Radionuclide Angiocardiography in the Clinical Evaluation of Cardiac Malpositions in Situs Solitus in Adults

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A right-sided position of the heart in the chest in situs solitus is an abnormal feature easily discernible from a plain chest radiograph. This cardiac malposition may be due to cardiac displacement (dextroposition), which is usually a feature of lung disease, or a structural abnormality of the heart (dextrocardia). Because each condition has different clinical pathologic implications, it is important to distinguish them. Chest films, however, often provide no conclusive information. We performed radionuclide angiocardiography (RNA) in six adults with a cardiac malposition in situs solitus. It was found that morphologic data obtained from the serial images may distinguish dextroposition from dextrocardia. In addition, these images permitted us to diagnose congenitally corrected transposition, a cardiac anomaly which occurs with increased frequency in situs solitus with dextrocardia. Quantitative shunt detection performed during this procedure is helpful in the differential diagnosis of dextroposition and able to distinguish uncomplicated dextrocardia from dextrocardia associated with other cardiac abnormalities. RNA therefore is a valuable and easily performed method in the analysis of cardiac malpositions in adults.


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Situs solitus implicates a normal location of both atria and abdominal organs. In this situation, the greatest portion of the heart is normally located in the left side of the chest. Sometimes, however, the cardiac silhouette is found to be predominantly in the right hemithorax. In situs solitus this may be principally due to two possible causes: (a) displacement of the mediastinum and cardiac structures to the right (dextroposition), and (b) abnormal rotation of the embryonic cardiac loop resulting in a right-sided location of the cardiac apex (dextrocardia) (1–4).

These cardiac malpositions in situs solitus are usually detected on plain chest films. However, radiologically it may be difficult to determine if the malposition is primarily due to displacement or an intrinsic cardiac abnormality. It is important to distinguish these conditions because each may be associated with a different spectrum of malformations (2–8).

We performed radionuclide angiocardiography (RNA) in six adults. Five patients presented with nonspecific cardiac murmur, one patient with slight dyspnea on exertion. Chest radiographs routinely obtained as part of the initial clinical workup, showed a cardiac malposition in situs solitus in all these patients. Sequential imaging of the central cardiovascular system by RNA allows a morphologic study of the different cardiac compartments and great vessels (9,10) (Fig. 1).

In our studies, we found that RNA is a useful technique in the first analysis of a cardiac malposition in situs solitus offering scintigraphic differentiation between dextroposition and dextrocardia. Additional hemodynamic findings, such as shunts, also diagnosed by RNA, may influence the clinical management of these patients. Moreover, congenitally corrected transposition (CCT), a complex congenital heart disease, that is known to occur in >50% of patients with situs solitus and dextrocardia (5,11), can be correctly diag-

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nosed. Scintigraphic findings in our patients, all confirmed by other techniques, are presented and discussed.

MATERIALS AND METHODS

Studies were carried out with a large field-of-view gamma camera, equipped with a low-energy, all purpose collimator. Data are stored in a 64 x 64 matrix on disk in an MDS-A\(^2\) dedicated nuclear medicine computer system. Patients were examined in supine position. A bolus injection of 7.5 mCi (277.5 MBq) of technetium-99m (\(^{99m}\)Tc) citrate\(^1\) was administered in the basilic vein of the right arm. This radiopharmaceutical was chosen as it is rapidly eliminated by the kidneys, thus minimizing radiation dose (\(12\)). Anterior images were obtained with an exposure of one image/sec during 30 sec, and were immediately recorded on transparent nuclear medicine film after the injection. The adequacy of the bolus injection was subjectively judged on the time-activity curve derived from a region of interest over the superior vena cava. The average count rate in one image amounts to about 3,500 cps over the first ten images of the study.

Shunt quantitation occurred according to the method described by Maltz and Treves (\(13\)) with a computer program provided by the supplier. Review of the hemodynamic pattern took place by data retrieval from the disk. A detailed study of the scintigraphic morphologic findings was performed.

RESULTS

Chest film and scintigraphic findings are summarized in Table 1. The ultimate diagnosis was established by operation (Case 5) and by other techniques including cardiac catheterization (Cases 2, 5, and 6), cardiac two-dimensional real-time ultrasonography (Cases 1, 3, and 4), and bronchoscopy (Case 1). The values of the shunts in Cases 2 and 5 established by catheterization correlated well with the results of the scintigraphic studies.

