Partial Situs Inversus with Levocardia

An Unusual Combination of Anomalies

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An instance of a unique combination of anomalies was recently encountered in an infant with cyanotic congenital heart disease who died at the age of 13 months. The clinical course and pathologic findings are described.

Case History

The patient was a female baby, born at full term after an uneventful pregnancy. At birth she weighed 6 lbs. and was cyanotic. Heart disease was first diagnosed at the age of 3 months. The parents had noted poor weight gain, delayed development, and generally poor health. The family history was nonecontributory.

At 7 months she weighed 9 lbs. 7 oz. and could not sit up. Physical examination revealed marked cyanosis and clubbing, perhaps more marked in the toes. The heart rate was regular at 150 per minute; the heart was not enlarged. A grade II systolic murmur was heard in the pulmonary area with a normal second sound. The liver was felt two fingers enlarged in the epigastrium. The femoral pulsations were normal.

On fluoroscopy the heart was slightly larger than normal, the pulmonary conus was not remarkable. The upper mediastinal shadow was broad and extended well to the right, suggesting extreme dextroposition of the aorta. The lung fields were oligemic. In the right anterior oblique view the aorta arched to the left. In the left anterior oblique position the large supracardiac shadow to the right was confirmed as a large dextroposed aorta. The right ventricle was diminutive and did not extend as far forward as the aorta above. The pulmonary window was not clear and the left ventricle was enlarged.

An x-ray confirmed the fluoroscopic findings (fig. 1). The liver appeared transposed with the larger lobe on the left, although with barium the stomach was on the left. Serial electrocardiograms were taken (fig. 2). Examination of the blood revealed a hemoglobin of 12 Gm. per 100 ml. and a hematocrit level of 56 per cent. The white cell count was 7,000 per cu. mm.

Figure 1
Posteroanterior view of chest showing enlarged heart, broad superior mediastinum, and oligemic lung fields.

At 10 months of age, on February 13, 1960, the patient’s weight was 10 lbs. 4 oz. and she was in frank congestive failure with peripheral edema, moist sounds in the lungs, marked dyspnea, and enlargement of the liver (three fingers below the costal margin). She was digitalized and given diuretics with only temporary relief. On February 23, 1960, the heart rate became slow. The electrocardiogram (fig. 2) suggested nodal rhythm. She was too ill for any investigative procedures and died on March 31, 1960.

At autopsy deep cyanosis, clubbing of the digits, and slight edema of the feet were present. The heart was in normal position in the thorax. The pericardium was normal. A right innominate vein entered a left superior vena cava that entered the left atrium posteriorly (fig. 3). The right superior vena cava was absent. The inferior vena cava also opened into the left atrium. The coronary veins drained into the left atrium through two openings.

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one the normal opening of the coronary sinus and another smaller opening lower down via the thebesian vein. The four pulmonary veins all drained separately into the right atrium. There were two atrial septal defects situated anteriorly, separated by a thin band of tissue, with a crescentic edge concave anteriorly as viewed from the left (fig. 4). One was about 1 cm. in diameter, the other was smaller. The fossa ovalis could not be identified; these two septal defects presumably represented an ostium secundum. The mitral and tricuspid valves were normal (fig. 5). The position of the left atrial wall resembled the normal right atrial position. The right ventricle was small and there was no trace of the pulmonary artery arising from this ventricle. There was a high ventricular septal defect about 8 mm. in diameter. The left ventricle was dilated and hypertrophied; the wall was 1 cm. thick. A large trunk arose from this ventricle and had three semilunar cusps situated rather higher than normal (fig. 6). The coronary orifices were identified behind the posterior and anterolateral cusps. This large trunk was wider than the normal aorta and just before giving off the three main branches gave off a pulmonary artery from its posterior surface that branched into two, one for each lung. The aorta arched to the left; there was no trace of a ductus arteriosus. There were no large bronchial arteries. The liver was greatly enlarged. The left lobe was as large as the right and extended to the left hypochondrium. The gallbladder was on the right and the stomach and intestines were in the usual position. The spleen was bilobed and weighed 25 Gm.

The anatomic diagnoses at autopsy were partial situs inversus with levocardia, persistent left superior vena cava (absent right superior vena cava), transposition of the atria, two atrial septal defects, high ventricular septal defect, diminutive right ventricle with no pulmonary artery arising from it and trunca arteriosus with pulmonary arteries arising from it.

Discussion

This case would appear to be an instance of the group of malformations associated with situs inversus (partial or complete) and levocardia. The large left lobe has been considered to represent partial situs inversus,1 Taussig2 stated that in this condition a corrected transposition is common, i.e., transposition of the atria and of the great vessels, one abnormality correcting the other. Young and Griswold3 in a review of eight cases of situs inversus with levocardia showed evidence of anomalies of the systemic venous return in seven, of the pulmonary venous return in four, and suspected in one more. Defects of the atrial septum were found in four and suspected in two. In all eight cases there was evidence of some dextroposition of the aorta; complete transposition was present in two and postulated in three more. The first of the eight cases that came to autopsy had a corrected transposition with pulmonary stenosis. The second case that was autopsied had transposition of the atria, of the A-V valves, and of the great vessels. There were pulmonary stenosis, a large ventricular septal defect, and an overriding pulmonary artery. Two other cases in their series that came to operation had similar findings. In the others the diagnosis was based on cardiac catheterization or angiographic data. Only one case in the series showed left ventricular preponderance.

