Situs Inversus of the Abdominal Viscera with Levocardia

Report of Eight Cases Submitted to the Blalock-Taussig Operation

By Maurice D. Young, F.R.C.P. (Canada), and Herbert E. Griswold, M.D.

A search of the literature has revealed the reports of only 14 cases of situs inversus of the abdominal viscera with levocardia. Eight additional cases are presented in this paper. Angiocardiography and/or heart catheterization was carried out on 6 of these and a Blalock-Taussig operation was performed subsequently on each patient. The autopsy findings in 2 instances, together with the results of special investigations in surviving patients, indicate that the malformations are of a bizarre nature. Anomalies of the venous return to the heart are particularly common and variable degrees of transposition of the great vessels are postulated.

Inversion of the abdominal viscera with levocardia is a rare anomaly in which the abdominal viscera are transposed but the heart occupies its normal position in the left chest. Forgaes has recently reported two instances of this malformation and in a review of the literature found 12 previously reported cases; in every case there was a congenital anomaly of the heart. He implied that due to a paucity of reports no conclusion could be reached concerning the incidence of normal hearts associated with isolated transposition of the abdominal organs.

Inasmuch as only 14 cases have been reported in the literature, it seemed worthwhile to analyze 8 cases of situs inversus of the abdominal viscera with levocardia which have been studied in the cardiac clinic of the Harriet Lane Home. Taussig has emphasized that the cardiac malformation is usually extremely complicated in such patients. Therefore, with two exceptions, special studies were carried out. In addition to the usual clinical examination and routine studies, angiocardiography* and/or cardiac catheterization† was performed on most of these patients. A Blalock-Taussig operation was carried out subsequently on each patient for the relief of inadequate pulmonary blood flow.

The cases are presented in abbreviated form with the autopsy findings in 2 cases and the postoperative progress of each of the remaining 6 patients. The blood analyses are summarized in table 1.

Case 1. C. F. HLH #A 48417: A 23 month old white female with moderate cyanosis and episodes of loss of consciousness. Examination revealed no cardiac enlargement. There was a soft systolic murmur but no palpable thrill. Fluoroscopy and x-ray films showed enlargement of the right ventricle, a concavity in the region of the pulmonary conus, a right aortic arch, and clear lung fields (fig. 1). Blood analysis revealed polycythemia and an extremely low oxygen saturation of arterial blood of 7.5 per cent. (See table 1.)

The clinical diagnosis was tetralogy of Fallot with extreme pulmonary stenosis and a right aortic arch. A Blalock-Taussig operation was performed on the left side by Dr. Alfred Blalock on June 21, 1946, but the infant died during closure of the incision. At operation the superior vena cava was noted to lie on the left side.

Autopsy 82005. (Performed by Dr. J. H. Rid-
The pertinent findings involved the thoracic and abdominal viscera. On opening the abdomen a complete situs inversus of the abdominal viscera was found. There was a perfect mirror image of the usual relations: the stomach and spleen lay on the right side, the liver on the left, and the intestines were transposed. The left pleural cavity contained about 100 cc. of bloody fluid. The left lung was collapsed and was composed of two normally developed lobes and an additional rudimentary upper lobe. The right lung was made up of only two lobes but otherwise appeared normal to the naked eye. Microscopic sections showed patches of intrapulmonic hemorrhage, edema, and atelectasis. Only the alveoli of the right upper lobe were normally aerated.

The heart was predominantly on the left side. The aorta was seen to lie anteriorly to the pulmonary artery. The superior and inferior venae cavae entered the left or posterior auricle. The foramen ovale was patent and fenestrated. The pulmonary artery arose from the left or posterior ventricle, was atretic at its origin, and passed to the left lung only. The infundibulum and pulmonary valve were absent. The right or anterior auricle received the pulmonary veins. From this auricle the blood passed into a large thick-walled (9 mm.) right or anterior ventricle. The aorta was large and overrode the ventricular septum and arose primarily from this ventricle. The aorta ascended and descended to the right of the esophagus. Arising from the aorta through a wide orifice there was a short...