Cases 1 and 2

The chest films of Cases 1 and 2 are shown in Figs. 2A and 3A. Figure 2B demonstrates the scintigraphic findings of Case 1, Fig. 3B of Case 2. The right atrial-ventricular complex projects to the right of the superior vena cava (SVC), which is in normal position. The long axis of the left ventricle has a normal direction (right cranial to left caudal) and the apex points to the left. The anterior view clearly demonstrates that the descending aorta crosses the left ventricle at the apex, while under normal conditions this point is found to

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Sex</th>
<th>Age (yr)</th>
<th>Presumptive chest film diagnosis</th>
<th>Morphologic scintigraphic findings</th>
<th>Left-to-right shunt</th>
<th>Presumptive scintigraphic diagnosis</th>
<th>Ultimate diagnosis</th>
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<tr>
<td>1</td>
<td>F</td>
<td>29</td>
<td>HLS(^*)</td>
<td>CD(^{**})</td>
<td>–</td>
<td>HRL(^1)</td>
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<tr>
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<td>F</td>
<td>52</td>
<td>HLS</td>
<td>CD</td>
<td>+</td>
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</tr>
<tr>
<td>3</td>
<td>F</td>
<td>14</td>
<td>NC</td>
<td>D(^{1})</td>
<td>–</td>
<td>D</td>
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<tr>
<td>4</td>
<td>M</td>
<td>35</td>
<td>NC</td>
<td>D</td>
<td>–</td>
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<td>D</td>
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<tr>
<td>5</td>
<td>M</td>
<td>26</td>
<td>HLS</td>
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<td>+</td>
<td>D, ASD(^3)</td>
<td>APVR to SVC and IVC</td>
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<td>6</td>
<td>M</td>
<td>32</td>
<td>NC(^{11})</td>
<td>CCT</td>
<td>–</td>
<td>CCT(^{**})</td>
<td>Isolated CCT</td>
</tr>
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</table>

\(^*\) HLS = Hypogenetic lung syndrome.
\(^1\) HRL = Hypoplasia of right lung or agenesis of a right lobe.
\(^2\) APVR = Abnormal pulmonary venous return.
\(^3\) ASD = Atrial septal defect.
\(^{**}\) CD = Cardiac displacement.
\(^{1}\) D = Dextrocardia.
\(^{11}\) NC = Not conclusive (differential diagnosis of HLS, HRL, D, and CCT was given).
\(^{**}\) CCT = Congenitally corrected transposition.
are consistent with situs solitus and dextrocardia. In Cases 3 and 4 no shunt was detected. In Case 5 a left-to-right shunt was found to be present (QP/QS = 2.10).

**Case 6**

Figure 6A shows the chest film of this patient. Figure 6B demonstrates the scintigraphic findings. The right-sided atrial-ventricular complex has an oval shape. The pulmonary trunk is central in position. The left-sided ventricle also has an oval configuration and its axis is vertically oriented. The aorta ascends on the left and its downward course is almost vertical, not crossing the

**FIGURE 2**

A: Case 1. PA chest film, showing right-sided position of heart. B: Case 1. Scintigraphic images of right and left side of heart, showing normal direction of long axis of left ventricle. Dextroposition. Compare anatomic relation of descending aorta and left ventricular apex with Fig. 1

be at the middle third part of the left heart chamber (Fig. 1).

These observations are consistent with cardiac displacement to the right. In Case 1 no shunt was detected. In Case 2 a left-to-right shunt was present (QP/QS = 1.60).

**Cases 3, 4, and 5**

The chest films of Cases 4 and 5 are shown in Figs. 4A and 5A. Figure 4B demonstrates the scintigraphic findings of Case 4. Fig. 5B of Case 5.

The right atrial-ventricular complex projects to the right of SVC. The long axis of the left ventricle has an abnormal direction (left-cranial to right-caudal), the apex points to the right and is located to the right of the midline. The relations and locations of the great arterial vessels are normal. These scintigraphic findings

**FIGURE 3**

A: Case 2. PA chest film, showing right-sided position of heart. Scimitar vein is demonstrated at base of right lung. B: Case 2. Scintigraphic image shows normal direction of left ventricular long axis. Left-to-right shunt, dextroposition
midline. The ventricular septum is perpendicular to the plane of imaging indicated by the side-by-side relation of the ventricles. A left-to-right shunt could not be detected. The scintigraphic findings demonstrate the typical characteristics of CCT.

**DISCUSSION**

A right-sided position of the heart in the chest of a patient with situs solitus, is an abnormal feature easily discernible from a plain chest radiograph. In this situation, it is important to determine if the cardiac malposition is due to displacement (dextroposition), or the result of a structural abnormality of the heart (dextrocardia) (1,2).