Campbell and Forgacs1 described 14 cases of transposition of the abdominal visceras with levocardia and compared them with 19 similar cases reported in the literature. There were only 10 necropsies in the whole series but 24 cases had been investigated by angiography or cardiac catheterization. They described two groups: (1) those with superior vena cava and venous atrium on the left, i.e., transposed; (2) those with a venous atrium on the right and where the cyanosis was due to associated defects. In cases with venous atrium transposed to the left, 

P

1 was inverted and there was a single left superior vena cava with a right-sided aortic arch. The aorta and pulmonary trunks were partially or completely transposed as a rule, with associated septal defects and pulmonary stenosis or atresia. In the cases with venous atrium on the right, the P wave was generally upright in lead I and there were often two superior vena cavae, the left generally entering the right atrium through the coronary sinus, aortic arch on either side, partial or complete transposition of aorta and pulmonary artery, atrial and ventricular septal defects, and, generally, pulmonary stenosis or atresia.

Campbell and Reynolds,4 in a study of the significance of the direction of the P wave in dextrocardia or isolated levocardia, con-
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Figure 3

cluded that in isolated levocardia the P wave in lead I was always inverted when the venous atrium and the superior vena cava had been transposed with the abdominal viscera. They noted, however, that the pattern was found less constantly in isolated levocardia than the opposite pattern is in isolated dextrocardia. Also, upright P₁ and inverted P₂ and P₃ was a common pattern in isolated levocardia (this was found in our case, during the second admission, associated with bradycardia). Campbell and Reynolds⁴ thought that this might have been due to the sinoatrial node lying low in the atrium adjacent to the coronary sinus.

Aguilar⁵ described a similar case in which there were right axis deviation, peaked P₁ and P₂, an x-ray suggestive of absent right ventricle, and oligemic lung fields. At autopsy there were A-V communis, absent pulmonary artery and valve, truncus arteriosus, multiple interatrial septal defects, anomalous systemic and pulmonary venous return, right aortic arch, partial situs inversus, and agenesis of the spleen.

In the present case the transposition of the atria was beyond question. Of the great vessels one was entirely absent and the single functioning vessel that gave rise to the pulmonary arteries must be termed a truncus arteriosus.

Figure 4
Heart showing right atrium and right ventricle. 1-15. As in figure 3. 16. Tricuspid valve.

Figure 2
Serial electrocardiograms. 11.19.59: Rate 150. Left axis deviation. P₁ biphasic P-R 0.12 second; T₁ inverted. P in aV_L inverted, P in aV_R upright. SV₂ + RV₂ = 70 mm. Left ventricular hypertrophy. 2.25.60: Rate 75. P₂ and P₃ inverted, P₁ upright. P-R 0.12 second. T₁ inverted. P in aV_R and P in aV_L inverted, and P in aV_L upright. 3.26.60: Rate 75. P waves follow QRS except in V₁. R-P interval 0.08 second. T₁ inverted. Electrocardiogram on 3.31.60 showed no significant change.

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The association of a diminutive right ventricle is also a feature not hitherto described. The case therefore presents an unusual combination of anomalies, i.e., transposition of the atria; but instead of a transposition of the great vessels, there was a truncus arteriosus with a diminutive right ventricle and a ventricular septal defect. It would appear that the aortopulmonary septum was not formed at all.

Forgaes\(^6\) postulated a theory for the cardiac malformations associated with abdominal situs inversus. He thought that when either the abdominal viscera or the heart was transposed, inversion of the atria was usual. The right horn of the sinus venosus grew rapidly in early embryonic life, eventually forming part of the right atrium. Abdominal venous channels shunted blood from the left side of the abdomen to the opposite side, so that the venous drainage was to the right horn of the sinus venosus via the vena hepatis communis. Reversal of the normal relationship between the heart and liver would cause a reversal of this process with the left horn of the sinus venosus receiving the venous return from the abdomen, and the inferior vena cava eventually terminating in the left atrium. He therefore postulated that the associated malformations resulted as a faulty developmental torsion of the aortopulmonary septum in an attempt to divert the venous return into the correct outflow channel.\(^6\) This explanation seems a satisfying one, as it explains the partial situs inversus seen in the present case with a total anomaly of the venous return.

**Summary and Conclusions**

A case of cyanotic congenital heart disease in a baby who died at the age of 13 months is described. The patient had deep cyanosis and a grade II systolic murmur over the base. On x-ray the lungs were oligemic, and there was a broad aorta with a diminutive right ventricle. The electrocardiogram showed left ventricular hypertrophy and inverted P\(_1\) in the first tracing, but the subsequent ones
showed inverted P₂ and P₃ with bradycardia. Partial transposition of the abdominal viscera was suspected.

Autopsy revealed partial situs inversus, transposition of the atria, a truncus arteriosus giving off the pulmonary trunk, a diminutive right ventricle, high ventricular septal defect, and two atrial septal defects. This is believed to be the only case so far described with such a combination of anomalies.

References


The Receding Horizon

Let us approach the problems before us in a humble spirit, recognizing the temporal nature of our deductions, remaining aware of the areas of unenlightenment, and leaving room for doubt. The greatest lesson that science has taught is how much more there is to learn.—Carl J. Wiggers. Introductory Remarks. Cardiovascular Effects of Nicotine and Smoking. Part I. The Absorption and Fate of Nicotine. Ann. New York Acad. Se. 90: 6, 1960.
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