Relationship of the Azygos Venous System to Normal Atrial Situs in Levocardia

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RESULTS of clinical evaluation of patients with dextrocardia or levocardia associated with situs inversus are often confusing. Recently Van Praagh, Van Praagh, and Keith correlated revealingly the embryologic folding of the primitive heart tube, the position of the bulbus cordis, and the septation of the arterial trunk with the clinical relationships of the aorta and the right ventricle in a large number of patients with dextrocardia. Their emphasis of the developmental events as a basis for understanding the anatomic variations provides much insight into the study of patients with levocardia (this term also implying abdominal visceral situs inversus.) Their findings may be combined helpfully with the knowledge that rightward or leftward position of the liver in the abdomen determines, in most instances, the side on which the inferior vena cava will develop. The situs of the inferior vena cava in turn seems to determine the position of the right atrium. Thus, in the patient with abdominal situs inversus and mirror-image dextrocardia, the functional right atrium is positioned to the left. In most instances of abdominal visceral situs inversus associated with levocardia, the functional and anatomic right atrium is positioned to the left, following the position of the liver. The majority of these patients with levocardia are known to have complex intracardiac anomalies.

We would like to call attention to a developmental peculiarity that occurred in two patients with levocardia and resulted in simple intracardiac defects that were surgically corrected. Both of these patients, one an adult...
woman (fig. 1), the other a child of 2 years (fig. 2), had abdominal situs inversus, absence of the inferior vena cava with the venous drainage from the lower part of the body returning by way of an enlarged azygos system which in turn emptied into the superior vena cava at its usual position. In addition, both of these patients had large atrial septal defects, but with definite atrial septation. In both patients surgical correction was accomplished without difficulty and no additional intracardiac malformation was identified. In the adult patient the electrical axis of the P wave was directed from right to left at approximately 0°, indicating that the atrial pacemaker was near the usual position and suggesting that the right atrium was indeed the rightwardly placed atrium. In the child, there was a wandering pacemaker making conclusions difficult, but the electrocardiogram suggested the origin of atrial excitation to be in the rightward atrium (figs. 3 and 4). In both patients examination of the interior of the right atrium at the time of surgical exploration disclosed no inferior vena cava; however, there was a small orifice that apparently led only into the hepatic veins. Neither patient had desaturation of the systemic arterial hemoglobin postoperatively. The exact relationships of the hepatic veins to the atrium were not demonstrated angiographically, however.

Radiographically, in the adult patient, a prominent shadow was apparent just above the right atrial silhouette along the superior
Figure 4
Electrocardiogram of the child. Note the P waves in leads III, aVp, and aVF.

Figure 5
Radiograph of the chest of the adult. Note position of the stomach bubble.

Figure 6
A radiograph of the adult showing the position of the opacified aorta. Note shadow of the azygos vein along the superior vena cava.
vena cava (figs. 5 and 6). At the time of surgical repair, this was identified as the greatly dilated azygos vein.

The development of this unusually large azygos system in place of an inferior vena cava is apparently the only exception to the clinical rule that the situs of the right atrium follows the situs of the liver. Further, when the azygos system does allow the usual rightward position of the systemic venous atrium in a patient with levocardia, the associated intracardiac defects may be simple and surgically remedial as in these two patients. In fact, absence of the inferior vena cava has been described in association with a normal heart. While we have not studied a similar patient with isolated dextrocardia and normal abdominal visceral situs, the presence of a well-developed left azygos or hemiazygos system would suggest that the atria may be in “mirror-image” position.

Summary

Levocardia with abdominal visceral situs inversus is associated with complex intracardiac malformation and most often with inversion of the atria.

In patients with levocardia, if the azygos venous system replaces the inferior vena cava as the principal or only avenue of venous return from below the diaphragm, the position of the right and left atria may be normal.

Patients with levocardia associated with simple ostium secundum atrial septal defects that were surgically corrected are briefly presented.

References

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