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**Table 1.—A Summary of the Blood Investigations Performed in Eight Patients with Situs Inversus of the Abdominal Viscera and Levocardia.**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Time Interval</th>
<th>Blood Count</th>
<th>Arterial Blood Sample</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>R.B.C. (X 10^6 per cu.mm.)</td>
<td>Hgb. (Gm.)</td>
</tr>
<tr>
<td>1. C. F.</td>
<td>Preoperative...</td>
<td>7.4</td>
<td>20.5</td>
</tr>
<tr>
<td>2. P. K.</td>
<td>Preoperative...</td>
<td>8.95</td>
<td>23.2</td>
</tr>
<tr>
<td></td>
<td>34 mos. Postop.</td>
<td>7.5</td>
<td>18.0</td>
</tr>
<tr>
<td></td>
<td>7 mos. Postop.</td>
<td>5.14</td>
<td>16.0</td>
</tr>
<tr>
<td>3. C. H.</td>
<td>Preoperative...</td>
<td>9.55</td>
<td>23.0</td>
</tr>
<tr>
<td></td>
<td>3 mos. Postop.</td>
<td>7.5</td>
<td>21.0</td>
</tr>
<tr>
<td></td>
<td>4 mos. Postop.</td>
<td>7.75</td>
<td>18.5</td>
</tr>
<tr>
<td>4. T. B.</td>
<td>Preoperative...</td>
<td>9.05</td>
<td>20.1</td>
</tr>
<tr>
<td></td>
<td>2 wks. Postop.</td>
<td>5.2</td>
<td>14.5</td>
</tr>
<tr>
<td></td>
<td>2 yrs. Postop.</td>
<td>5.5</td>
<td>15.0</td>
</tr>
<tr>
<td>5. R. C.</td>
<td>Preoperative...</td>
<td>8.4</td>
<td>23.5</td>
</tr>
<tr>
<td></td>
<td>9 mos. Postop.</td>
<td>6.8</td>
<td>17.0</td>
</tr>
<tr>
<td></td>
<td>21 mos. Postop.</td>
<td>7.75</td>
<td>15.8</td>
</tr>
<tr>
<td>6. G. W. K.</td>
<td>Preoperative...</td>
<td>8.45</td>
<td>22.5</td>
</tr>
<tr>
<td></td>
<td>2 wks. Postop.</td>
<td>5.3</td>
<td>14.2</td>
</tr>
<tr>
<td></td>
<td>19 mos. Postop.</td>
<td>4.27</td>
<td>13.5</td>
</tr>
<tr>
<td>7. J. E.</td>
<td>Preoperative...</td>
<td>7.7</td>
<td>22.3</td>
</tr>
<tr>
<td></td>
<td>1 yr. Postop.</td>
<td>6.16</td>
<td>15.5</td>
</tr>
<tr>
<td>8. B. D.</td>
<td>Preoperative...</td>
<td>9.9</td>
<td>24.2</td>
</tr>
<tr>
<td></td>
<td>2 mos. Postop.</td>
<td>5.9</td>
<td>17.0</td>
</tr>
<tr>
<td></td>
<td>1 yr. Postop.</td>
<td>4.27</td>
<td>13.5</td>
</tr>
</tbody>
</table>

* By oximetry.
wide ductus arteriosus which bifurcated, sending one branch to the rudimentary pulmonary artery and the other branch directly to the right lung. The left subclavian artery arose from a left innominate artery. The surgical anastomosis of this vessel to the side of the left pulmonary artery was patent. One large bronchial artery passed to the right lung posteriorly.


![Fig. 2. Case 2, P. K. Roentgenogram of chest showing cardiac outline with prominent pulmonary conus and right aortic arch.](image)

Case 2. P. K. HLH #A 54017. A 10 year old white boy with moderate cyanosis and limited exercise tolerance. Examination revealed a harsh systolic murmur and a slight systolic thrill; there was no cardiac enlargement. X-ray films and fluoroscopy revealed slight prominence in the region of the pulmonary conus and slightly increased lung markings but no pulsações were visible in the lung fields (fig. 2). There was a right aortic arch. The stomach was seen to lie on the right side and the liver lay on the left side. Blood analysis revealed a marked polycythemia; the oxygen saturation of an arterial blood sample was 55.5 per cent. (See table 1.) The electrocardiogram showed right axis deviation and hypertrophy of the anterior ventricle.

The angiocardiograms were interpreted as showing an overriding aorta and a high ventricular septal defect; the pulmonary artery was poorly visualized. The standard exercise test showed a drop in the oxygen consumption. The pulmonary capillary blood flow determined by the indirect test was low.

This boy was considered to have situs inversus of the abdominal viscera with levocardia, a pulmonic stenosis with a ventricular septal defect and an overriding aorta. A Blalock-Taussig operation was performed successfully by Dr. Alfred Blalock on April 8, 1947. Following operation the patient gradually developed cardiac failure, from which he died seven months later.*

Autopsy $\#3894-47$. (Performed by Dr. A. H. Wells and Dr. H. H. Jaffe.) The chief interest centered about the heart and abdominal organs. The heart was in its normal position, with the apex toward the left. There was obvious hypertrophy of the right side. The heart was enlarged and weighed 230 Gm. (normal 110 to 135 Gm.). There was complete absence of the auricular septum and absence of the upper third of the ventricular septum. The common auricular cavity was grossly enlarged to about three times the combined size of two normal auricles, and the cavity of the right ventricle was about three times its normal size. The cavity of the left ventricle was quite small and admitted the tips of two fingers.

