Hepato-Atrial Dissociation
Report of Two Cases and Clinical Implications*

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Two clinical cases are reported illustrating the rare condition in which the liver does not follow the situs of the right atrium. Both patients had right-sided liver and cardiac apex, a condition usually associated with severe intracardiac defects. Both proved to have mirror-image dextrocardia with normal intracardiac anatomy or correctable lesions (one case). The behavior of the inferior vena cava in these cases is illustrated. Embryologic considerations are presented concerning the hepato-cavo-atrial concordance rule.

In clinical cardiology, when abnormalities of location of the cardiac cavities are suspected, atrial situs (position of the morphologic atria) may most often be predicted by knowledge of the liver situs. When the major lobe of the liver is the right one, atrial situs is predicted to be normal. Conversely, when the left hepatic lobe is dominant, the atria are expected to be inverted.1 Another useful generalization is the association of complex intracardiac malformations with situs solitus dextroversion2 or situs inversus levoversion.2,3 The present report describes two patients in whom the atrial situs did not correspond to the liver situs. Both patients had either normal cardiac physiology and anatomy or correctable lesions, even though the clinical diagnosis of dextroversion had suggested complicated malformations nonamenable to surgical correction.

CASE REPORTS

CASE 1

This female patient was studied at the age of five years, because of a history of heart murmur. On physical examination she appeared in good general health, without cyanosis or any sign of congestive heart failure. The apex was found in the fifth right intercostal space at the mid clavicular line. A grade 3/6 holosystolic murmur was best heard at the apex and a separate ejection murmur, grade 4/6, was present in the second right intercostal space, radiating to the back. A single loud second sound was heard. The liver was palpated below the right costal arcade and was of normal size. Results of hematologic tests were normal; no Howell-Jolly bodies were seen. The x-ray films of the chest confirmed that the apex of the heart was in the right chest. There was mild cardiomegaly, without selective chamber enlargement. The pulmonary vascular markings were normal. The aortic knob was on the left side. Barium swallow confirmed the clinical impression of a normally-placed stomach, while the liver was seen in a normal right-sited position (Fig 1). In the electrocardiogram, the P wave had an axis of +110° and was notched, findings consistent with left atrial enlargement in situs atrialis inversus. There was first degree AV block (P-R = 0.20 s, for a heart rate of 120/min). The mean QRS axis was +180°. There were no initial Q waves in the right precordium. In mirror image dextrocardia, these electrocardiographic findings are consistent with ventricular inversion (Fig 2).

The catheterization data are presented in Table 1. During the test the patient had a junctional rhythm with retrograde P waves which followed the QRS. The venous catheter inserted in the right basilic vein reached the heart to the right of the spine and followed an oblique course reaching a venous atrium on the left. It was possible to catheterize the inferior vena cava, positioned to the left of the spine, and also to enter a rightwardly located arterial atrium. The right-sided ventricle was reached through the arterial atrium. Following injection of contrast material into the inferior vena cava (IVC) a group of suprahepatic veins were seen to fill directly from the IVC at its entrance into the atrial cavity (Fig 3). After injection of contrast material into the left-sided ventricle, a morphologic left ventricle was visualized. A ventricular septal defect was present through which the morphologic right ventricle was filled to the right (Fig 4). The great vessels had abnormal parallel courses, with an anterior and right-sided aorta and a posterior and left-sided pulmonary artery. The aortic arch

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1 The terms "dextroversion" and "levoversion" are used in this paper to mean abnormal positions of the apex for a given situs atrialis.1

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HEPATO-atrial DISSOCIATION

Table 1—Hemodynamic Data of Case 1.

