LÆVOCARDIA WITH TRANSPOSITION OF THE ABDOMINAL VISCERA

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Received April 30, 1953

The revival of interest in congenital heart disease is reflected by the increasing number of reported cases with transposed abdominal viscera and a normally situated heart.

Incidence. The condition is rare in ordinary practice. After viewing over 250,000 miniature chest radiographs, Nash (1952) cannot remember having seen such a case, and the experience of Morgan (1952), based on 150,000 miniature films and 50,000 full-size radiographs, is similar. They are careful to point out the limitations of this method in discovering the abnormal situation of the abdominal viscera, and this is in accordance with our own experience, that the shadow of gas in the stomach may sometimes be invisible on standard films so that transposition may be apparent in only one of a series of pictures. The rarity of this condition is, however, confirmed by Hills (1952) who cannot remember seeing a single case in the course of more than 20,000 routine barium meals.

We have studied 14 patients under our care and 19 others reported since the subject was reviewed by one of us (Forgacs, 1947) in an attempt to discover whether these defects conform to any common pattern. Our 14 patients were seen among 1130 patients with congenital heart disease, 12 among 670 cyanotic, and 2 among 460 acyanotic patients. In 4 of these the results of necropsies were available; angiocardiology had been carried out in 6 others and cardiac catheterization in 5 of them, but in 4 no special investigations have yet been performed so that the diagnosis must remain very uncertain. The 19 reported are single cases by Del Carril *et al.* (1949), Robinson and Garfinkle (1950), Tanner-Cain and Crump (1951), Case 2 of Hardy (1948), two cases by Thomson (1950), five cases by Doliopoulos and Maillet (1952), and eight cases by Young and Griswold (1951). Case 1 of Hardy is excluded because subsequently he was investigated by us and the findings and necropsy are reported as our Case 9. The two cases of Donzelot *et al.* (1950) were included as Cases 1 and 2 of Doliopoulos and Maillet (1952).*

Of this total of 33 cases, 10 came to necropsy (Tables I and II): among the others cardiac catheterization and angiocardiology were carried out in 9 and angiocardiology alone in 6 (Tables I and III); and in 7 of these and in 1 other the findings at operation were also available. The data are sufficient for analysis of at least some of the features of this uncommon type of heterotaxy.

Associated Defects. When the abdominal viscera are transposed and the heart is on the left, it is nearly always abnormal. There are two or three reported cases where the heart is said to have been normal but Geipel's Case 2 is the only one with necropsy and the only reference to this that we have found (Schelenz, 1909) gives no details. Among the recent cases with necropsies that of Tanner-Cain and Crump (1951) is the only one that makes no mention of any congenital defect. This was a child who was cyanosed at birth and died when three days old: at necropsy "the heart

* Doliopoulos has kindly informed us that in his paper the case references to Donzelot *et al.* (1951) should be to Doliopoulos and Maillet (1952), and that in Case 3 the superior vena cava was on the right and entered the right atrium and the ductus arteriosus was not patent.

was normally placed and extremely dilated with immense thickening of the myocardium and endocardium" but the relationship of the great veins to the atria or of the ventricles to the main arteries is not described.

Generally in these cases there are complicated defects and an accurate diagnosis is notoriously difficult. Our attempt to discover a common pattern has succeeded only partially but they can be divided into two groups. In the first the superior vena cava and the venous atrium follow the abdominal viscera and are transposed, which obviously leads to great difficulty in the development of a heart that can function reasonably well. In the second the superior vena cava and the venous atrium are not transposed and should, therefore, have a normal relationship to the rest of the heart so that the circulation should be normal—a finding that is, in fact, very rare. In the former the P wave in lead I is inverted, while in the latter it is upright (Campbell and Reynolds, 1952). In the former a veno-arterial shunt is left-to-right, and in the latter it is right-to-left as normally. In both groups there are likely to be atrial and ventricular septal defects, sometimes with a single atrium or ventricle, complete or partial transposition of the aorta and pulmonary trunk, and pulmonary stenosis or atresia: there may also be complicated abnormalities of the great veins. We think it important to discuss these groups separately.

CASES WITH AN INVERTED PI AND THE VENOUS ATRIUM ON THE LEFT

Our cases from this group with other reported cases where similar investigations had been carried out are summarized in Table I.

Case 1 (D.P. O405). This boy, aged 3, was cyanosed from infancy and had never crawled. As he was anæmic (hæmoglobin 62%), the obvious cyanosis and clubbing must have indicated a very low oxygen saturation; and he had always been liable to attacks, in which he was almost unconscious, often four or five times a day. He died suddenly at the age of 3 during one of these attacks.

No murmurs were heard and the pulmonary second sound was faint. The heart was large (cardio-thoracic ratio (c.t.r.) 62: m.t.d. 10/16 cm.) with a very broad pedicle (Fig. 1A); it was on the left side but the liver and stomach were transposed. Radioscopy showed oligæmic lung fields and displacement of the aorta forward and to the right. The electrocardiogram (Fig. 2) showed some right ventricular preponderance: PI was small and inverted and PII and PIII were large and upright.

Necropsy. Both the superior and inferior venæ cavæ were on the left and entered the left atrium. There was only a small slit patency of the atrial septum. The left atrium communicated through a

FIG. 1.—Teleradiograms from three cases with the venous atrium on the left. (A) Case 1, with a large heart and a broad pedicle. (B) Case 2, with a rather small heart. (C) Case 3, with a normal-sized heart. The aortic arch is right-sided in all these three, and this is nearly always so when the venous atrium is on the left. A barium swallow showed that the stomach was on the right in all these patients and the air bubble can be seen in (A) and (C).

FIG. 2.—Case 1. Electrocardiogram showing inverted PI and less evidence of right ventricular preponderance than is usual. Standardization 1 mV.=0.5 cm. in chest leads, V4R to V7.

tricuspid valve with the larger ventricle that was placed posteriorly and on the left side. The outflow tract ran to the right to a high ventricular septal defect and the pulmonary trunk over-rode this.

The right atrium was small and posteriorly placed; it received the pulmonary veins and communicated through a *bicuspid valve* with the smaller ventricle on the right. The aorta arose from the extreme right of this ventricle and formed a right-sided arch. The pulmonary trunk arose behind and to the left of the aorta and over-rode the septal defect; the pulmonary valve was *bicuspid* and stenosed.

The lungs were transposed like the other viscera, the left having three lobes and weighing more than the right. There was *situs inversus* of all the abdominal viscera.

Case 2 (K.L. O428). This girl, aged 19, was cyanosed from the age of six weeks, and did not walk until she was 4 years old. She had always been breathless and, though she could walk a mile, had to stop every 50 yards for a rest. She did not squat. She had some degree of *arachnodactyly*.

Cyanosis and clubbing of the fingers were severe. A soft systolic murmur was loudest in the fourth left intercostal space close to the sternum, and the pulmonary second sound was normal. The heart was not enlarged (c.t.r. 46: m.t.d. 11.0/24 cm., Fig. 1B). Radioscopy confirmed the reversed position of the stomach and liver, oligæmic lungs, and a right-sided aortic arch. The cardiogram (Fig. 3) showed an inverted PI and S waves only from V1 to V5 so there may have been some slight hypertrophy of both ventricles. The red cell count was 8.7 million and hæmoglobin 137 per cent.