The aorta arose from the right or anterior ventricle and did not override the ventricular septal defect. It lay anteriorly and to the right of the pulmonary artery which arose from the left or posterior ventricle. The pulmonary valves were fused so that there was only a slitlike opening measuring 8 mm. in length. The pulmonary artery was very small beyond the valve and did not measure more than 1 cm. in diameter at any point in its course.

The aorta measured approximately 2 cm. in diameter in its ascending portion. The proximal end of the innominate artery was anastomosed to the side of the left pulmonary artery. The anastomosis was patent and the intimal surface of the vessels was quite smooth and apparently covered by endothelium. The ductus arteriosus was obliterated.

A large vein, believed to be a left superior vena cava, crossed from left to right anteriorly to the aortic arch and received the pulmonary veins from both lungs. This vein entered the right superior vena cava which passed into the right side of the single auricle. The inferior vena cava was in its normal position. The origin of an anomalous vein thought at operation to be the inferior vena cava was not determined.

There were 300 cc. of clear fluid in the right pleural cavity. The right lung was well aerated and slightly emphysematous. The left lung was covered by dense fibrous adhesions and was almost completely atelectatic. There was moderate congestion throughout

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* For the interval history and for the report of the autopsy findings we are indebted to Dr. Roy C. Pederson, Dr. A. H. Wells, and Dr. H. H. Jaffe of Duluth, Minn.
both lungs, but no consolidation. Both lungs had three lobes.

Each of the abdominal viscera lay in a transposed position so that there was a perfect mirror image of the normal relations. The liver extended 6 cm. below the left costal margin; it weighed 1450 Gm. (normal 700 to 800 Gm.) and showed severe chronic passive congestion.


Comment. These 2 cases illustrate the complicated anatomy which may be present in a patient who has situs inversus of the abdominal viscera and levocardia. In each patient there was a mirror image of the normal relations of the abdominal viscera and a complicated malformation of the heart. Certain features were similar; namely, the inadequate pulmonary blood flow with considerable enlargement of the right ventricle, the anomalous pulmonary venous return to the right side of the heart, the transposition of the great vessels, and the ventricular septal defect.

In other respects the malformations were different. In case 1, the foramen ovale was patent and the systemic veins entered the left auricle. The aorta arose primarily from the right ventricle but overrode the ventricular septum. There was pulmonary atresia so that the pulmonary circulation was maintained through a patent ductus arteriosus, a rudimentary pulmonary artery, and the bronchial arteries. Theoretically, the patient should have benefited from an increase in the blood supply to the lungs, but she died suddenly at the conclusion of the operation. The cause of death was not apparent, but it would seem that the production of an artificial ductus arteriosus placed too sudden a strain on the heart. In case 2, there was a complete absence of the auricular septum and the systemic veins entered the single auricle in their normal position. The right superior vena cava received blood from the pulmonary veins by way of the left superior vena cava. The aorta arose entirely from the right ventricle and did not override the ventricular septum; the ductus arteriosis was obliterated. The left ventricle was small and underdeveloped. There was marked pulmonic stenosis, but apparently there had been sufficient blood flow through the pulmonary vessels to maintain life. Immediately following operation this patient developed cardiac failure from which he died seven months later. In this instance, therefore, death was not immediate, but the altered circulation led to slowly progressive heart failure.

These patients are the only 2 in this series on whom autopsies have been performed and concerning whom the exact nature of the malformations is known. However, in each of the remaining patients, with but one exception, investigations demonstrated the existence of a complicated cardiac malformation.

Case 3. C. H. HLH SA 60487: A 15 month old white female with moderate to marked cyanosis, a harsh croupy cough, and attacks of paroxysmal dyspnea. There was no cardiac enlargement; no thrill was palpable and no murmurs were audible. The liver edge was palpable under the left costal margin. Fluoroscopy showed that the heart was boot-shaped in outline and there was a left aortic arch; the lung fields appeared abnormally clear; a pulmonary artery was visible on the right side only; the stomach and liver were transposed. Electrocardiography displayed right axis deviation and hypertrophy of the anterior ventricle. There was marked polycythemia and a low arterial blood oxygen saturation of 28.5 per cent. (See table 1.)

Angiocardiography demonstrated a left superior vena cava and a small pulmonary artery on each side, and the films were interpreted as showing an auricular septal defect, pulmonic stenosis and an overriding aorta. (See fig. 3.) The preoperative diagnosis was situs inversus of the abdominal viscera with levocardia, tetralogy of Fallot with an auricular septal defect, a left aortic arch and a left superior vena cava.