<table>
<thead>
<tr>
<th>Location</th>
<th>% O₂ Sat</th>
<th>Pressure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inferior vena cava</td>
<td>74</td>
<td></td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>72</td>
<td></td>
</tr>
<tr>
<td>Right atrium (high)</td>
<td>73</td>
<td>‘a’ wave 25* mean 8</td>
</tr>
<tr>
<td>(left-sided) (mid)</td>
<td>85</td>
<td></td>
</tr>
<tr>
<td>(low)</td>
<td>73</td>
<td></td>
</tr>
<tr>
<td>Right ventricle</td>
<td>87</td>
<td>85/0-5</td>
</tr>
<tr>
<td>(right-sided)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>88</td>
<td>24/6 (11)</td>
</tr>
<tr>
<td>Left atrium</td>
<td>93</td>
<td>‘a’ wave 25* mean 8</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>87</td>
<td>85/0-5</td>
</tr>
<tr>
<td>Right femoral artery</td>
<td>86</td>
<td>95/55 (75)</td>
</tr>
</tbody>
</table>

*During junctional rhythm, with P waves following the QR complex.

situs solitus of liver and stomach; mirror-image dextrocardia, with corrected transposition; persistent right superior vena cava draining into the coronary sinus; absent left superior vena cava; discordance between the situs of the liver and that of the inferior vena cava; atrial septal defect (ASD), possibly secundum type; ventricular septal defect (VSD); and pulmonic valvular stenosis (Fig 5).

This patient was advised to have surgical treatment (ASD and VSD repair, with pulmonary valve commissurotomy) at a future time, when symptoms appear.

Case 2

This patient was studied when she was 27 years old because of abnormal cardiac findings, although she was asymptomatic. On physical examination the liver was palpated below the right costal margin, and the cardiac apex was in the fifth right intercostal space at the mid clavicular line. The heart sounds were normal, and no murmur was heard. There were no signs of congestive heart failure nor

Figure 1. (Case 1) Heart is mildly enlarged. Apex is in right chest. Aortic knob is on left side of spine. Pulmonary vascular markings are within normal limits. Liver is on right side and stomach on left.

Figure 2. (Case 1) ECG shows first degree AV block (P-R = 0.20 sec). P waves are consistent with left atrial enlargement in situs inversus. Precordial leads suggest right-position of apex. Extreme right precordial leads are not available, so that no definitive diagnosis can be made of situation of ventricles.

Figure 3. (Case 1) Venous catheter was passed through right superior vena cava draining into coronary sinus, to right atrium (left-sided) and inferior vena cava, which lies on left side of spine. few suprarepatic veins are visualized on left side of IVC, at entrance into right atrium.

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cyanosis. No Howell-Jolly bodies were found. The x-ray films of the chest showed a predominantly right-sided liver. The stomach was on the left side. The heart was essentially in the right chest, having normal size. The aortic knob was on the right. The pulmonary vascular markings were normal (Fig 6). Sinus rhythm was seen in the ECG. The shape of the P waves and QRS complexes were consistent with mirror-image dextrocardia and normally situated ventricles in situs inversus (Fig 7).

Cardiac catheterization was performed, with the venous catheter introduced into the right basilic vein. The catheter passed through a right superior vena cava (without innominate vein) and the coronary sinus into a left-sided venous atrium. The right ventricle and both pulmonary arteries were entered from this atrium. Because of normal pressures and oxygen saturations in the pulmonary artery and in a systemic artery, "left heart" catheterization was not done (Table 2).

The angiographic study confirmed the abnormalities previously suspected by catheter course and proved the absence of septal defects. The inferior vena cava was found to have a peculiar course. The suprahepatic segment, after penetrating the diaphragm on the right side, abruptly turned to the left.
and entered the left-sided right atrium (Fig 8). At the level of the diaphragm there was a localized stricture (without pressure gradient) below which the IVC had the usual course for a right-sided inferior vena cava. A group of hepatic veins drained into the inferior vena cava just proximal to the stenosis. A selective injection in one of these veins filled an intrahepatic channel which connected with veins of the left lobe and then drained directly into the left-sided right atrium (Fig 9). The catheterization of this vein from the right atrium was not achieved. After opacifying a normal RV and the pulmonary vascular bed, the contrast material filled a normal left atrium and ventricle (Fig 10). The great vessels were seen to be normally related for a mirror-image dextrocardia.

**Final Diagnosis**

The final diagnosis was: situs inversus of atria and lungs; situs solitus of liver and stomach; persistent right superior vena cava draining into the coronary sinus; atrophic innominate vein; right-sided infradiaphragmatic portion of the inferior vena cava; direct connection of the left hepatic veins to right atrium; uncomplicated mirror-image dextrocardia (Fig 11).