Catheterization showed that the superior and inferior *venæ cavæ* drained into the left atrium (O_2 saturations 61–67%). This communicated through a sizeable septal defect with the right atrium which received the pulmonary veins. From it the catheter entered the right (mixed arterial) ventricle where the systolic pressure was 120 mm. Hg and the arterial O_2 saturation 84 per cent compared with 75 per cent in the systemic arteries: it could not be passed into any exit from this or into the left (venous) ventricle.

Angiocardiography confirmed filling of the left atrium from a left-sided superior vena cava (Fig. 13A). Four seconds after the injection the aorta was opaque and at five seconds the pulmonary arteries were outlined.

FIG. 3.—Case 2. Electrocardiogram showing inverted PI and no certain right ventricular preponderance, as there is low voltage in the standard and unipolar limb leads, large S waves only, from V1 to V5, and smaller S but quite insignificant R waves from V5R to V3R. Standardization 1 mV.=1.0 cm. in all leads.

A complete diagnosis cannot be made. The atria are reversed and there is a large atrial septal defect. Pulmonary stenosis is supported by the oligæmic lungs. There is a free shunt from the venous side to the aorta but it is not clear whether it arises from the left (venous) ventricle or overrides this, or even from a right ventricle that is partly venous from a left-to-right shunt through the atrial septal defect. Two years later Mr. R. C. Brock was unable to find any pulmonary artery on the left side for a subclavian-pulmonary anastomosis. Attempts were therefore made to improve the collateral circulation over the pleura and six months later she seems to be somewhat improved.

Case 3 (L.H., P105). This boy, aged 14, was cyanosed from birth. When seen, he was attending an ordinary school but could only walk a mile on the level and was not playing any games.

He was small, deeply cyanosed, and had severe clubbing of the fingers. A harsh systolic murmur was widely conducted and was heard best in the third left intercostal space close to the sternum; the second sound was accentuated. The blood pressure was 110/80. The heart was normally situated and was not enlarged (c.t.r. 51: m.t.d. 12.5/24.6 cm. Fig 1C), the stomach was under the right costal margin, and the liver was palpable on the left side. The cardiogram (Fig. 4) showed right ventricular preponderance, PI was inverted (except in one record) and PII and PIII were upright. The hæmoglobin was 110 per cent and the red blood cells were 6.4 million.

Cardiac catheterization from the left arm proved that the superior and inferior venæ cavæ were on the left and drained into the left atrium. From here it entered a venous left ventricle but could not be advanced far (Fig. 5A). The left atrium also communicated through a large septal defect with the right atrium which received one and probably all the pulmonary veins (Fig. 5B). The catheter passed from the right atrium into the right ventricle both of which contained blood of a much higher oxygen saturation (78–80%) than the left side of the heart (66–68%). Neither the aorta nor the pulmonary trunk could be entered, but judging by the O₂ saturation of the aorta which was 78 per cent, it came mainly from the right ventricle, which had the same systolic pressure as the systemic arteries.

The angiogram confirmed that the left atrium filled from a left superior vena cava but was at a rather high level for a venous atrium (Fig. 13B). There was a large communication between this and the right atrium, so that the heart seemed generally filled at 3 seconds. The aorta, situated well to the right and with a right-sided arch, became opaque at 4 seconds and the pulmonary arteries

FIG. 4.—Case 3. Electrocardiogram showing inverted PI and the more usual evidence of right ventricular preponderance. Standardization 1 mV. = 1.0 cm. in all leads.

FIG. 5.—Catheter demonstration of the venous atrium on the left side. (A) The catheter has passed down the left S.V.C. to the left atrium, and to the left (venous) ventricle. (B) The catheter has passed through an atrial septal defect to the right atrium and thence into a pulmonary vein. Case 3.

were opaque between 2–4 seconds. The pulmonary trunk was not seen, but the pulmonary blood flow was thought to be little, if at all, diminished.

The diagnosis is incomplete. The arterial and venous sides of the heart are reversed; there is a large atrial septal defect; and the aorta is thought to arise from the right ventricle. He is now 17 and has kept well during the last three years.

Case 4 (R.W. O556). This girl, aged 7, was cyanosed from birth and until the age of 5 years could not walk more than 50 yards and used to squat. She then improved and, when seen at the age of 7, could walk a quarter of a mile. She was mentally backward and moderately cyanosed with severe clubbing of the fingers and toes. There was a prominent venous pulse in the neck.

TABLE I
CASES WITH THE VENOUS

Case	Age and sex	Necropsy N Angioc. A Catheter C Operation O	Inf. V.C.	Sup. ven. cav.		Pulm. veins	A.S.D.	V.S.D.	Aortic arch
				R.	L.				
Our Case 1	3M	N	LA	Abs.	LA	RA	Slit P.F.O.	++	R
Young and Griswold, Case 1	2F	NO	LA	Abs.	LA	RA	P.F.O.	++	R
Our Case 2	19F	ACO	LA	—	LA	RA	+++	?	R
Our Case 3	14M	AC	LA	—	LA	?RA	++	?	R
Our Case 4	7F	ACO	LA	—	LA	RA	++	?	R
Our Case 5	4F	A	?LA	?	LA	?	++	?	R
Doliopoulos and Maillet (1952), Case 4	6F	A	?LA	?	LA	?	?	?	L

* Neither aorta nor pulmonary trunk were entered on cardiac catheterization.

† Generally inverted but sometimes upright with PII and PIII deeply inverted.

A harsh systolic murmur and a loud single second sound, which was probably aortic, were heard in the pulmonary area. The heart was normally situated and large (c.t.r. 65, m.t.d. 12·8/19·4 cm., Fig. 7A) with a broad pedicle: on screening both ventricles seemed large and the aorta arose well to the left and anteriorly; it was broad and unfolded and passed nearly horizontally to the right, forming a right-sided aortic arch. The lungs were oligæmic. The reversed situation of the liver and stomach was confirmed. The cardiogram showed right ventricular preponderance and the P waves were generally inverted in lead I but sometimes there was a change of pacemaker and then they were upright in lead I and inverted in leads II and III. The hæmoglobin was 136–160 per cent.

Catheterization from the left arm showed that the superior and inferior venæ cavæ entered a left venous atrium (Fig. 6A). From it the catheter passed directly into a venous ventricle on the left and thence into the aorta: the oxygen saturation at these three sites was much the same (77%, 77%, 80%). The right atrium (O₂ saturation 89%) and one of the pulmonary veins were entered through an atrial septal defect (Fig. 6E). The pressure in the left (venous) atrium (19/–4) was a little higher than in the right (arterial) atrium (14/–2 mm. Hg). Neither the second ventricle nor the pulmonary trunk could be entered.

Angiocardiography, from the right arm, confirmed that the left superior vena cava drained into the left atrium (Fig. 13C). Diodone passed at once to the right atrium through a large septal defect, and after 3 seconds the large ventricular cavity that extended to the apex and the arch of the aorta was opaque. The pulmonary arteries began filling at 4 seconds and were well seen at 5 seconds, though it was not clear how they arose.