An anastomosis was performed between the end of the right subclavian artery and the side of the right pulmonary artery by Dr. Alfred Blalock on May 4, 1948. As expected, no superior vena cava was found on this side. The immediate postoperative progress was uneventful and there was temporary improvement in the patient's condition. Subsequently she developed generalized edema, cardiac enlargement and evidence of renal failure. She died six months after operation from cardiac failure and anuria with uremia and convulsions. Permission for autopsy was not granted.

Comment. This patient is the third in this series who died following a Blalock-Taussig...
operation. Cardiac failure with progressive cardiac enlargement was gradual in onset, but as in case 2 the patient died approximately six months after operation. In this instance the urinary findings suggested that death was due to a combination of cardiac failure and renal insufficiency. The exact nature of the cardiac malformation is not known. Clinical and radiologic evidence pointed to the existence of an anomalous systemic venous return, an auricular septal defect, and pulmonic stenosis. The aorta was seen to arise anteriorly and far to the right both in the angiocardiogram and at operation. It may be postulated, therefore, that there may have been a transposition of the great vessels as in case 2.

**Fig. 3.** Case 3, C. H. Angiocardiogram. Injection through right external jugular vein. *Top left:* Anteroposterior view (1 to 2 seconds) showing left superior vena cava and early filling of both auricles. *Top right:* Anteroposterior view (2 to 3 seconds) showing early visualization of aorta which appears to arise more to the right than normal, sweep upwards to the right before crossing to form a left aortic arch, and descend on the left. *Bottom left:* Oblique view (1 to 2 seconds) showing filling of both auricles. *Bottom right:* Oblique view (2 to 3 seconds) showing early visualization of aorta which appears to arise anteriorly.

**Case 4.** T. B. HLH $A-53589$. A 4 year old white boy with marked cyanosis and brief periods of unconsciousness. On examination a systolic thrill was palpable and a harsh systolic murmur was audible over the precordium. Fluoroscopy revealed that the heart was not enlarged; there was a slight concavity in the region of the pulmonary conus; the lung fields appeared clear; the aortic arch was on the right side; the stomach was on the right, and the liver shadow was on the left side. The electro-
cardiogram showed right axis deviation and a predominance of the anterior ventricle. Blood analyses revealed marked polycythemia, and an arterial blood oxygen saturation of 67.6 per cent. (See table 1.)

Angiocardiography, performed on two occasions on the right and left sides respectively, revealed bilateral superior venae cavae, one to each auricle, a normal origin of the pulmonary artery, a ventricular septal defect, and an overriding aorta. Cardiac catheterization, also carried out on each side, confirmed the presence of two superior venae cavae and anomalous return of the pulmonary and systemic circulations. A Blalock-Taussig operation was performed by Dr. Alfred Blalock on April 1, 1947; the postoperative progress was uneventful. The patient was reported to be remarkably improved 22 months after operation.* He was able to walk an almost unlimited distance and had only residual cyanosis. At that time a humming continuous murmur was audible over the base of the heart. His heart had enlarged but there was no evidence of cardiac failure. The results of blood examination are included in table 1.

The results obtained from this investigation were as follows:

<table>
<thead>
<tr>
<th></th>
<th>Pressure (mm. Hg.)</th>
<th>Blood O₂ Content (vols. %)</th>
</tr>
</thead>
<tbody>
<tr>
<td>R. sup. vena cava</td>
<td>15.4</td>
<td>80/12</td>
</tr>
<tr>
<td>R. auricle</td>
<td>15.3</td>
<td>15.1</td>
</tr>
<tr>
<td>R. ventricle</td>
<td>10.3</td>
<td>25/11</td>
</tr>
<tr>
<td>L. sup. vena cava</td>
<td>20.4</td>
<td>120/8</td>
</tr>
<tr>
<td>L. auricle</td>
<td>19.9</td>
<td>19.9</td>
</tr>
<tr>
<td>Femoral Artery—content 19.9 vols. %</td>
<td>capacity 26.5 vols. %</td>
<td>saturation 75%</td>
</tr>
</tbody>
</table>

On the assumption that the blood returning from the lungs was 96% saturated, the oxygen content of the blood in the pulmonary veins was calculated to be 25.6 vols. per cent.

The results were interpreted as indicative of inadequate pulmonary blood flow and an anomalous return of at least one pulmonary vein to the right superior vena cava.

The preoperative diagnosis was situs inversus of the abdominal viscera with levocardia, pulmonary stenosis, overriding aorta with a right aortic arch and situs inversus of the upper extremities. The patient was a boy, 3 years old.

