., superior and inferior vena cæ; P.A., pulmonary trunk; P.V., pulmonary veins; A, aorta.
If so, the circulation in these six patients with atrial inversion would have been impossible without the additional malformations.

The aorta arched to the right in all our earlier 6 cases and in 3 of the 5 later ones, but to the left in the other 2 of these. It is, therefore, much commoner to find a right-sided (9) than a left-sided (2) arch.

**Other Associated Malformations.** An atrial septal defect was present in 10 of these 11 cases, and in 1 it was large enough to produce what was virtually a common atrium (Case 16). A ventricular septal defect was thought to be present in all except one, but was proved in only 5 of them, because when there is an A.S.D. it may be difficult to be sure. In 2 (Cases 18 and 20, both with necropsies) there was a common ventricle.

Transposition of the great trunks has been discussed. The presence of pulmonary stenosis was proved in 4 and suspected in 2 others: certainly 2 and probably 1 other had pulmonary atresia making 9 of the 11 with probable pulmonary stenosis or atresia. A tenth was thought to have tricuspid atresia (Case 5) and this was present as well as pulmonary atresia in one of those already mentioned (Case 18); and the last (Case 6) was acyanotic.

Apart from these generalizations, each patient has so many different individual features that short notes of the new Cases 16–20 are given in an appendix. One of the few exceptions is that Case 19 hardly needs description, because of the very close resemblance to Case 17. Case 6 of our earlier series has since been fully investigated and, as she is our only acyanotic patient who has been investigated and as she had unusual features, she is described fully.

**Case 6.** A girl, aged 4 (1952), looked healthy and had no disability or cyanosis, though a loud systolic murmur had led to a diagnosis of congenital heart disease when she was a few weeks old. Her apex beat was felt well out to the left though her abdominal viscera were inverted. The provisional diagnosis was isolated levocardia and aortic stenosis.
When she was 12 (1960) she still seemed just as well and had angiocardiography. The S.V.C., I.V.C., and venous atrium and ventricle were all left-sided (Fig. 2) though morphologically the atrium was almost certainly the right atrium and the ventricle probably the right ventricle. There was no visible obstruction to the outflow tract or at the pulmonary valve (Fig. 2). The blood returning from the lungs entered a right-sided (morphological left) atrium (Fig. 3) and a right-sided ventricle that on its smooth outline was thought to be a morphological left ventricle. The aorta filled from this ventricle and lay to the right and arched to the right. The ventricular septum seemed intact and lay almost vertically in the sagittal plane, so that the two ventricles lay to the right and to the left rather than in their normal antero-posterior relation. There was corrected transposition of the great trunks, the aorta being to the right and the pulmonary trunk to the left and posterior (Fig. 4). The circulation was thought to be normal, though a diagnosis of left-sided A-V (tricuspid) regurgitation was made, as the murmur was now heard better towards the apex and was not like an ejection murmur.
II. Right-sided Vena Cavae and Venous Atrium (Cases 7–15: Group LIV)

Of these 9 cases 6 were fully investigated and 2 of them had necropsies also. In 3 of the 6 the right-sided S.V.C. and I.V.C. both drained to the right atrium (Cases 8, 10, and 11), and in one of these a left S.V.C. also reached this chamber through the coronary sinus. In 2 others there were bilateral S.V.C. but no I.V.C. (Cases 7 and 15): the azygos vein carried the blood from the lower part of the body and reached the right side of the common atrium in each case, in one directly through the right S.V.C. and in the other through the left S.V.C. and the coronary sinus. In the sixth (Case 9), the right S.V.C. reached the right side and the pulmonary veins reached the left side of the common atrium, but the I.V.C. and the left S.V.C. also reached the left side so it is less certain that this case should be included here.

The pulmonary veins drained into the left atrium in 3 cases, though in one of these (Case 8) its only exit was through an atrial septal defect to the right atrium. They drained into the left side in 2 (Cases 7 and 9) but not in the third (Case 15) of the 3 that had a common atrium.

The P wave in lead I was upright in 8 of the 9 in this group, but in Case 15 with a common atrium it was small and biphasic. With the qualifications that are necessary in the presence of a common atrium, we think that the atria were not inverted in any case in this group.