**Discussion**

In the clinical analysis of complex malformations of the heart with abnormal position of the cavities, the position of the atria is the first point that needs to be established. This is because the atria are the reference pattern for the whole heart. As known, the situs of the atria is decided very early in embryogenesis. At the time when right and left cardiogenic areas are determined, the atrial situs is decided. In fact, when the heart is eventually formed from the fusion of two symmetrical tubes, the atrial level is the only one where the right and left tubes maintain their original individualities. This is also the reason why the term "atrial inversion," used to classify cases with "normal" position of the ventricles but inversion of the atria, is not proper: situs atrialis inversus with atrioventricular discordance or ventricular inversion is more correct terminology.

It is a common observation that there is a consistent relationship between the atrial and visceral situs, the rule being that the right atrium is homolateral to the larger lobe of the liver and the trilobar lung, opposite to the stomach and the
spleen. In clinical cardiology, this has been applied as the "hepato-atrial" concordance rule because the liver is the only one of these visceral organs in which laterality is easily recognized by clinical means.

There are rare exceptions to this rule, originating essentially from two situations: (first) heterotaxic syndromes, where the body laterality is lost, in which case the liver is usually symmetrical and there is a common atrium; and (second) the cases in which there is a primary "error" in the concordance of the right atrium and the liver.

The cases presented in this report are examples of the second event. In both the anatomic right atrium was to the left, even when the major lobe of the liver was to the right.

The laterality of the inferior vena cava is an interesting point in these circumstances. The most significant portion of this vein, from an embryologic viewpoint, is the suprahepatic segment. The suprahepatic portion of the inferior vena cava develops from one of the two hepatocardiocar channels (om-phalomesenteric veins or vitelline veins). Usually the right persists when the situs atrialis is solitus and the left when the situs atrialis is inversus. The caudal portion of the inferior vena cava is embryogenetically unrelated to the suprahepatic portion, and various anomalies are possible. One of the more frequent is inferior vena cava continuation into theazygos or hemiazygos veins. In this anomaly the hepatic portion of the IVC is absent, and the suprahepatic drains only the hepatic veins directly into the anatomic right atrium. The so-called dissociation of the inferior vena cava from the right atrium should refer only to the caudal portion of the inferior vena cava since the suprahepatic IVC always connects to the anatomic right atrium.

For clinical purposes it can be concluded that localization of the anatomic right atrium may be easily accomplished by demonstration of the drainage of the suprahepatic veins with or without the caudal inferior vena cava.

We are not aware of any true exception to this rule, if cases of heterotaxia are excluded. Of the four cases described in the literature as "drainage of the inferior vena cava into the left atrium," only two had autopsy study. These two are consistent with overgrowth of the right valve of the sinus venosus which anomalously joined the septum secundum, leading to secondary functional drainage into the left atrium. This anatomic arrangement is not an exception to the rule that the suprahepatic portion of the inferior vena cava is always connected with the anatomic right atrium.

Another common observation is that the persisting hepatocardiac channel is usually homolateral with the major lobe of the liver. In cases of hepatocordial discordance the infrahepatic portion of the inferior vena cava may be connected with the azygos or hemiazygos systems, or to the suprahepatic portion. In the latter case it may be situated on the same side as the anatomic right atrium and opposite to the major lobe of the liver (as in our Case 1) or it may course on the opposite side (homolateral with the major hepatic lobe). When homolateral to the liver, it crosses the midline at the diaphragmatic level, as in our Case 2 and in the three cases of Hasterreiter and colleagues. In this last type there is usually a persisting contralateral hepatocardioc channel of varying size, which joins the right atrium independently or connects with the horizontally crossing segment of the inferior vena cava just below its junction with the right atrium. It is our impression that also in our Case 1 a right-sided hepatocardioc channel should have persisted, but this was not demonstrated by catheter studies.
These rare cases are the exception which confirms the general rule that the major lobe of the liver is homolateral to the hepatic drainage. This generalization suggests an embryologic hypothesis, which could explain the usual concordance in laterality of the right atrium and the major lobe of the liver.