Case 5. R. C. HLH #A 54377: An 8½ year old white boy with limited exercise tolerance in whom heart disease was diagnosed at the age of 1 year, but in whom cyanosis did not appear until between the ages of 3 and 4 years. The heart was enlarged to the left; a systolic thrill was palpable and both a systolic murmur and a faint diastolic murmur were audible. The liver was palpable under the left costal margin. Fluoroscopy revealed a generalized cardiac enlargement with a broad shadow at the base; the lung markings were slightly prominent but no pulsations were visible; the aortic arch was on the right side; the stomach was on the right, and the liver was situated on the left. X-ray films of the chest confirmed these observations (fig. 4). The electrocardiogram revealed left axis deviation and predominance of the posterior ventricle. There was polycythemia; the arterial blood oxygen saturation was 66.2 per cent (see table 1).

Angiocardiography revealed a left superior vena cava and prompt filling of the aorta. On cardiac catheterization, performed on two occasions, blood oxygen saturation was 67.6 per cent. (See table 1.)

* For this report, dated March 16, 1949, and for permission to refer to it, we are indebted to Dr. M. Cooperstock of Marquette, Michigan.
catheterization the catheter passed down a left superior vena cava and appeared to enter the right auricle through the coronary sinus and thence passed to the left auricle; it was impossible to pass the catheter into a ventricle. The oxygen content of the superior vena cava blood was 18.1 vols. per cent, of the right auricle blood 20.5 vols. per cent, and of the left auricle blood 29.4 vols. per cent. The oxygen content of the blood in the pulmonary vein (obtained by catheterization) was 29.4 vols. per cent. The total pulmonary capillary blood flow was calculated to be 1730 cc. per minute per square meter of body surface.* These findings indicated the presence of a left superior vena cava, an auricular septal defect, and a low pulmonary blood flow.

In view of the pulmonic stenosis and the right to left intracardiac shunt, a Blalock-Taussig anastomosis was performed by Dr. Alfred Blalock on April 25, 1947. At operation a large vein, considered to be the inferior vena cava, was found to enter the left superior vena cava.

This boy's postoperative progress was uneventful.† His exercise tolerance has increased greatly. Examination nine months after operation revealed only mild cyanosis without clubbing of the fingers; a loud continuous murmur was audible; the heart was almost identically the same size as before operation and there was no evidence of cardiac failure. The results of postoperative blood examinations are shown in table 1.

Case 6. G. W. K. HLH #A 57195: A 94 year old white boy with marked cyanosis and limited exercise tolerance. At the age of 8 years he had contracted a severe throat infection complicated by a septicemia, heart failure and auricular fibrillation. Upon examination the heart was normal in size; a systolic thrill was palpable and a widely transmitted harsh systolic murmur was heard. The liver was felt on the left side and the stomach resonance was on the right. Fluoroscopy revealed clear lung fields; the heart was not enlarged in the anterior-posterior view but in the left anterior oblique position there appeared to be enlargement of the anterior ventricle; there was a left aortic arch. Blood count revealed a polycythemia; the arterial blood oxygen saturation was 64.8 per cent. (See table 1.) The electrocardiogram showed a tendency to right axis deviation and hypertrophy of the anterior ventricle.

Angiocardiography demonstrated a left superior vena cava; the pulmonary artery appeared to arise posteriorly to the aorta.

A standard exercise test showed a fall in the oxy-

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† According to the formula of Bing and associates.¹

* For this report and for permission to refer to it we are indebted to Dr. Marvin Schwartz of Portland, Oregon, and for the arterial blood analysis we wish to thank Dr. Elton Watkins of the University of Oregon Medical School.

† For this report, dated April 29, 1949, and for permission to refer to it, we are indebted to Dr. N. B. Livingston, Jr., of Mohall, North Dakota.
systolic thrill and a high-pitched widely transmitted systolic murmur; the heart was not enlarged; the liver was palpable under the left costal margin. Fluoroscopy revealed a concavity in the region of the pulmonary conus but the pulmonary arteries and the pulmonary vascular markings were more prominent than expected. There was a right aortic arch. The stomach was seen on the right side. The electrocardiogram revealed right axis deviation and preponderance of the anterior ventricle. Blood examination showed a polycythemia; the arterial blood oxygen saturation was 72 per cent. (See table 1.)

The angiocardiogram was interpreted as demonstrating a left superior vena cava, small pulmonary arteries and a large aorta which appeared to arise far anteriorly; the appearances were considered to be consistent with the diagnosis of tetralogy of Fallot.