At thoracotomy, Mr. Holmes Sellors thought there was a common arterial trunk, possibly with the right pulmonary artery arising from it: a right subclavian-pulmonary anastomosis was made.

ATRIUM ON THE LEFT SIDE

Origin of aorta	Origin of pulm. trunk	Pulm. orifice	Lungs	EC	PI	Remarks
From R of RV	Behind aorta from RV; over-riding LV	PVS	Oligæmic	Some RVP	Inv.	RA, RV arterial. LA, LV venous. Aorta from RV but PT stenosed and over-riding
From RV mainly but over-riding LV	Fibrous cord from LV	Atresia	Oligæmic	RVP	No EC	RA, RV arterial. LA, LV venous. Pulm. atresia and PDA
Receives venous blood *	??	?PS	Oligæmic	S only V1 to V6	Inv.	RA arterial. LA venous. Large ASD. ?PS
From RV mainly. Receives venous blood *	??	?	?Normal	RVP	Inv.	RA, RV arterial. LA, LV venous. Large ASD
Partly from LV (passage of catheter)	?	?PS	Oligæmic	RVP	Inv.†	RA, RV arterial. LA, LV venous. Aorta from LV
Receives venous blood	?	?	Oligæmic	LPV	Inv.	?RA, arterial. LA venous. ?Tricuspid atresia. ??Dextrocardia
From R. side. Receives venous blood	?	?	?Normal	RVP	Inv.	?

Six months after the operation her capacity and colour were better and after two years there was further improvement.

In this case the reversal of the arterial and venous atria was confirmed by catheterization and angiocardiology. There was a large atrial septal defect, and the lungs were oligæmic.

Case 5 (S.M., O266). This girl was cyanosed from birth and was first seen at the age of 7 months. During the first few months of life she had several attacks in which she became stiff and panted for breath, but these subsequently ceased. When 4 years old she was still unable to walk. She had intense cyanosis with severe clubbing of the fingers.

A systolic murmur was heard, loudest towards the apex, and a slight thrill was felt. The pulmonary second sound was normal. The heart was normally situated but rather central: it was not generally enlarged (c.t.r. 46, m.t.d. 7.6/16.5 cm., Fig. 7B) but on screening the left ventricle was enlarged, and the lungs looked oligæmic. The stomach was on the right but the liver seemed to be, at least partly, on the right also. The cardiogram showed an inverted PI and left ventricular preponderance.

Angiocardiology showed that the superior and inferior venæ cavæ and the venous atrium were all on the left (Fig. 13D). Diodone passed from the left atrium to the right and the aorta filled quickly. It was so much on the right that a right-sided arch was diagnosed and, although it curved to the left, it passed downwards on the right side, at least as far as the level of the diaphragm. The pulmonary trunk was not seen but the left pulmonary artery was filling at 1½ seconds.

The clinical diagnosis is tricuspid atresia, though pulmonary stenosis with an unusual degree of rotation of the heart is not excluded. Although the heart is thought to be on the left it is possible that there is really dextrocardia with the heart rotated back to the left side.

FIG. 6.—Catheter demonstration of the venous atrium on the left in Case 4. (A) Catheter passing down the left S.V.C. through the left atrium into the I.V.C. which also entered the left atrium. (B) Catheter passing from the left atrium to the left ventricle. (C) Catheter passing on from the left ventricle to the aorta and round the right-sided arch into the descending aorta which lies on the left below the diaphragm. (D) and (E) Catheter passing from the left to the right atrium through an atrial septal defect and outlining the border of this chamber (D) and entering a pulmonary vein (E). (F) Catheter passing from the left atrium, possibly via the right atrium, to the hepatic veins shown up by the injection of diodone. The hepatic veins may enter a different atrium from the inferior vena cava or the left atrium may extend more to the right in its lower part.

FIG. 7.—Teleradiograms from three cases with the venous atrium on the left. The aortic arch was right-sided in all three. (A) Case 4, with a rather large heart and broad pedicle. (B) Case 5, with a rather small heart and probably with tricuspid atresia. (C) Case 6, with some enlargement of the heart and left ventricular preponderance: she was one of the rare acyanotic patients and was thought to have congenital aortic stenosis.

OTHER CASES WITH P INVERTED IN LEAD I

One other case of our own and Case 1 of Thompson (1950) belong to this group because of the inverted PI, but they are not included in the discussion or in Table I, because no special investigations had been carried out.

Case 6 (B.T., O851). This girl, aged 4, looked healthy and had no disability, but congenital heart disease had been diagnosed when she was 3 weeks old. When 4 years old there was no cyanosis nor clubbing of the fingers. A thrill and systolic murmur were found, maximal in the right second intercostal space, and the second sound was normal. Her blood pressure was 100/70. The heart was normally situated and enlarged (c.t.r. 56: m.t.d. 9/16 cm.), mainly the left ventricle (Fig. 7C). The aortic arch was right-sided. The blood supply to the lungs was normal. The cardiogram showed left ventricular preponderance and the P wave was generally inverted in lead I, but once it was upright, probably with a change of pacemaker. The liver was palpable under the left costal margin, and on screening the stomach was seen under the right diaphragm. The provisional diagnosis was congenital aortic stenosis.

CASES WITH AN UPRIGHT PI AND WITH THE VENOUS ATRIUM ON THE RIGHT AS NORMALLY (TABLES II AND III)

Case 7 has been fully reported (Campbell *et al.*, 1952). A frail little boy, 5 years old at the time of his death, he had been cyanosed from birth and could not walk much more than 25 yards. The heart was large (c.t.r. 57) and there was a continuous murmur high up on the left side. The cardiogram showed an upright PI, inverted PII and PIII, and evidence of right ventricular hypertrophy. He died after a subclavian-pulmonary anastomosis.

Necropsy. The heart was bilocular. There were gross abnormalities of the veins, as the drainage from the lower half of the body was through the azygos vein to the right superior vena cava (Fig. 14A). A left superior vena cava was also present and passed through a greatly dilated coronary sinus to enter the right side of the common atrium. The aortic arch was left-sided.

Case 8 has been fully reported (Case 3 of Campbell *et al.*, 1953). She was 4 years old and had been severely cyanosed from birth. She could only walk 100 yards, but did not squat. There was a harsh systolic murmur and a faint thrill in the third left space. The heart was enlarged (c.t.r. 61) and sabot-shaped with a deep pulmonary bay. The aortic arch was left-sided. The cardiogram supported right ventricular preponderance and PI was upright. The hæmoglobin was 138 per cent. Angiocardiography showed that the superior vena cava and the venous atrium were on the right (Fig. 14B). She died after a subclavian-pulmonary anastomosis.

Necropsy. The heart lay normally on the left side. The systemic veins drained normally into a right atrium which opened by a tricuspid valve into a single ventricle. The pulmonary trunk arose to the left of the aorta, and there was pulmonary valvular stenosis. A single pulmonary vein from both lungs drained into the left atrium, from which the only exit was a slit-like opening in the atrial septum to the right atrium and thence to the common ventricle. The right lung had four lobes and the left had three. The stomach was on the right side and the gall bladder was on the left, but the liver lay on both sides.