In Case 15 the ventricles were not inverted and we think they were not inverted in Cases 10 and 14 but probably were in Case 11. In Cases 7, 8, and 9, with necropsies, there was a common ventricle. It seems probable, therefore, that the ventricles are generally not inverted in this group though they may be sometimes.

In Cases 7, 8, and 9 there was anatomical transposition of the great trunks though functionally this was not significant because of the common ventricles, and in one of them because of pulmonary atresia also. In Case 10 there was complete transposition of the great vessels, in Case 11 we could not say, and in Case 15 and probably in Case 14 there was not.

The aortic arch was right-sided in 5 and left-sided in 4 of these cases, so there was no preponderance either way as there was in the previous group where it generally arched to the right. Of the 6 investigated cases, 5 had bilateral S.V.C. and the last a right S.V.C. only, while in the previous group bilateral S.V.C. were found only once. Campbell and Forgacs (1953) drew attention to this difference.

Other Associated Malformations. In 6 of 8 cases there was a defect of the atrial septum and in 3 this was large enough to produce a common atrium (Cases 7, 9, and 15). In 4, or probably 5, of the 8, there was a defect of the ventricular septum and in 3 this was enough to produce a common ventricle (Cases 7, 8, and 9).

In 3, or probably 4, of the 8 there was pulmonary stenosis, and in 2 others pulmonary atresia (Cases 7 and 12). The seventh had pulmonary hypertension and anomalous pulmonary venous drainage (Case 15), and the eighth, the only one who was acyanotic, had aortic regurgitation probably from bicuspid aortic valves (Case 14). We have not included a ninth, though clinically we thought she had pulmonary atresia (Case 13). In Cases 7 and 15 there was no inferior vena cava, the drainage from the lower part of the body being through an azygos vein.

Case 15 is reported in more detail since she had unusual malformations, i.e. anomalous pulmonary venous drainage, a common atrium, an absence of the inferior vena cava, and pulmonary hypertension. Without the first two, her circulation would have been normal functionally in spite of the isolated left atrioventricular.

Case 15. A girl, aged 15 in 1954, was cyanosed and moderately disabled. Both S.V.C. reached the right side of the common atrium, the small right one directly and the large left one through an enormous coronary sinus (Fig. 5A). There was no I.V.C. and the drainage from the lower part of the body was through a left azygos vein that joined the left S.V.C. The pulmonary veins also drained into the right side but posteriorly. There was such a large A.S.D. that there was virtually a common atrium with much mixing of venous and arterial blood in the atrium. Hepatic veins from the right and left sides of the liver drained directly into the right and left sides of the atrium, and as the liver was inverted the veins on the left were the larger.
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FIG. 5.—Angiocardiogram from Case 15. (A) From left femoral vein showing opaque media passing through the left azygos vein, the large left S.V.C. and coronary sinus, to the right side of the common atrium. (B) Left ventricular angiocardiogram (from femoral artery) showing a small left ventricle which from its smooth outline was thought to be a morphological left ventricle in spite of the apparent outflow tract. The small aorta, arching to the left, is well opacified.

With the exception of the left hepatic veins, all the systemic veins drained into the right side so that this would have compensated for the inversion of the abdominal viscera, but the pulmonary veins also reached the right side, so that the venous and arterial blood would have been mixed even if the atrial septum had been normal. The P wave in lead I was small and biphasic so did not help in deciding about inversion of the atria.

The large pulmonary trunk arose from a right-sided ventricle. The small aorta arose from a left-sided ventricle, which was long and slim (Fig. 5B) with a rather narrow outflow tract, but its smooth outline favoured it being a morphological left ventricle. We have assumed, therefore, that the ventricles were not inverted. The aortic arch was left-sided and the descending aorta lay on the left side as normally.

When she was 22, she was increasingly disabled so had a thoracotomy, but nothing could be done as the pulmonary arterial pressure (140/80) was slightly higher than that in the aorta (130/80). Her condition has not changed during the year that has elapsed since.

ASSOCIATED MALFORMATIONS AND PROGNOSIS OF OUR CASES

The other malformations associated with isolated laevocardia seemed to be much the same in our two groups, though a common atrium was more frequent in group L.IV (3 of 8 against 1 of 11 for group L.III). A common ventricle was also more frequent but the difference was less (3 of 8 against 2 of 11 in group L.III). Other differences were slight and the numbers are hardly large enough to be sure about the common atrium and common ventricle.