The following results were obtained from cardiac catheterization:

<table>
<thead>
<tr>
<th>Pressure</th>
<th>Blood O2 Content</th>
</tr>
</thead>
<tbody>
<tr>
<td>mm. Hg.</td>
<td>vols. %</td>
</tr>
<tr>
<td>R. sup. vena cava</td>
<td>17.8</td>
</tr>
<tr>
<td>R. auricle</td>
<td>19.6</td>
</tr>
<tr>
<td>R. ventricle</td>
<td>92/2</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>8/0</td>
</tr>
<tr>
<td>Pulmonary vein (catheterized)</td>
<td>29.6</td>
</tr>
</tbody>
</table>

Femoral artery—content 21.2 vols. %

—capacity 30 vols. %
—satisfaction 71%

From these figures the pulmonary artery blood flow was calculated to be 1620 cc. per minute per square meter of body surface, and the systemic blood flow was calculated to be 4100 cc. per minute per square meter of body surface; thus there was a right to left shunt of 2480 cc. On the basis of the low pulmonary artery flow and the low pulmonary artery pressure associated with the right ventricular pressure, it was felt that there was pulmonic stenosis. The difference between the oxygen content of the blood in the superior vena cava and that in the right auricle suggested that there was either an aberrant return of one or more of the pulmonary veins to the right auricle and/or an auricular septal defect.

In view of the inadequate pulmonary blood flow and the evidence of a right to left intracardiac shunt, a Blalock-Taussig operation was performed by Dr. W. P. Longmire on April 6, 1948. Following operation the patient developed a high fever, and blood cultures yielded organisms identified as either alpha or gamma streptococci. After intensive penicillin therapy the bloodstream infection eventually was cured. The patient was discharged home 2 months after operation.

Since discharge from the hospital her progress has been very satisfactory. She tires less easily and is less dyspneic. She is able to walk a mile with ease and has been riding a tricycle. Examination 11 months after operation revealed minimal clubbing of the fingers and barely perceptible cyanosis at rest. A well-marked blowing continuous murmur was audible over the left upper chest anteriorly and posteriorly. Her heart had increased in size but there was no evidence of cardiac failure. Blood count at that time revealed a mild polycythemia; the oxygen saturation of an arterial blood sample was 77 per cent.

Case 8. B. D. HLH # A 61973: A 5 year old boy with marked cyanosis and considerable limitation of activity. An attempt to perform a Blalock-Taussig anastomosis in France had been unsuccessful. Upon examination he was poorly nourished and poorly developed. The heart was slightly enlarged. No thrill was palpable and only a faint systolic murmur was audible. The liver was palpable in the epigastrium. Fluoroscopy revealed a slightly enlarged heart, boot-shaped in outline; there was a concavity in the region of the pulmonary conus and the lung fields appeared quiet. There was a right aortic arch. The stomach was seen on the right side and the liver lay on the left. The electrocardiogram showed right axis deviation and anterior ventricular hypertrophy. The blood count revealed marked polycythemia; the oxygen saturation of an arterial blood sample was 58.5 per cent (see table 1).

The clinical diagnosis was situs inversus of the abdominal viscera with levocardia and tetralogy of Fallot. In April 1948 Dr. Alfred Blalock made an anastomosis between the end of the left subclavian artery and the side of the left pulmonary artery. The patient's postoperative progress was relatively uneventful. Digitalis therapy was instituted because of tachycardia and some enlargement of the liver.

Following operation the patient showed distinct improvement. Examination two months later revealed an easily palpable systolic thrill, a loud blowing systolic murmur, and in addition a well-marked blowing continuous murmur. The polycythemia was less and there was a considerable rise in the arterial blood oxygen saturation. (See table 1.) A follow-up report* a year after operation stated that he was then able to walk 11 miles without fatigue. No cyanosis was visible at rest, he was not dyspneic, and he had gained weight. A continuous murmur was easily heard. There had been a further drop in the blood count.

**Discussion**

An analysis of the cases presented in this series supports the impression that cardiac mal-

* For this report, dated April 11, 1949, and for permission to refer to it, we are indebted to Prof. Donzelot, Dr. Cottraux, and Dr. A. Pithon of the Broussais Hospital, Paris.
### Table 2—Summary of Findings

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Age</th>
<th>Clinical Diagnosis</th>
<th>Aort. Arch</th>
<th>ECG</th>
<th>Exercise Test</th>
<th>Heart Catheterization</th>
<th>Angiocardiology</th>
<th>Preoperative Diagnosis</th>
<th>Operative Findings</th>
<th>Operative Pathology</th>
<th>Operation</th>
<th>Remarks</th>
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<tbody>
<tr>
<td>No.</td>
<td>Sex</td>
<td>Age</td>
<td>Diagnosis</td>
<td>Side</td>
<td>RAD</td>
<td>RVH</td>
<td>Findings</td>
<td>Tetralogy</td>
<td>Lesion</td>
<td>Postoperative Comments</td>
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</table>

formations are complicated when they exist in combination with situs inversus of the abdominal viscera and levocardia. The findings in the individual patients are summarized in table 2.