*Dextroposition* usually results from reduction of the volume of the right lung (1,2). In most patients, this is due to acquired lung disease (such as atelectasis, fibrosis, or operation), that will often be apparent from the chest film and clinical history of the patient. Reduction of the right lung volume with consequent dextroposition, however, may also be due to congenital hypoplasia of this lung or agenesis of a lobe (2). This may be an isolated uncomplicated disorder. Hypoplasia of the right lung with consequent cardiac displacement to the right, however, may also be part of the hypogenetic lung syndrome. This always includes partial anomalous pulmonary venous return to the inferior vena cava (IVC), resulting in a left to right shunt. Bronchial abnormalities are virtually always present (7,8,14). This syndrome, also known as the "scimitar syndrome," is associated with an increased incidence of cardiovascular disease of which the secundum type of atrial septal defect is the most frequent (7,8).

*Dextrocardia* in situs solitus is an intrinsic cardiac abnormality, due to a rotation disorder of the embryonic cardiac loop (2). In this situation, associated cardiac malformations are frequently present and often severe (2,5,6,11). Patients with this cardiac malposition therefore usually will present in early childhood. Adult patients with dextrocardia and situs solitus may have a structurally normal heart, but there is an increased incidence of left-to-right shunts and CCT, which justifies a careful analysis (2,4–6,11). In CCT, the right atrium communicates with the morphologic left ventricle located on the right side, while the transposed pulmonary trunk arises from this ventricle. The left atrium communicates with the morphologic right ventricle, located on the left side. The ascending aorta originates from this ventricle, coursing left and anterior to the pulmonary trunk (15).

Dextrocardia and dextroposition in situs solitus are differentiated by analyzing the direction of the base-apex axis of the heart (1). Dextroposition is characterized by a normal direction of the base-apex axis, oriented to the left and inferior. In dextrocardia, the base-apex axis points to the right and inferior (1–3). Determination of the direction of the base-apex axis may be difficult from plain chest films as illustrated in our cases.

In four of six cases, chest radiography was inconclusive or misinterpreted as to the direction of this axis, leading to a false diagnosis.

In our six patients, sequential imaging of the central cardiovascular circulation by RNA provided sufficient morphologic information to determine the direction of this axis, offering scintigraphic differentiation between dextroposition and dextrocardia.
Once a diagnosis of dextrocardia or dextroposition in situs solitus is made by RNA, one has to distinguish the uncomplicated disorders from those with associated abnormalities. In this respect, the detection of a left-to-right shunt and the demonstration of an abnormal relation of the great arterial vessels, are crucial. These features can be evaluated during the same study.

A scintigraphic diagnosis of dextroposition without a shunt implicates uncomplicated hypoplasia of the right lung, that generally necessitates no further clinical attention (Case 1). Dextroposition in combination with a left-to-right shunt, however, favors a diagnosis of the hypogenetic lung syndrome. The shunt is due to anomalous pulmonary venous return to the IVC (scimitar vein) and may be aggravated by an associated atrial septal defect, that is frequently present in this syndrome (Case 2). The magnitude of this shunt will influence further clinical management.

A scintigraphic diagnosis of dextrocardia without a shunt and normal related great arterial vessels in an adult excludes major associated cardiac abnormalities (Cases 3 and 4). Other than the rotation disorder, these patients are considered to have normal hearts.

The demonstration of a shunt in dextrocardia indicates an associated cardiac abnormality such as an atrial septal defect, ventricular septal defect, partial anomalous pulmonary venous return or a combination of these, requiring further analysis (Case 5).

A careful morphologic study of the images is indicated to exclude CCT. Constant scintigraphic features of this congenital heart disease are the left ascending aorta, central position of the pulmonary trunk and perpendicular position of the ventricular septum in the anterior projection, due to the side-by-side relation of both ventricles. When these features are encountered in a patient who presents in adulthood, this is virtually pathognomonic of CCT (16,17) (Case 6). Regular follow-up of these patients should be considered, because abnormalities of the conduction tissues and left atrioventricular valve may lead to symptoms in many cases (14,18).

In conclusion, it is shown that RNA provides suffi-
consider RNA a valuable and easily performed method to evaluate patients with a cardiac malposition.

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FOOTNOTES

* Toshiba Medical Systems Europe, (Toshiba 40A) Delft, The Netherlands.
  † Solco-citrane, Solco, Basel, Switzerland.

REFERENCES

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FIGURE 6

A: Case 6. PA chest film shows right-sided heart and situs solitus. B: Case 6. Scintigraphic images of right and left side of heart, showing typical characteristics of CCT (central position of pulmonary trunk, left ascending aorta, and side-by-side relationship of both ventricles)