Taking the two groups together, atrial septal defects are present in about three-quarters, including one-quarter with a single atrium. Ventricular septal defects are found in at least half and perhaps in three-quarters, again including one-quarter with a single ventricle. Pulmonary stenosis or atresia was proved to be present in about 80 per cent. Transposition of the great trunks, generally complete and often corrected, was present in more than half our cases. Other malformations, including
tricuspid atresia and anomalous pulmonary venous drainage, and probably many others in a larger series, may occur, but none of these is common in the way that septal defects and pulmonary stenosis and atresia are. In cases that have been reported by others the findings are generally similar.

The age incidence of our patients has been given in Table I. Most of them (80%) were under 10 years and only one was over 20 years when first seen. As for other groups with malformations of the heart, we have seen relatively few affected infants aged 3 years and under, but think the proportions of children of other ages and of adults are representative of the total number living. The early mortality must therefore be heavy and probably at least three-quarters of these patients die in the first ten years and half in the first five years of life.

Prognosis. Though one more patient in our group L.IV had died (5 of 9) than in group L.III (4 of 11), we do not think there is any significant difference in the prognosis of the two groups. We have, therefore, considered it for both together, but have taken the acyanotic cases (one from each group) separately from the cyanotic ones.

Cyanotic Cases. Of the 18 cyanotic cases, 9 had died and 9 were still alive when last heard of (most of them in the last two years). The prognosis is poor, since 4 died before they were 5, generally in cyanotic attacks (Cases 1, 12, 13, and 18), and another 4 died after operations before they were 7 and did not seem likely to have lived very long without this (Cases 7, 8, 9, and 20). The last (Case 3) kept much the same for six years, but died at the age of 20 after recovering from the immediate effects of subacute bacterial endocarditis (Campbell, 1960).

Of the 9 living cyanotic cases, 1 has done extremely well for 12 years after a subclavian-pulmonary anastomosis when he was 6 (Case 16); 1 severely cyanosed woman has hardly changed from 19 to 30 years of age (Case 2); 5 did not change much during six years of observation (Cases 4, 5, 10, 11, and 19) but the eldest was only 12 years old; and the last 2 have lost a good deal of ground from 9 to 17 years and from 15 to 23 years of age respectively (Cases 15 and 17). Case 16 was the only patient in our series who benefited from operation, though the results of Young and Griswold (1951) were more favourable. A subclavian-pulmonary anastomosis might be expected to help in most cases with a diminished blood supply to the lungs, though probably less than in uncomplicated cases of Fallot's tetralogy, but this has not been so in our experience.

Acyanotic Cases. Only 2 of the 20 were acyanotic, one with and one without atrial inversion. Both have continued to lead a normal life during 10–13 years of observation and the girl (Case 6) is now 14 and the man (Case 14) 39 years old. The latter (group L.IV) had no symptoms and was thought to be too well to need any special investigations: moderate enlargement of his heart was attributed to some degree of aortic regurgitation that may have been due to bicuspid aortic valves. His superior vena cava was on the right side and his aorta was normally placed and arched to the left, so that we think his circulation was completely normal—an unusual example of what might be expected more often. In Case 15 also the circulation would have been functionally normal if the pulmonary and systemic veins had not both reached the right side of a common atrium. Case 6 (Group L.III) has been reported fully. Here the venous atrium was inverted to the left side and joined a left-sided morphological right ventricle from which the pulmonary trunk arose, since it also showed inverted (corrected) transposition. One of the earliest reported cases (Hardy, 1833) may have been of this type as he was a man who died of cholera in Calcutta: details about the heart are scanty but it seems that the venous atrium was on the left side. In Case 14 there was no inversion of heart or blood vessels (only the abdominal viscera being inverted), while in Case 6 there was complete inversion of the large veins and arterial trunks and of the chambers of the heart but not of its position.

Schmidt and Korth (1954) agree that both varieties are generally accompanied by other major malformations, but found uncomplicated cases by no means rare. Their 71 cases were reported ones and so may not give a true picture of incidence, but in solitus levocardia (our group L.IV with Case 14) uncomplicated cases formed nearly double the 10 per cent we have found, while in levoversion (group L.III) they found only one example. Rosenbaum, Pellegrino, and Treciokas (1962) have, however, described three cases without any disability as acyanotic levocardia. Two of these, a man
Dextrocardia and isolated levocardia must be considered together because of their mirror-image relationship, so a fuller discussion will be deferred until we have reported our cases of dextrocardia. Complete situs inversus is accepted as a mirror-image of the normal. Less emphasis is generally laid on the fact that each form of isolated dextrocardia is a mirror-image of a form of levocardia, though it is often implicit in the authors’ discussions and has been stressed by Shaher and Johnson (1963).