The presence of situs inversus of the abdominal viscera may be easily overlooked, particularly in the patient who is severely incapacitated and whose tolerance is low since under such circumstance examination has to be rapid. Such an oversight occurred in case 1. Unless the liver is palpable, clinical examination offers no clue to the existence of situs inversus. In order to ensure correct diagnosis it is therefore advisable to make a practice of lowering the fluoroscope screen after each esophagram in order to locate the position of the stomach. When a patient with a congenital cardiac defect is found to have situs inversus of the abdominal viscera, a complicated malformation should be suspected and further investigations should be carried out in an attempt to elucidate more accurately the nature of the malformation.

The mortality from operation in this group is high. Three of the 8 patients died. This is distinctly higher than the mortality with the Blalock-Taussig operation performed by the same group of surgeons in cases of simple tetralogy of Fallot, but is to be expected from the bizarre nature of the malformation. Although statistics in such a small group are apt to be erroneous, the serious risk of operation deserves mention. In another respect the group resembles the cases of simple tetralogy of Fallot in that the deaths occurred at the time of operation or within about six months thereafter; those who survived the first six months have continued to progress satisfactorily.

Evidence is abundant that there was inadequate pulmonary blood flow in every patient in this series. Clinical examination and fluoroscopy indicated that this was so; heart catheterization demonstrated it in cases 4, 5, 6, and 7; angiography revealed small pulmonary arteries in cases 2, 3, 4, and 7; and in every instance at operation the pulmonary artery pressure was considered to be low. In the 2 cases which came to autopsy complete atresia of the pulmonary artery was found in case 1 so that the pulmonary circulation was maintained through a patent ductus arteriosus and a large bronchial artery, and in case 2 there was a marked pulmonary stenosis.

It is noteworthy that the aorta arched to the right in 6 of the 8 patients. As is usual in a person with a right aortic arch, in each of these 6 cases the innominate artery was situated on the left, i.e. the aorta and its main branches lay in the mirror image of the normal relations. In each of 2 instances (cases 4 and 8) an obliterated ductus arteriosus was seen at operation to extend from the left subclavian artery to the pulmonary artery.

Analysis of the 7 cases in which data in addition to the clinical findings are available reveals certain striking features. Anomalies of the systemic venous return to the heart were found in all 7 cases. Bilateral superior venae cavae were found in 4 patients (cases 2, 4, 6, and 7). In case 4 the right and left superior venae cavae were known to have entered respectively the right and left auricles, while the same was considered probable in both case 6 and case 7. In case 2 the left superior vena cava crossed to enter the right superior vena cava and both emptied into the right auricle. In cases 1, 3, and 5, a single superior vena cava was demonstrated on the left side in each instance. The venae cavae entered the left auricle in case 1, the superior vena cava emptied into the right auricle through the coronary sinus in case 5, and in case 3 the site of termination of the superior vena cava could not be ascertained.

Anomalies of the pulmonary venous return were demonstrated in 3 instances (cases 1, 2, and 4) and were suspected in 2 others (cases 6 and 7). In both cases 1 and 2, all pulmonary veins were found at autopsy to enter the right side of the heart. In case 4, at least one pulmonary vein was demonstrated by heart catheterization to enter the right superior vena cava; in case 6 one or more of the pulmonary veins appeared to drain into the right auricle since the catheter apparently entered a pulmonary vein directly from the right auricle. However, the possibility that the catheter slipped through an auricular septal defect into the left auricle and thence into a pulmonary vein cannot be entirely excluded. It was suspected on heart
catheterization that one or more of the pulmonary veins entered the right auricle directly in case 7, but this could not be proved as an auricular septal defect may produce similar findings. An auricular septal defect was known to exist in each of 2 patients as it was demonstrated by angiocardiography in case 3 and by heart catheterization in case 5; in case 1 there was a patent foramen ovale and in case 2 a single auricle was found at autopsy. Thus of the 7 cases under discussion, a communication between the auricles was known to exist in 4 patients, was suspected in one (case 7) and could not be excluded in a sixth (case 6).

Prior to operation in every instance the aorta was believed to override the ventricular septum. However, at autopsy the aorta was found to be transposed in case 2 and to arise mainly from the right ventricle in case 1. The possibility that the position of the aorta was a factor in the failure of the heart to withstand the strain of the altered circulation after operation has to be considered in each of these cases. A similar possibility has to be borne in mind in case 3, as the aorta was seen in the angiocardiogram to be dextroposed and a diagnosis of transposition of the aorta was considered. In case 6 the angiocardiogram demonstrated that the aorta lay anteriorly to the pulmonary artery and in case 7 the aorta was seen to arise far anteriorly. In each of these last 2 instances transposition of the aorta is therefore a possibility, but both these patients have progressed well since operation in contrast to the first 3 patients, so that there must be additional factors involved.