Case 9 (A.M., O146). Cyanosis was first noticed when he was 6 months old. On a warm day he could sometimes walk 50 yards but in cold weather he could not get about at all: he squatted sometimes but not regularly. He was first seen when he was one year old and his condition was unchanged for the next three years, but became worse during his fifth year.

He was deeply cyanosed and had severe clubbing of the fingers. A widely conducted systolic murmur, followed by a single, loud and palpable second sound was heard in the pulmonary area. There was also systolic pulsation here, but after radioscopy this was rightly thought to be due to the aorta and not the pulmonary trunk. Screening showed a small, normally situated heart (c.t.r. 48: m.t.d. 7.3/15.2 cm., Fig. 8A) and an aorta that arose anteriorly and had a right-sided arch. The lungs were oligæmic and the pulmonary arteries were small.

The cardiogram with R and S waves of equal size in V4R and in V1 and with S waves only in V6 was thought to indicate some right ventricular preponderance, and the P waves were upright in the standard leads (Fig. 9). The diagnosis was pulmonary atresia.

FIG. 8.—Teleradiograms from three cases with the venous atrium on the right. Those for Cases 7 and 8 have already been published. (A) Case 9, with a small central heart which was only slightly inclined to the left: the bilateral superior venae cavae do not show on the X-ray film. (B) Case 10 with a rather large heart and a broad pedicle that was partly due to bilateral superior venae cavae, both draining into the right atrium. (C) Case 11, with a rather large heart, a broad pedicle, and an unexplained indefinite shadow on the left. The aortic arch was right-sided in all these though it may be on either side when the venous atrium is on the right. All these three had bilateral superior venae cavae. The gas bubble in the stomach is well seen on the right in (A) and (C) and this was confirmed by barium in (B) though the liver was thought to be on both sides in this case.

FIG. 9.—Case 9. Electrocardiogram showing upright PI. The standard leads show the sSS pattern and the chest leads show mainly S waves though R is about the same size as S from V4R to V1 and this, perhaps, indicates some right ventricular preponderance. Standardization 1 mV.=0.5 cm. in chest leads V4R to V6.

Angiocardiography was abandoned because of the severe fall of blood pressure when it was started. As he was getting worse, thoracotomy was decided on. The left superior vena cava seemed to empty into the left atrium, and the right superior vena cava was not seen, although in fact it was present. The left superior pulmonary vein seemed to pass behind the left atrium to join the right atrium so that the atria and venous system were thought to be transposed as the abdominal viscera were, and the apex of the heart pointed almost completely downwards. No pulmonary artery could be found for an anastomosis. The

findings at operation can not, therefore, give the detailed anatomy of these complex hearts. He had a stormy convalescence, including an attack of hemiplegia that was diagnosed as cortical thrombo-phlebitis, but recovered enough to go home and died rather suddenly three months later.

Necropsy. The heart was very central but just pointed to the left (Fig. 10). There were bilateral superior venæ cavæ and the left was joined by an azygos vein. The inferior vena cava entered the atrium below on the left and only a large hepatic vein entered on the right. Some hepatic veins also entered the inferior vena cava on the left. The atrium was large and single: above the ridge (see Fig. 11B) there was a primitive septum but below this the two sides communicated freely so that there was a single atrium. The two superior venæ cavæ entered the single atrium on either side of the primitive septum almost symmetrically but the right a little more obliquely and anteriorly: the left entered nearer to the entrance of the pulmonary veins (Fig. 11) and not through the coronary sinus. For some time no coronary sinus was found, but in the left side of the atrium some way below the entry of the left superior vena cava (B in Fig. 11B) there was a small opening, well-covered by a valve, leading to a vein that ran in the heart wall. The position suggested that it might have been a coronary sinus and, if so, it implies that the primitive venous system became obliterated in its middle part leaving the left superior vena cava patent above and a coronary sinus draining into the left side of the atrium below. The direction of flow through this vessel was, however, unusual for a coronary sinus.

There was a single bicuspid A-V valve with its longer axis lying transversely. There was also a single ventricle though there was a ridge that may have represented the septum. The only exit from the ventricle was through a rudimentary outflow chamber (Fig. 10) lying anterior and to the left and this could only be reached by the blood passing at the side of or through the fenestrations of the cusp of the A-V valve. At first no pulmonary trunk could be found because it was hidden directly behind the aorta, i.e. transposed. There was a moderately severe pulmonary valvular stenosis and some infundibular stenosis.

The pulmonary veins joined to form one from each lung and entered the single atrium above and on the left side, and the oxygenated blood passed through the single A-V valve to the common ventricle. The aorta and pulmonary trunk both arose from the rudimentary outflow chamber; the former was centrally placed and ascended vertically to arch to the right and the descending and abdominal aorta were also on the right.

Both lungs had three lobes. The stomach was on the right and the duodenum was also abnormal as it passed between the neck of a very large gall bladder and the liver. The liver appeared to be almost equally on both sides but the gall bladder was just to the right of the mid-line. No spleen was found. There was ante-mortem thrombosis of the right renal vein. The brain contained a fairly large area of softening in the left external capsule. This seemed to have followed from thrombosis and had been present for some months, possibly but not certainly before his operation. There was a much smaller area of softening in the right frontal pole that had probably been present only for days.

Case 10 (R.W., H126). This girl, aged 8, had been cyanosed from birth and was only able to walk a mile slowly. She had not squatted. There was moderate clubbing of the fingers.

There was a systolic murmur and a thrill in the third and fourth spaces in the left. There was no diastolic murmur, the pulmonary second sound was loud normal, and the blood pressure was 95/70. There was striking pulsation of the veins of the neck. Her heart was moderately enlarged (c.t.r. 61: m.t.d. 11/18 cm.) and was on the left (Fig. 8B); and later it was realized that the abdominal viscera were transposed. On screening the pedicle was very broad and both ventricles seemed large: the pulmonary arteries were easily seen and the lungs did not look oligæmic. The aortic arch was high and right-sided. The cardiogram showed right ventricular preponderance without right axis deviation and PI was large, broad, and upright. Her hæmoglobin was 122 per cent, the red cells 7.0 million, and the hæmatocrit 65 per cent.

Four years later, her condition and the physical signs were much the same. Although the liver seemed palpable on the right side, X-rays confirmed that the stomach was on the right and that the gall bladder was on the left so probably the liver was on both sides.

Fig. 10 a.

Fig. 10 b.

FIG. 10.—The heart of Case 9 from the front with the rudimentary outflow tract on the left and the central aorta opened. The small pulmonary trunk with a valvular and infundibular stenosis arose behind this. (A) Photograph of the heart. (B) Drawing of the outflow chamber. The aorta, which had three cusps, is seen opened. The small pulmonary trunk arose behind this, in the cleft marked A. The only way from the main cavity of the ventricle to this chamber was by the side of (marked B) and through the fenestrations of the anterior cusp of the A-V valve.

Fig. 10b

Fig. 11 b.