Until the second week of life, the human embryo has a straight heart tube, at the caudal extremity of which the sinus venous is connected. At this stage, two symmetrical omphalomesenteric veins drain blood from the yolk-sac to the sinus venosus, in close relation to the liver, which is developing in the septum transversum.

About four weeks later,16 the primitive atrium begins to differentiate into its sections. According to its situs, the anatomic left side receives connections from the pulmonary veins, while the sinus venosus is gradually absorbed into the posterior wall of the anatomic right atrium. At this stage the venous circulation, previously symmetrical, acquires asymmetry. One of the changes that takes place is the reabsorption of the omphalomesenteric vein contralateral to the anatomic right atrium and the enlargement of its homolateral counterpart,17 probably for hemodynamic reasons. At the same time the liver abandons its original symmetry, with enlargement of the lobe homolateral to the persisting omphalomesenteric vein.

This sequence of events, if experimentally proved, would relate atrial situs to the most significant asymmetrical abdominal organ (the liver) and would support the thesis that the organ, which rules the laterality in visceral asymmetry, is the heart at its atrial level.

A final comment should be made about the finding of a completely normal heart, even when the apex is directed to the same side as the major lobe of the liver (Case 2). It is common observation that in the presence of "dextroversion" the heart is most frequently severely deformed.23 Since this condition is characterized by a right-sided liver and a right-sided apex, our patient would have been expected to have intracardiac defects, as would the cases presented by Hastreiter in "levversion."11

Due to the hepatocardial dissociation, our patient did not have "dextroversion," but mirror-image dextrocardia, as correctly suspected from the electrocardiogram. Two of Hastreiter's cases11 were the mirror-image of those reported here as were the two cases previously reported by one of us (R.D.L.).10 A third case11 was analogous to ours.

Our first patient had a complex malformation, but it was compatible with normal physiology (situs atrialis inversus with corrected transposition and ventricular inversion). This was complicated by septal and valvar defects, all correctable by surgical treatment.

A recent example of hepatocardial dissociation was reported by Clarkson and colleagues.8 In this case the hepatocaval relationship corresponded exactly to our Case 2, but atrioventricular discordance was associated with normally related great vessels, an anatomic complex resulting in "functional transposition."

In consideration of the few cases reported, it is difficult to conclude that with hepatocardial dissociation the heart will be normal.11 From our cases and those published in the literature it appears that there may be a significant incidence of atrioventricular discordance. Clinically, it is important to recognize that "dextroversion" and levoversion are not necessarily associated with complex intracardiac defects. Indeed, demonstration of an anomalous position of the suprahepatic portion of the inferior vena cava with respect to the liver may confirm the electrocardiographic suspicion that there is a normal heart or mirror-image dextrocardia.

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The Art of Ballet

Ballet, a word which has its roots in the Italian "ballare" (to dance), had its beginnings in Italy almost five centuries ago. When Catherine de Medici became the Queen of France, she brought to the French court the Italian's fondness for dancing and dance entertainments. By her command, on October 15, 1581, the Ballet Comique de la Reine, usually considered the first real ballet, was given. Here, in fact, was a new concept of dance theater, for these were not isolated divertissements but, rather, a theater work with a continuously developed theme, with choreographic disciplines making it a dramatic entity. An immensely important contribution to the development of ballet was to be found in a book called Orchégraphie and published in 1588. Its author was a priest, Thoinot Arbeau, and the book described the dances, along with musical notes, popular during that century. Here, the turn-out of the legs and feet was recorded, and this, of course, heralded the establishment, a century later, of the five positions of the feet, the absolute technical base of ballet itself. In the year of 1661 Louis XIV, King of France, commanded that the Académie Royale de la Danse be established in Paris which in 1661 produced the world's first professional ballerina, Lafontaine. The ballerinas, of course, were not by any means solely responsible for the technical development of ballet. The danceurs most certainly added to ballet's technique. But the ballet masters, the choreographers, even the ballet theoreticians, were the ones whose vision required more and more of the dancers.

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