The pulmonary artery arose from the left ventricle in case 1 and also in case 2. In these cases, therefore, there was a complete transposition of the great vessels, although in case 1 the aorta did override the septum to a mild degree. Since the pulmonary venous return was to the right auricle and the systemic venous return was to the left auricle in case 1, the malformation amounted to a corrected transposition of the great vessels with an overriding aorta. In case 2 the pulmonary veins entered the superior vena cava so that blood from both the systemic and the pulmonary circulations entered the right side of the single auricle.

In this instance, therefore, there was a partially corrected transposition of the great vessels inasmuch as the aorta, pulmonary artery and pulmonary veins were transposed. A similar but not identical malformation may have been present in both case 6 and case 7. In the former, the origin of the pulmonary artery was seen in the angiocardiogram to lie posteriorly to the aorta, and it was known after heart catheterization that one or more of the pulmonary veins probably entered the right auricle; furthermore, there was a left superior vena cava which appeared to empty into the left auricle. These findings led to the consideration that a corrected transposition of the great vessels may have been present. That this was not the complete picture was demonstrated by the fact that there was also a right superior vena cava which entered the right auricle. Almost identical possibilities were present in case 7, except that the exact relation of the aorta to the origin of the pulmonary artery was not clearly visualized in the angiocardiogram although the aorta was seen to arise far anteriorly.

In case 3, there is less evidence to support consideration of the possibility of a transposition of the great vessels. Nevertheless, the aorta was seen to arise far to the right both on angiography and at operation. In addition the superior vena cava was seen in the angiocardiogram to be on the left side and at operation no superior vena cava was found on the right side. Therefore, there is at least a suggestion of the possibility that a corrected transposition of the great vessels was combined with some other anomaly.

Thus, in brief, a corrected transposition of the great vessels combined with pulmonary stenosis existed in case 1, and a partially corrected transposition of the great vessels was present in case 2 and may be postulated in each of cases 3, 6, and 7.

The pathogenesis of the cardiac defects in cases of isolated inversion of the abdominal viscera has been discussed by Forgacs. Of the 14 cases in his review, the auricles were transposed in 8, there was a common auricle in each of 2, and in 4 cases the disposition of the auricles could not be ascertained. In his article he attempted to correlate the findings in this group
with those in cases of isolated dextrocardia, and pointed out that transposition of the auricles may be regarded as the fundamental defect in most cases of each group. According to his argument, the transposition of the auricles is dependent upon the embryonic abdominal venous channels. A reversal of the normal relation between heart and liver is associated with a corresponding change in the hepatic venous channels, with the result that in cases of situs inversus of the abdominal viscera the venous drainage is to the left instead of to the right side of the heart. This theory was also advanced by Lochte.\textsuperscript{4}

On the whole, our series supports this view. However, it is apparent that in some cases the transposition is incomplete. In consequence various degrees of transposition of the venous return to the heart are to be expected in combination with various degrees of transposition of the great vessels. Some of the variations are exemplified by the cases in this series.

**Summary**

1. Eight cases of congenital malformations of the heart of the cyanotic group occurring in association with situs inversus of the abdominal viscera with levocaridi have been presented in abbreviated form.

2. The bizarre nature of the malformations has been demonstrated by heart catheterization, angiocardiography, operative findings, and in 2 cases at autopsy.

3. A summary of the findings in each case is given in table 2.

4. Analysis of the cases has been undertaken wherever available data have permitted. This analysis reveals that (a) Situs inversus of the abdominal viscera may be easily overlooked. (b) The mortality from operation in this group is relatively high. (c) Inadequate pulmonary blood flow was demonstrated in every case in our series. (d) The aorta and its main branches bore a mirror image relationship to the normal anatomy in 6 of the 8 cases. (e) Anomalies of the systemic venous return to the heart are very common in this type of malformation, and in this series occurred in 7 of the 8 cases. (f) Anomalies of the pulmonary venous return were demonstrated in 4 cases and suspected in one additional case. (g) A communication between the auricles was found in each of 4 cases and was suspected in 2 additional cases. (h) The aorta was dextroposed in each case; it was completely transposed in one case, and mainly transposed in another; the possibility of transposition of the aorta is postulated in 3 further cases. (i) Transposition of both great vessels was demonstrated in 2 cases and postulated in 3 other cases.

5. The likelihood of a corrected transposition of the great vessels in each of 5 of the 8 cases is discussed.

6. The possible pathogenesis of the cardiac defects is briefly mentioned.

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Situs Inversus of the Abdominal Viscera with Levocardia: Report of Eight Cases
Submitted to the Blalock-Taussig Operation
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