A classification that will be generally useful and widely used must be based on features that can be recognized in life. We have, therefore, based it on three features: (1) the side on which the heart and its apex lie, (2) the side on which the superior vena cava and the venous atrium lie, and (3) the position of the abdominal viscera, normal or inverted. Sometimes the heart is rather centrally placed and it is not easy to say whether the apex points to the right or the left. Thus Case 16 was wrongly diagnosed as having situs inversus until his angiocardiogram was studied, and in Case 20 the original correct diagnosis was wrongly changed to that of situs inversus at his first admission to hospital. Because of this, some authors have introduced a further group of mesocardia, but generally the true position should be clear after radioscopy, and certainly after angiocardiography.

When the heart is normally placed on the left, the venous atrium may be on the right, as normally, or on the left, and in each case the abdominal viscera may be normally placed or inverted, making four possible patterns. These are shown on the left of Fig. 6 and the four patterns of dextrocardia on the right, each being an exact mirror-image of one of the patterns of levocardia, D.I. of L.I., D.II of L.II etc. Only the forms where the atria and ventricles are both normally placed or both inverted are shown in Fig. 6.

Theoretically the ventricles may be “inverted” when the atria are not or vice versa, and this allows another 8 patterns—the mixed levocardia and mixed dextrocardia of Lev (1954) and Lev and Rowlett (1961). The patterns shown in Fig. 6 where the atria and ventricles behave in the same way are indicated in the text as L.IIIa, L.IVb, etc. The patterns not shown in Fig. 6, where the atria are normally placed and the ventricles are inverted or vice versa, are indicated as L.IIIb, L.IVb, etc. We agree that this classification according to the true morphology of the ventricles is the ideal that should be aimed at, but it is often difficult to apply to living patients because of the uncertainty about inversion of the ventricles or because of the presence of a common ventricle.

The term inversion has been used in two senses. Inversion of the heart (or abdominal viscera) means a complete mirror-image with reference to the mid-line of the body. When we speak of "inverted" ventricles (or atria) we mean inverted with reference to the long axis of the heart. Bulbo-ventricular inversion is the best known example of inversion of the ventricle without inversion of the heart as a whole. If this process were not possible we should be limited to 8, rather than to 16, patterns. Strictly speaking we should refer to mirror-image inversion of the heart as a whole, and axial inversion of the chambers alone, but the context should make the sense clear without this.

L.Ia is the pattern of all normal hearts and of those with any malformations other than dextrocardia or isolated levocardia. L.Ib is the pattern of the common form of bulbo-ventricular inversion. D.I is the pattern of all hearts with complete situs inversus, whether or not there are additional malformations.

L.II is the pattern with everything normal except for inversion of the heart chambers. We have
These 5 forms are common or relatively so.

FIG. 6.—Diagrammatic representation of the eight possible patterns, where (1) the position of the heart, (2) the position of the chambers, and (3) the position of the abdominal viscera may be normal or inverted. In all these 8 patterns the ventricles behave in the same way as the atria, i.e. both normal or both inverted. Theoretically there are another 8 patterns where the ventricles behave differently from the atria, i.e. are inverted if the latter are normal and vice versa.

seen no example of this type and they would have to be looked for among examples of bulboventricular inversion rather than those of isolated levocardia. We shall, however, be reporting one example of D.II.

L.III, with the venous atrium on the left, is the pattern of one common form of isolated levocardia (11 of our 20 cases; 2 of L.IIIa; 3 certainly and 3 others possibly of L.IIIb pattern; and 3 uncertain). D.III is the pattern of the common form of isolated dextrocardia that is generally called dextroversion.

L.IV, with the venous atrium on the right as normally, is the second common form of isolated levocardia (9 of our 20 cases: 2 of L.IVa, 1 of L.IVb; 3 uncertain; and 3 not placed because of a common ventricle). D.IV is a much less common pattern of isolated dextrocardia.