FIG. 11.—The heart of Case 9 from behind to show the single atrium. The hila of the lungs have been turned upwards to display the pulmonary veins. These unite to form one vein from each lung both of which then enter the single atrium on the left side. Above the ridge marked A in the drawing there was a primitive septum but below this the two sides were in free communication with each other and joined the single ventricle through the single A-V valve. (A) Photograph of the heart. The crossed probes marked P are in the two pulmonary veins. The more vertical probes are in the two superior venæ cavæ. (B) Drawing of the heart. Probes have been passed down the right and left superior venæ cavæ and pass respectively into the right and left sides of the common atrium.

Angiocardiography showed bilateral superior venæ cavæ and both entered the venous atrium on the right (Fig. 14C). Early opacification of the left atrium suggested an atrial septal defect. The aorta filled early at 2 sec. before the left ventricle and the pulmonary arteries filled slowly without much change in the density of the lungs.

On catheterization the pressure in the right ventricle (90/0 mg. Hg), and probably in the left also, was the same as in the systemic arteries. The pressure in the pulmonary trunk was much lower (4 mm.), confirming pulmonary stenosis. The O₂ saturations were difficult to understand: in the right atrium and ventricle (75–80%) they were higher than in the superior vena cava (66%) left ventricle? (68%), and brachial artery (64%), suggesting that a pulmonary vein entered the right atrium or that there was a left-to-right shunt through the atrial septal defect rather than the right-to-left shunt seen on angiocardiography. Only a partial diagnosis is possible. There are arterio-venous and veno-arterial shunts; the aorta over-rides or rises from the right ventricle; and there is also pulmonary stenosis, though at one time Eisenmenger's complex was suspected.

Case 11 (H.R., O116). This boy was under observation for five years. At the age of 3 years he only walked 100 yards: he had a high colour but no convincing cyanosis, though it had first been noticed at 3 months. When 5, there was a little cyanosis at rest and the fingers and toes showed early clubbing: he was able to attend an ordinary school. When 8, he had improved further and could walk a mile at his own speed and cycled 6 miles to and from school.

There was a loud, widely-conducted systolic murmur and thrill in the pulmonary area. The pulmonary second sound was always loud and became louder, a short diastolic murmur was sometimes heard towards the apex, and the blood pressure was 90/60. The heart was on the left (Fig. 8C) and both ventricles seemed large (c.t.r. 60, 10·8/18·0 in 1948; 57, 12·6/22 in 1953): the aortic arch was right-sided. Pulsation was seen in the pulmonary arteries and at first beyond these. The pedicle was broad and there was an ill-defined shadow above the heart on the left gradually fading upwards, so that drainage of some pulmonary veins into a superior vena cava (Snellen and Albers, 1952; Gardner and Oram, 1953) was considered but seemed unlikely, and catheterization was unsuccessful. The cardiogram showed right ventricular preponderance with rather wide notched QRS complexes (0·10 sec.); PI was large and upright, PII inverted, and PIII generally inverted, but not in this record (Fig. 12). The hæmoglobin was 128 per cent and the red cell count 7·0 million with a hæmatocrit of 59 per cent. The stomach was under the right diaphragm and the liver was on the left.

FIG. 12.—Case 11. Electrocardiogram showing upright PI and the more usual signs of right ventricular preponderance. Standardization 1 mV=1·0 cm. in all leads.

TABLE I
CASES WITH THE VENOUS ATRIUM ON THE RIGHT

Case	Age and sex	Necropsy N Angico. A Catheter C Operation O	Inf. V.C.	Sup. ven. cav.		Pulm. veins	A.S.D.	V.S.D.	Aortic arch
				R.	L.				
Our Case 7	5M	NACO	Absent. Azygos to R. S.V.C.	R. side of single atrium	R. side through coron. sinus —	Single atrium	++++*	++++*	L
Our Case 8	4F	NAO	RA	RA	—	LA	Slit-like PFO	++++*	L
Our Case 9	5M	NO	Left of single atrium	R. of single atrium	L. of single atrium	L. of single atrium	++++*	++++*	R
Doliopoulos and Maillet (1952) Case, 1	4F	NA	RA	RA	—	LA	+++	++++*	R
Doliopoulos and Maillet (1952), Case 3	1F	N	LA	RA	—	LA	++	++	L
Young and Griswold (1951) Case 2	10M	NAO	R. side of single atrium	R. side of single atrium	To R. S.V.C. by innominate Abs.	To L. S.V.C.	++++*	++	R
Robinson and Garfinkle (1950)	6daysM	N	RA	RA	—	LA	+	++	?L
Tanner-Cain and Crump (1951)	3daysF	N	?	?	?	?	No	No	?

* +++++ denotes such a large defect that it was a single chamber.

Angiocardiography showed a superior vena cava and a venous atrium on the right (Fig. 14D) and suggested that a pulmonary vein entered the right atrium. The diiodone passed from the right to the left atrium so that all the heart was quickly opacified. The aorta was large and arched to the right; it arose well over to the left but it was difficult to say from which ventricle. The pulmonary arteries filled slowly—less well than was expected from radioscropy—and too little blood passed through the lungs to opacify the left atrium again or the unusual shadow to the left of the aorta.

He was thought to have an atrial septal defect, at first with a shunt that was mainly left-to-right; cyanosis then appeared at rest and pulsation in the pulmonary arteries decreased as the shunt became reversed. The increasingly loud pulmonary second sound suggests that a rise of the pulmonary blood pressure was responsible for this.

OTHER CASES WITH P UPRIGHT IN LEAD I

Three of our cases and four others—Case 2 of Hardy (1948), Case 2 of Thompson (1950), Case 5 of Doliopoulos and Maillet (1952), and probably Case 8 of Young and Griswold (1951)—belong to this group because of the upright PI. They are not included in Table III or used in the discussion because special investigations were not made or were too incomplete. Short notes of our three cases follow.

Case 12 (R.T., O014). He was seen when a year old. He was never able to walk and was cyanosed from birth, with early clubbing of the fingers. From 2 years onwards he had several attacks of intense dyspnoea, prostration, pallor, and coughing, and he died suddenly at the age of 3 in one of these attacks.

A soft systolic murmur was heard in the pulmonary area and the second sound was normal. The heart was large and sabot-shaped (c.t.r. 60: 9.7/16 cm.) and the lungs looked oligæmic with bronchial arteries carrying the main blood supply. The aortic arch was probably right-sided. The cardiogram showed right

SIDE AS NORMALLY (ALL WITH NECROPSIES)

Origin of aorta	Origin of pulm. trunk	Pulm. orifice	Lungs	PI	EC	Remarks
From R. of single V.	Dimple just L. of aorta	Pulm. atresia	Oligæmic	Upr.†	RVP	Bilocular heart. Pulm. atresia. Lungs supplied through PDA
From R. of single V.	L. of aorta	PVS	Oligæmic	Upr.	RVP	Mitral atresia. Blood reached single ventricle through PFO and RA.
Centrally from single V.	Posterior to aorta	PVS	Oligæmic	Upr.	Some RVP	Heart almost bilocular and very central
Over-riding RV. Almost single V.	L. of aorta	Pulm.	Oligæmic	Upr.	RVP	Single ventricle. Large ASD. Pulmonary atresia. PDA.
From RV.	From RV posterior to aorta	?Normal	?Normal ?Oligæmic	Flat	RAD	ASD, VSD and aorta from RV with or without pulm. stenosis
Over-riding, mainly RV	From LV	Pulm. stenosis	Oligæmic	—	RVP	Single atrium. Anomalous pulm. veins. Pulm. stenosis
From RV	From LV	Normal	?Congested	—	No EC	ASD, VSD, PDA, Transposition
?From LV	?From RV	Normal	?	?	?	

