CASE REPORTS

ISOLATED LÆVOCARDIA WITH SITUS INVERSUS VISCERUM

BY

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In their recent study, Campbell and Forgacs (1953) divide cases of isolated lævocardia with situs inversus viscerum into two groups. In the first, the venous atrium and the venæ cavæ are on the right side of the heart as in the normal subject; in the second group, which has been reported less frequently, the venous atrium and the venæ cavæ are left-sided. These authors reported only two necropsies concerning the second group: their own, and that of Young and Griswold (1951). We have found three further reported cases with necropsy studies on the second group, and details of a sixth are recorded here.

Case Report

A girl, 2 years old, was admitted to hospital with symptoms of intracranial abscess. She had been cyanosed since birth but showed normal physical development. The apex beat of the heart was in its normal position. X-ray examination showed evidence of transposition of the abdominal viscera, the gas bubble in the stomach lying on the right side (Fig. 1). The patient died suddenly a week after admission.

Necropsy. An abscess, 2 cm. in diameter, filled the left cerebellar lobe. The abdominal organs, and the intrathoracic structures other than the heart, were transposed. The right lung had two lobes and the left three; the aortic arch was right-sided and the azygos arch left-sided. The circulatory arrangement is summarized in Fig. 3.

The left-sided, venous atrium showed the usual pattern of crista terminalis and muscoli pectinati, and the sinu-atrial node was in normal relationship to the crista. No coronary sinus could be found. The only visible cardiac vein crossed the posterior atrioventricular groove to enter the venous atrium. The right-sided arterial atrium received a persistent right superior vena cava in addition to the four pulmonary veins. The left atrioventricular valve was tricuspid and the right was bicuspid. The corpus of the left ventricle was the same size as that of the right: its conus, however, was absent, the cavity of this chamber being bounded above by a large crista supraventricularis. The only exit was through a ventricular septal defect that measured 7×5 mm. into the right ventricle; and the septal cusp of the tricuspid valve united through the foramen with the anterior mitral cusp. Both arterial trunks arose from the right ventricle (Fig. 2). The aortic valve had three cusps; the valve at the stenotic pulmonary orifice had two thick cusps. A patent ductus arteriosus joined the right branch of the pulmonary artery.

Discussion

Campbell and Forgacs' (1953) case is very similar, but in our case there was a right superior vena cava which entered the arterial atrium at the point where the oblique vein of the atrium normally commences. The absence of a coronary sinus suggests that the left horn of the sinus venosus was absorbed into the common atrium during development. McCotter (1916) has described an earlier phase in the absorption process, with a persistent caval vein entering a coronary sinus which communicated freely with both atria.

Comparison of the present case with the five others alluded to, reveals that the major differences between them concern their outflow channels; these are depicted in Fig. 4. These variations evidently arise from the manner in which the aortico-pulmonary septum has been laid down. In the hearts described by Hickman (1869) and Young and Griswold (1951), torsion of the septum seems to have been normal. In that recorded by Taussig (1947), failure of torsion has led to uncorrected transposition of the great vessels. The heart

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FIG. 1.—Chest X-ray, showing gas bubble in the stomach on the right side, and lævocardia.

FIG. 2.—Photograph of the interior of the right ventricle. C.S., crista supraventricularis (right extremity). I.V.F., interventricular foramen. P.O., pulmonary orifice. S.V.C., superior venæ cavæ.

here described links these three with those of Campbell and Forgacs (1953) and Muscovitz *et al.* (1952) in which the septum has undergone counter-torsion, returning the pulmonary trunk in the last case to full communication with its own ventricle.

The coronary artery in the case of Muscovitz *et al.* was single. They postulate detorsion followed by countertorsion of the aorticopulmonary septum to account for the distribution of the artery. The other cases quoted here showed no evidence of any such migration of the septum. The coronary cusps in the present case were some 290° out of alignment with the normal (Fig. 4), but the distribution of the two vessels showed no unusual features.

Summary

Necropsy findings in a case of isolated lævocardia with situs inversus viscerum are reported. The heart showed transposition of its chambers. Comparison with the findings in five similar cases indicates a great variability in the disposition of the aorticopulmonary septum in this condition.

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FIG. 3.—Diagram of the circulation.

FIG. 4.—Diagrams of the outflow channels of the six autopsied cases of isolated lævocardia with transposition of cardiac chambers. The smaller vessel in each is the pulmonary artery. The arrows indicate the disposition of the aorticopulmonary septum. The interventricular septum in the last case (Muscovitz *et al.*) was rudimentary.

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