In D.III the heart is generally assumed to have “swung over” to the right instead of to the left as normally; or more likely, as suggested by Grant (1958), remained in the earlier embryological position when it was pointing to the right. Whether this is the correct explanation of cases of “dextroversion” will be discussed later; but, if it is, the pattern L.III is probably produced in the same way by inversion of the heart as well as of the abdominal viscera, and then by a second abnormality of development, i.e. the heart remaining in its earlier embryological position (in the same way as D.III but pointing to the left instead of to the right because of the earlier inversion). Schmidt and Korth (1954) have accepted this explanation and called one group “levoversion” (see
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later). Campbell and Forgacs (1953) assumed that the venae cæ and atria were inverted with the abdominal viscera, though when Case 6 was first examined by radioscopy, we wrote that she looked like an example of complete situs inversus where the heart had been rotated through 90° back to the left-sided position.

The concept of levoversion may seem less likely because it depends on two separate errors of development and the explanation of Campbell and Forgacs may be correct. If so, dextroversion (D.III) could be explained as inversion of the heart as a whole and of the ventricles, but not of the atria because they have remained in their normal position in the same way as the abdominal viscera.

The reported cases of isolated levocardia have been reviewed by Forgacs (1947), Doliopoulos and Maillet (1952), Moscovitz, Gordon, and Scherlis (1952), and Campbell and Forgacs (1953), but the most complete review is by Schmidt and Korth (1954). They found reports of 91 cases and divided them into three groups: (1) levoversion cordis, where there was situs inversus that was complete except that the heart was turned to the left (31 cases); (2) solitus levocardia, where the abdominal viscera, or at least the stomach and liver, were inverted but the heart, including the atria, was normally placed (40 cases); and (3) levositiotio cordis where there was situs inversus totalis with the heart displaced to the left as the result of congenital or acquired changes in adjoining organs (21 cases).

We have not considered cases of their third group where organs other than the heart are mainly responsible for the condition. In their first group corresponding with our group L.III, they say that PI is generally inverted and that the aortic arch is generally right-sided, with both of which statements we are in agreement. In their second group corresponding with our group L.IV, they say that PI is upright, as we find, but they add that the great vessels are radiologically normal with the arch left-sided, while we find it about as common for the aorta to arch to the right as to the left.

The paper of Lev and Rowlatt (1961) on mixed levocardia is hardly pertinent though it contains so many cases, because most of them have a heart on the left and normally placed abdominal viscera, i.e. bulbo-ventricular inversion (our pattern L.Ib). Only 2 of their own 13 (Cases 11 and 13) had inversion of the abdominal viscera, the former being of our L.IIb pattern; the latter was in their rare third type where the chambers were normally placed but wrongly connected. We have no example of this last type with isolated levocardia but think we have one with dextrocardia to be reported. Lev, Licata, and May (1963) have shown that in one case with ventricular inversion, the A-V node and the conducting system also were inverted.

The Circulation. Whether the circulation can be effective depends on the relationships of the venae cæ to the pulmonary trunk and of the pulmonary veins to the aorta, without much regard being paid to the chambers in between. We know of no evidence that a morphological right atrium cannot deal equally adequately with the systemic or pulmonary venous return, and the right ventricle at birth is as well equipped as the left to deal with the systemic pressure. Some of the features on which a ventricle is identified as morphologically right or left may depend on the flows and pressures to which it has been subjected, i.e. on its functional work rather than on its embryological origin.

It is sometimes thought that transposition (corrected transposition) is necessary to allow the circulation to function reasonably, but this is true only under certain conditions. If a venous right atrium connects with a morphological right ventricle, transposition of the great trunks would make the circulation impossible without other malformations. If on the other hand it connects with a morphological left ventricle transposition of the great trunks will be a great help in allowing the circulation to function. If there is a venous left atrium, the same considerations apply with appropriate modifications.

Transposition of the great vessels is common in all our groups of isolated levocardia and dextrocardia. This is probably not because patients with transposition are more likely to survive, but because the abnormal positioning of the ventricles makes it difficult for the complex spiral septation of the truncus arteriosus to join accurately with the ventricular septum.

The position can be summarized by saying that if there are no changes from normal positioning
to inversion of different parts of the heart (as normally and as in cases of complete situs inversus) or two changes, the circulation should be normal in the absence of additional malformations. If, on the other hand, there is one such change only, the circulation will be very abnormal, and impossible in the absence of additional malformations.