† Inverted PII and PIII. This patient had duodenal atresia.

ventricular preponderance with an upright PI. The hæmoglobin was 110 per cent, the red cell count 7·3 million. The gastric air bubble was under the right diaphragm, but the liver was probably on both sides.

A tentative diagnosis of pulmonary atresia with bronchial artery blood supply to the lungs was made. There was no necropsy.

Case 13 (J.L., O823). She was cyanosed from birth and severely so with marked clubbing of the fingers when she was seen at the age of 4 years. Her physique was very poor and she was unable to stand.

A trivial systolic murmur in the pulmonary area was followed by a double second sound, and there was a triple rhythm at the apex. The heart was large (c.t.r. 65: m.t.d. 10·1/15·5 cm.), the vascular pedicle was very broad, and the aortic arch was right-sided. The lungs were oligæmic. The stomach was seen under the right diaphragm, but the liver may have been partly on both sides. The cardiogram showed right ventricular preponderance, and the P waves were upright in lead I and deeply inverted in leads II and III. The hæmoglobin was 120 per cent.

The findings, with the exception of the double second sound, were consistent with pulmonary atresia. She died a year later in a fit while her mother was bathing her, having got much worse after measles some months before and having other convulsive fits in the weeks before her death: there was no necropsy.

Case 14 (N.R., P222). A fit man, now aged 32, was the Case 1 of Forgacs (1947). We need only add that he has got on well during the subsequent six years and his heart is no larger (c.t.r. 54: m.t.d. 15·7/29 cm.). He has put on too much weight and is breathless playing tennis singles or running upstairs but not otherwise. He has consistently shown left ventricular hypertrophy on radiology and cardiography and the average of many readings of his blood pressure is 145/75. A faint aortic diastolic murmur has been heard at some visits but not regularly. The aortic arch is left-sided. He has always shown an upright PI and inverted PII and PIII.

We do not now accept the explanation that was then suggested. He probably has aortic incompetence and this may be due to bicuspid aortic valves. This, the most natural explanation, was rejected in 1946 because no aortic diastolic murmur had then been heard.

TABLE I
CASES WITH THE VENOUS ATRIUM ON THE LEFT

Case	Age and sex	Necropsy N Angioc. A Catheter C Operation O	Inf. V.C.	Sup. ven. cav.		Pulm. veins	A.S.D.	V.S.D.	Aortic arch
				R.	L.				
Our Case 10	8F	AC	?RA	RA	RA through cor. sinus	?	++	?+	R
Our Case 11	8M	A(C)	?RA	RA	—	?	++	?	R
Doliopoulos and Maillet (1952), Case 2	3F	A	?RA	RA	?Abs.	?LA	?	++	L
Young and Griswold, Case 3	15m.F	AO	?RA	Abs.	RA through cor. sinus	?	++	?+*	L
Young and Griswold, Case 4	4M	ACO	?	RA	LA	LA but one PV to Rt. SVC	+	?++	R
Young and Griswold, Case 5	8M	ACO	To L. S.V.C.†	?Abs.	RA through cor. sinus	LA	++	?+*	R
Young and Griswold, Case 6	9M	ACO	?RA	RA	RA through cor. sinus §	?RA†	+†	+*	L
Young and Griswold, Case 7	6F	ACO	?RA	RA	?LA ?RA through cor. sinus	?RA†	+†	?*	R
Del Carril <i>et al.</i> (1949)	21m.F	AC	RA	RA	RA through cor. sinus	?	+	?	R

* Only evidence is that aorta was thought to be over-riding.

† At least one pulm. vein to RA or left-to-right shunt through A.S.D.

‡ I.V.C. probably absent with large azygos vein to left S.V.C.

§ Personal communication from Dr. Taussig. Single atrium not excluded in this or in Young and Griswold, Case 3.

DISCUSSION OF GROUP WITH THE VENOUS ATRIUM ON THE LEFT

Five of our cases belong to this group but very few of the others reported so that there are only seven in all and only two with necropsies (see Table I). These seven have certain features in common: the superior and inferior venæ cavæ are on the left, and so far as we know on this side only, and enter a venous atrium on the left; the pulmonary veins enter the right atrium; and the aortic arch is right-sided. If the structure of the heart were otherwise normal and the aorta arose from the left ventricle, the venous blood would be returned to the systemic circulation and the patient could not survive. If the aorta and pulmonary trunk were transposed without any other abnormalities the circulation would be functionally normal.

In our Case 1 where the anatomical facts are known fully, the systemic veins entered a left (venous) atrium which had the general appearance and shape of a normal right atrium, and joined a left ventricle through a valve that had three cusps. The pulmonary veins entered a right (arterial) atrium, which had the general appearance and shape of a normal left atrium, and communicated with a right (arterial) ventricle through a valve that had two cusps. The aorta arose from this and not from the left ventricle. If the pulmonary trunk had arisen directly from the left ventricle the circulation, being a mirror image of the normal, would have been functionally normal, but there was pulmonary stenosis and a large ventricular septal defect and the pulmonary trunk over-rode this. There must also have been a large left-to-right (veno-arterial) shunt through the ventricular septal defect to the aorta to account for the severe anoxæmia: presumably the pressure was higher on the left side because of the pulmonary stenosis.

The only other case with a necropsy (Young and Griswold, Case 1) agrees fully with this in

LIGHT SIDE AS NORMALLY (NO NECROPSIES)

Origin of aorta	Origin of pulm. trunk	Pulm. orifice	Lungs	EC	PI	Remarks
From RV	??LV	Pulm. stenosis	?	Some RVP	Upr.	Pulmonary stenosis
?	?	?Pulm. stenosis	?Pleonaemic and later oligæmic	RVP	Upr.	ASD and ?pulmonary hypertension
Over-riding RV	?	Slight P.S.	?Oligæmic	RVP	Upr.	VSD and over-riding aorta with slight pulm. st.
Over-riding RV	?	?P.S.	Oligæmic	RAD	Inv. and Upr.	ASD. ?Fallot Died 6/12 after operation
Over-riding RV	?RV	?P.S.	Oligæmic	RAD	Inv. and Upr.	?Fallot Anomalous pulm. veins
Over-riding RV	?	?P.S.	Oligæmic	LVP	Upr.	ASD. ?Fallot
Over-riding RV	?Posterior to aorta	?P.S.	Oligæmic	RAD	—	?Fallot
Over-riding RV	RV	P.S.	Oligæmic	RAD	Inv. or biphasic	Pulmonary stenosis
?	?	?	?	RVP	Upr.	Diagnosis uncertain

the structure of the veins, atria, and ventricles but there was pulmonary atresia instead of stenosis and the aorta was mainly, instead of entirely, from the right ventricle. Unfortunately, the report does not say which A-V valve had two cusps and which had three cusps and the case of Robinson and Garfinkle (1950) (see Table II) does not help because both A-V valves had two cusps.