Applying these general considerations to our cases with isolated levocardia might lead one to expect that the circulation would be more normal and the prognosis better when the venous atrium was on the right as normally, provided there is not any transposition (see Case 14, p. 76). Our findings do not support this supposition; and transposition and other malformations, especially pulmonary stenosis or atresia with ventricular and often atrial septal defects, do not seem to be significantly different in our two groups (L.III and L.IV).

SUMMARY

We have put forward a classification of isolated levocardia and of dextrocardia that covers all the varieties of mirror-image inversion of the heart as a whole and axial inversion of its separate chambers. This emphasizes the exact mirror-image relationship of every form of levocardia, whether isolated or normal, with one form of dextrocardia.

We group them according to the position of (1) the superior vena cava and, when there is not a common atrium, on the position of the venous atrium also, in addition to the position of (2) the heart and of (3) the abdominal viscera. This means that there are four patterns of levocardia and four of dextrocardia. Several of these theoretical patterns are quite common, but others appear to be rare or unknown.

We have reported 6 new cases of isolated levocardia and discussed them with 14 earlier ones that were reported by Campbell andForgacs (1953). Our classification divides them into two roughly equal groups: (1) 11 with the venæ cæœ and venous atrium on the left side, i.e. inverted in the same way as the abdominal viscera (group L.III); and (2) 9 with the venæ cæœ and venous atrium on the right side as normally, in spite of the inversion of the abdominal viscera (group L.IV). In the former group, PI is generally inverted and the aortic arch is right-sided: in the latter, PI is generally upright and the aortic arch may be left- or right-sided.

There do not seem to be any great differences between these two groups of isolated levocardia as regards the additional malformations present; nor as regards the poor prognosis, though the second group (L.IV) might be expected to have a better outlook provided there were no additional malformations, because the venæ cæœ and atria are oriented correctly with respect to the heart.

About one-tenth of the patients (one in each of our two groups) are acyanotic; these get on well and lead a normal or near normal life, though generally they have some other minor malformation. The great majority of the patients, about nine-tenths, are cyanotic. Nearly all these have other major malformations of the heart in addition to the isolated levocardia, and in most of those of group L.IV this is the cause of their cyanosis. Pulmonary stenosis or atresia is present in four-fifths; atrial and ventricular septal defects are each found in three-quarters, and are large enough to produce a common atrium or common ventricle, each in about a quarter; transposition of the great trunks, either complete or inverted and often corrected, occurs in more than half; and many other malformations are not uncommon.

The prognosis is poor and probably more than three-quarters die in the first ten years of life. Those who reach this age may, however, carry on for years without much further deterioration. Subclavian-pulmonary anastomosis or pulmonary valvotomy would seem likely to help many of them, but we have rarely found that it does.

We have commented shortly on the effects of the two common forms of isolated levocardia on the circulation and on some other aspects. We have, however, deferred a fuller discussion, especially as regards the pathogenesis, until we have reported a study of our cases of dextrocardia, since the exact mirror-image relationships of levocardia and dextrocardia make it essential that the two conditions should be considered as a unity.
ADDENDUM  

Since completing this paper we have been shown a further case by Dr. M. Joseph which we mention as it widens further the spectrum of abnormalities found in these patients.

A girl aged 14 months presented with a history of recurrent bronchitis with cyanosis and clubbing. The pulse rate was only 40 a minute and complete heart block was confirmed. There was a long murmur over the base of the heart with a late systolic crescendo which split over into diastole. The chest radiograph confirmed levo-cardia with inversion of the abdominal viscera and showed great cardiac enlargement with asymmetry of the pulmonary vasculature, the right lung being relatively oligemic. The P wave was upright in I and aVL but inverted in II, III, aVF, and V1, suggesting so-called “coronary sinus rhythm”. Attempted angiocardiography from a saphenous vein demonstrated a left-sided I.V.C. which drained via the azygos system into a left S.V.C., thence via the coronary sinus to the atrium. Injection into the right subclavian vein showed a greatly dilated right S.V.C. from which opaque medium passed refluxu into a vessel passing medially and posteriorly, suggesting a common pulmonary vein. The right S.V.C. drained into an atrial chamber which seemed to occupy the position of both right and left atria. A huge dilated, apparently single ventricle filled from this. An aortic root arose from this leading to a left arch and descending aorta. A separate pulmonary trunk could not be identified but the definition of the roots of the vessels was poor; there was heavy filling of the left lung but poor filling of the right. The general picture suggested the possibility of a truncus arteriosus with a large left pulmonary artery but only small vessels to the right lung.

APPENDIX OF CASES 16–20

Case 16. A boy, aged 6, had a rather centrally placed heart which led at first to his inclusion as complete situs inversus with the heart rotated to the left; but re-examination of his angiocardiograms showed isolated levo-cardia. He was thought to have pulmonary atresia with a bronchial artery supply to the lungs, mainly the right. Diodone from the right arm passed to the right S.V.C. and from here via the coronary sinus to the left side of a common atrium. The left S.V.C. also entered this chamber. The coiling of the catheter right across the heart shadow confirmed a common atrium, all of which filled quickly.

From the left side of the atrium, the diodone passed to the left-sided ventricle and from here to the large aorta with a right-sided arch. Little could be seen of the small right-sided ventricle, and there seemed to be a ventricular septum. No pulmonary trunk was seen and the right pulmonary artery (P.A.) filled from the aorta through large bronchial arteries which produced a continuous murmur. A small left P.A. was found with difficulty at operation: it was anastomosed to the subclavian and he has done well for ten years.

Case 17. A boy, aged 6, was cyanosed and moderately disabled. The S.V.C. and I.V.C. and venous atrium were all left-sided. The catheter passed from this atrium through an A.S.D. to the right-sided (morphological left) atrium and entered a pulmonary vein. Probably from this right-sided atrium, it entered a right-sided ventricle with a pressure of 80/8 mm. Hg (femoral artery pressure 90/65 mm.). The catheter then entered the pulmonary trunk, with a pressure of 14/4 mm. Hg, confirming severe pulmonary stenosis.

Angiocardiography confirmed the findings as regards the atria, and showed a long narrow left-sided ventricle in a normal position: from here the diodone quickly filled the aorta which curved over to the right but had a left-sided arch. In the next film the pulmonary trunk was visible and had almost certainly filled from the right ventricle. In view of the large A.S.D., it was uncertain if there was a V.S.D. also. From their positions and connexions, it seems almost certain that the left-sided and right-sided ventricles were morphologically left and right ventricles respectively.

Case 18. A girl, aged 5 months. Inverted atria, but PI small and upright. The heart was rather centrally placed but had an apex pointing to the left, and a common ventricle.

Necropsy. Left (and only) S.V.C. and I.V.C. to left-sided (morphological right) atrium. Tricuspid atresia with mixing of venous and arterial blood in right-sided (morphological left) atrium. A large, but otherwise normal mitral valve on the right side. Common ventricle with transposition of the great vessels that was not of functional significance. Pulmonary atresia with a large persistent ductus joining the right pulmonary artery. Fully reported by Campbell and MacCarthy (1957).

Case 19. A boy, aged 4 when investigated in 1949, was cyanosed and moderately disabled. Almost every feature was the same as in Case 17, even to the catheter entering a pulmonary vein after passing from the left-sided atrium to the right-sided atrium. The only differences found were (1) that the pulmonary trunk was not entered and he was thought to have pulmonary atresia rather than severe stenosis, and (2)
that the left ventricle was of normal shape, as well as in a normal position, instead of being rather long and narrow.

**Case 20.** A boy, aged 3, was cyanosed from birth and never walked more than 50 yards; he had 8 attacks of unconsciousness with stiffness but no convulsions. Heart rather centrally placed. Catheterization showed that the S.V.C. and the venous atrium were on the left and from this a pulmonary vein was entered: the venous ventricular pressure was at systemic level. Angiocardiography showed the aorta and small pulmonary trunk filling at the same time. When 7, he was still very disabled and cyanotic and operation was decided on. There was a double fall of pressure at the infundibular and at the valve level and the obstructions were dilated. He died with cardiac arrest the next day.

**Necropsy.** Heart weight 290 g. Ventricular septum virtually absent. Aorta arose from right side of common ventricle and arch right-sided. Small pulmonary trunk, 1 cm. in diameter, arose behind it and to the left from common ventricle but through a narrow muscle-bound cleft. No true valve cusps were present and the opening had been moderately dilated. The pulmonary veins (and the S.V.C.) drained into the left-sided atrium.

**REFERENCES**