In the other five the arrangement of the systemic veins and atria was the same as in the two with necropsies (see Fig. 13). The aorta received venous blood, but it was not certain that it arose from or over-rode the right ventricle because there was a large left-to-right (veno-arterial) shunt through an atrial septal defect: this means that the pressure must have been higher on the left (venous) side, probably because of pulmonary stenosis.

In Case 4, who was very cyanotic and disabled, the aorta seemed to arise mainly from the left (venous) ventricle (see figures for O₂ percentage) and the catheter took this course, and in Case 5

FIG. 13.—Diagrams of the angiocardigrams from four cases where the venous atrium is on the left. These are taken from tracings of several angiocardigrams, so that the part filling first is shown in dark black and in some cases the spread beyond by dotted black. The aorta is shown from later records by hatched lines. In (B) and (C) the left atrium seems more in the normal position of the left atrium and in (A) and (D) more in the mirror-image position of a right atrium.

the angiocardigram favoured an aorta from the left ventricle; but in Case 3 who was less disabled the figures for the O₂ saturation suggested that the aorta arose from the right (arterial) ventricle, and in Case 2 it was uncertain. In all these four there was an atrial septal defect and this was probably the site of the left-to-right (veno-arterial) shunt. In several of these cases transposition of the aorta and pulmonary trunk seems to be the method used to overcome the abnormal venous return: it often fails because pulmonary stenosis produces a large veno-arterial shunt through a septal defect.

DISCUSSION OF GROUP WITH THE VENOUS ATRIUM ON THE RIGHT AS NORMALLY

When the systemic veins and venous atrium remain on the right as normally, there is no need teleologically for transposition of the great vessels and, if the aorta arose from the left ventricle, the circulation should be normal. This, however, is most unusual and only one of our eight (Case 14) is acyanotic and without much disability. The cyanosis and disability of the others may be entirely due to the associated defects but at least it means that there is some fundamental fault in development that makes these the rule when there is *lævocardia* and transposition of the abdominal viscera.

In discussing the full diagnosis only the cases with necropsies can be used and, omitting the one of Tanner-Cain (1951) for the reasons given, this leaves 7 with the venous atrium on the right (Table II). All these had multiple defects that were more than enough to account for their cyanosis and disability. Three or four, if we include our Case 8 where all the blood from the left atrium had to pass to the right atrium to reach the ventricle, had a single atrium and the other three an atrial septal defect. Four had a single ventricle and the other three a ventricular septal defect. In addition there was pulmonary atresia in two and pulmonary stenosis in three at least.

The venous blood from the systemic veins entered the right atrium or the right side of a single atrium (see Fig. 14), except that the inferior vena cava entered the left atrium in Case 3 of Doliopoulos and Maillet and the left side of a single atrium in our Case 9.

The pulmonary veins entered the left atrium or the left side of a single atrium in all these except one (Case 2 of Young and Griswold), where they passed via the left superior vena cava and the innominate vein to the right side of a single atrium.

In the three cases where there was not a single ventricle, the aorta was transposed and arose from the right ventricle in two (including the only one where the pulmonary trunk was not transposed) and mainly from the right but over-riding the left in one (Case 2 of Young and Griswold). The pulmonary trunk arose from the right ventricle in one (Case 3 of Doliopoulos and Maillet) but was transposed and arose from the left ventricle in two. There was partial transposition in most of those with a single ventricle, but this prevents transposition having its usual functional significance and makes it more difficult to say from which ventricle the aorta would have arisen. Transposition, complete or partial, was therefore an important cause of the cyanosis and disability

FIG. 14.—Diagrams of the angiocardigrams from four cases where the venous atrium is on the right. For explanation see Fig. 13. In (A) the angiocardigram was done from below through the azygos vein and the descending aorta in marked, as well as the arch, to show their relationship. In (C) the filling took place through bilateral *venæ cavæ* and the left was thought to enter the right atrium through the coronary sinus.

in these patients and with the other defects described was more than enough to explain them. All these defects, of course, often occur without transposition of the abdominal viscera.

The Venæ Cavæ. In addition to the 7 cases with necropsies (Table II) the 9 that had angiocardiology and often cardiac catheterization (Table III) can be used for this part of the discussion. We have included all these where a right-sided superior vena cava drained into a right atrium, even if the left superior vena cava was thought to drain into the left side. There was a right superior vena cava (S.V.C.) in all the cases with necropsy and in at least 7 of the 9 others: in all these it entered the right atrium or the right side of a single atrium. In the remaining 2 where there was no proof of a right S.V.C., the left S.V.C. entered the right atrium through the coronary sinus.

There was a left S.V.C. as well as a right S.V.C. in at least 8 cases, while in the previous group with the venous atrium on the left there was never proved to be one on both sides. In 4 of the 8 it drained into the coronary sinus and so reached the right atrium or in one instance the right side of a single atrium. In one instance it received the pulmonary veins and drained into the right S.V.C. (Case 2, Young and Griswold). In another (our Case 9 with necropsy) it went directly into the left side of a common atrium. In one or two, neither supported by necropsy proof (Young and Griswold, Case 4 and less certainly Case 7), it was thought to enter the left atrium. Bilateral venæ cavæ are perhaps even more common than would appear from these figures, for unless both sides are explored at angiocardiology and catheterization (and information about this is not always available) one of these veins may not be demonstrated.

The inferior vena cava presumably went into the right atrium in most of these cases but there was generally no proof of this unless there was a necropsy. In one, however, it went into the left atrium (Case 2, Doliopoulos and Maillet) and in our Case 9 into the left side of a single atrium. In our Case 7 it was absent and the lower half of the body was drained by the azygos vein which entered the right S.V.C. This was also probable in Case 5 of Young and Griswold where a large vein, probably the azygos vein, was seen entering the left S.V.C.

The Pulmonary Veins. In all the cases with necropsy the pulmonary veins drained into the left atrium or the left side of a single atrium, except in the one just mentioned where they drained into a left superior vena cava (Case 2, Young and Griswold). Probably this was also true in the cases who are living but there was less direct evidence except in Cases 4 and 5 of Young and Griswold and in the former one pulmonary vein drained into the right superior vena cava and so into the right atrium. In their Cases 6 and 7, either some pulmonary veins must have entered the right atrium or there must have been a left-to-right shunt through an atrial septal defect.

The Aortic Arch. In the previous group with the venous atrium on the left the aortic arch was always right-sided, but the reverse is not true. When the venous atrium was on the right, the aortic arch was left-sided in 7 and right-sided in 9 cases. The side of the aortic arch was not constantly correlated with the side of the superior vena cava but when there was only a right-sided superior vena cava the aortic arch was more often left-sided (4 of 6 cases) and when there were bilateral superior venæ cavæ the aortic arch was more often right-sided (6 of 8 cases). In the two cases where the only known superior vena cava was on the left but entered the right atrium through the coronary sinus, the aortic arch was right-sided in one and left-sided in one.

ATRIAL AND VENTRICULAR SEPTAL DEFECTS

Septal defects are common in both groups so they are combined in this section. A defect of the atrial septum, varying from a patent foramen ovale (3 cases) to a common atrium (3 cases), was present in 19 of the 24 cases and was thought probable in at least 2 others.

In the 10 cases that came to necropsy, a single ventricle was found in 4, a ventricular septal defect in 5, and no communication between the ventricle in 1 only. A ventricular septal defect was thought to be present in at least 7 of the other 10 cases. Except for the infant described by Tanner-Cain and Crump (1951) every case with necropsy control had a communication between

the atria and between the ventricles, and this seemed likely in many of the others as well. Labesse (1952) has mentioned the frequent occurrence of atrial and ventricular septal defects and that the superior venæ cavæ may be bilateral.

It is difficult to be certain about the arrangement of the ventricles and of the great arteries in cases investigated by clinical methods alone. In the six necropsied cases with both ventricles present the aorta originated from the right ventricle in three, mainly from the right but over-riding the septum in two, and from the left ventricle in one.

TRANSPOSITION OF THE ABDOMINAL VISCERA

In all these cases the abdominal viscera were thought to be transposed but the transposition was not always complete. In the four with necropsies it was complete in Case 1 but in the others the liver was rather central (Case 7) or on both sides (Case 8 with the gall bladder on the left and Case 9 with it just on the right). No spleen was found in Cases 8 and 9 and in Case 7 the spleen had four lobes. In some of the cases investigated clinically the liver was thought to be on both sides (Cases 5 and 10) but in the latter the gall bladder was on the left.

THE BLOOD SUPPLY TO THE LUNGS

The congenital heart disease is usually serious and, with two exceptions, all the cases reviewed were cyanosed. The cyanosis is generally striking and the disability severe. Death at an early age is likely, since only three of these patients were over 10 years of age at the time of examination.

The adequacy of the blood supply of the lungs is of interest from the point of view of surgical treatment. Pulmonary stenosis was found in 4 and pulmonary atresia in 3 of the 10 cases with necropsy, and the pulmonary blood flow was thought to be reduced in 16 of the 24 with necropsy or full investigation. Probably, therefore, most of these patients can be helped by operation in spite of the complex abnormalities.

The only series dealing with the surgical results is Young and Griswold's (1951), on each of whose eight cases a Blalock operation had been performed: three died within seven months of the operation, but the other five were much improved. Five of our patients have had operations: one (Case 4) had a subclavian-pulmonary anastomosis and two years later was a good deal better; two died soon after an anastomosis and in two, one of whom died later, no anastomosis was possible because the pulmonary artery could not be found.

THE RELATIONSHIP OF THE DIRECTION OF PI TO THE SIDE OF THE VENOUS ATRIUM

As in most cyanotic congenital heart disease, right ventricular preponderance is usual in the electrocardiogram. It was found in 22 cases, while left ventricular preponderance was found in four or five only.

Campbell and Reynolds (1952), from a study of these 14 cases and 23 others with complete heterotaxy or isolated dextrocardia, concluded that inversion of PI generally indicates a superior vena cava and a venous atrium on the left side of the heart. PI was inverted (as a rule) in 6 of our 14 cases (Fig. 2, 3, and 4) and in all the 5 where the information was available a left-sided superior vena cava entered a venous atrium on the left (Fig. 13). PI was upright (as a rule) in 8 of our 14 cases (Fig. 9 and 12) and in all the 5 where the information was available a right-sided superior vena cava entered a venous atrium on the right or the right side of a single atrium (Fig. 14).

Cases 1 and 2 of Doliopoulos *et al.* (1952) with an upright PI and Case 4 with an inverted PI support this view. In their Case 5 there was no comment on the position of the venous atrium so probably it was normal and PI was upright, and in their Case 4, PI was isoelectric so it does not help. Nor does the case of Del Carril *et al.* (1949) help for they state that PI was inverted but that a second record showed this was due to nodal rhythm.

Young and Griswold (1951) do not mention the P waves but Dr. Helen Taussig has kindly

given us some information. No notes are available of Cases 1, 2, and 6 and PI was sometimes inverted and sometimes upright in Cases 3 and 4: these five cannot therefore help us. In Cases 5 and 7 the venous atrium was on the right and the former supported our theory by having upright P waves in lead I but the latter opposed it as they were negative or possibly biphasic. In the reported cases where the information is available there were, therefore, four in favour of and one against the theory that the shape of PI indicates the side of the venous atrium.

SUMMARY

We have described 14 cases of congenital heart disease with transposition of the abdominal viscera but with lævocardia, and have compared them with 19 similar cases that have been reported recently. In a lesion of such complexity, only the 10 with necropsies can provide full details of the anatomical changes; but including these there are 10 of our cases and 14 others that have been investigated, generally by angiocardiology and by cardiac catheterization, and these 24 have been analysed and used as far as possible for discussion of the changes.

They fall into two groups (*a*) those where the superior vena cava and venous atrium are on the left, i.e. transposed with the abdominal viscera, and (*b*) those where the venous atrium is on the right as normally, but where the cyanosis and disability are due to the associated defects. The conclusion of Forgacs (1947) that congenital heart disease is the rule in this type of heterotaxy is confirmed, as is the transposition of the atria in the former group, but the assumption that transposition of the atria is the fundamental cause of the other defects is no longer tenable.

In those with the venous atrium transposed to the left, the P wave in lead I is inverted and there is generally a single superior vena cava, also transposed to the left, and a right-sided aortic arch. As a rule, the aorta and pulmonary trunk are partially or completely transposed, but this adaptation, which should be effective, is rendered ineffective by the presence of septal defects and pulmonary stenosis or atresia.

In those with the venous atrium on the right as normally—about half our cases but much more than half of the total number—the P wave in lead I is generally upright and there is often a superior vena cava on both sides, the left generally entering the right atrium through the coronary sinus. The aortic arch may be on either side. As in the first group, the aorta and pulmonary trunk are often partially or completely transposed and there are atrial and ventricular septal defects, often indeed a single atrium or single ventricle, and generally pulmonary stenosis or atresia.

In view of the different arrangement of the atria in these two groups, a different pattern of the other defects in the heart might be expected. They are, however, found to be very similar: atrial and ventricular septal defects, partial or complete transposition, and pulmonary stenosis or atresia are often found in both. Evidently the failure to decide on which side the development is to take place causes fundamental difficulties in the complex processes of growth and rotation that divide the heart into its two sides and bring the aortic and pulmonary rings opposite the correct ventricle.

A Blalock-Taussig operation should be helpful in most cases of both groups though perhaps the result cannot be expected to be as good as in uncomplicated cases of Fallot's tetralogy. If the patient is sufficiently disabled it can be advised whenever the blood flow to the lungs is diminished and suitable pulmonary arteries are present. In the cases reported by Young and Griswold (1951) the results have been good but our experience so far has been less fortunate.

We should like to thank the registrars of the National Heart Hospital for the investigations of Case 4, and Dr. D. Deuchar for the findings at cardiac catheterization, and Dr. T. H. Hills for the angiocardiology of the other cases. We are grateful to Miss M. Waldron of the Department of Medical Illustration for the drawings and Mr. C. Engel of the Photographic Department, Guy's Hospital, for the photographs."

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