Left Juxtaposed Atrial Appendages: Diagnostic Two-Dimensional Echocardiographic Features

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Left juxtaposition of the atrial appendages is a rare congenital malformation usually associated with cyanotic congenital heart disease. Recognition of this problem in complex congenital heart disease is important for complete anticipation of diagnostic or surgical management of these patients. The diagnosis of juxtaposed atrial appendages can only be suspected by certain nonspecific plain film roentgenographic (1-3) and electrocardiographic (4) findings. To date, angiography has been the only reliable means of preoperative recognition (3,5-7). However, the diagnosis is more commonly made incidentally at the time of surgery or autopsy (5,8-18).

In this report, we describe the noninvasive recognition of left juxtaposition of the atrial appendages by means of two-dimensional echocardiography. The diagnostic features are described and correlated with angiographic, surgical and autopsy observations. These observations suggest that two-dimensional echocardiography allows reliable noninvasive diagnosis of left juxtaposition of the atrial appendages.

Methods

Study patients. Ten patients (seven male and three female), aged 7 months to 17 years, with left juxtaposition of the atrial appendage are the subjects of this report (Table 1). In four patients, a prospective diagnosis of this condition was made by two-dimensional echocardiography. Two-dimensional echocardiograms were assessed retrospectively in the remaining six patients. Five patients were known to have left juxtaposed atrial appendages on the basis of other studies (angiography, four patients, and surgery, one patient). In one patient, the echocardiogram was reviewed after autopsy findings revealed juxtaposed atrial appendages. In all but one patient, the diagnosis was ultimately confirmed by surgery (nine patients) or autopsy (two patients), or both.

Echocardiography. Diasonics (Varian) 3000 or 3400 and Advanced Technology Laboratory (ATL) Mark V wide angle, two-dimensional sector scanners with 2.25, 3 or 5 MHz transducers were utilized. Echocardiographic figures in this article are 35 mm photographs of stop-action video images. The standard two-dimensional echocardiographic scanning techniques utilized have been described (19).

Two scanning planes were the most helpful in elucidating the anatomic features of left juxtaposition of the atrial appendages. 1) A diagnostic alteration of the plane of the atrial septum and visualization of the juxtaposed right atrial appendage were appreciated from the parasternal short-axis scan at the level of the great arteries (section 9 [19]). 2) A consistent but nondiagnostic alteration of the posterior atrial septal configuration was appreciated from the apical four
Figure 1. Schematic diagrams illustrating the anatomy and echocardiographic planes of section seen in left juxtaposition of the atrial appendages. A, Anterior view of the heart showing the right atrial appendage (RAA) lying posterior and to the left of the transposed great arteries (Ao = aorta; PA = pulmonary artery) and anterior and superior to the left atrial appendage (LAA). B. The right atrium and juxtaposed right atrial appendage have been unroofed. Note anteriorly that the floor of the right atrial appendage is oriented within the heart from right to left in a horizontal plane. A short-axis plane of section (plane drawn through the atria) demonstrates the abnormal anatomy visualized by a parasternal short-axis echocardiographic scan. An inset (above) shows the resultant two-dimensional echocardiographic image. The right atrial appendage lies posterior and to the left of the great arteries and anterior to the left atrium. The posterior portion of the atrial septum (a) is oriented normally; the anterior portion (b) is oriented transversely and parallel to the anterior chest wall. RA = right atrium; RV = right ventricle. C, Four chamber view of the heart as visualized by two-dimensional echocardiography. In this plane, the atrial septum curves toward the right. The left atrium (LA) and right pulmonary veins (PV) wrap around the posterosuperior aspect of the right atrium. The inset (above) represents the resultant two-dimensional echocardiographic image (that is, four chamber plane). LV = left ventricle.

chamber view (section 11 [19]). Other useful planes of section (19) that were helpful included the subcostal four chamber plane (section 15) and suprasternal long-axis views (section 18).

Results

Two-dimensional echocardiography. The features of left juxtaposition of the atrial appendages by two-dimensional echocardiography were based on two observations: 1) visualization of the malpositioned right atrial appendage (Fig. 1C); and 2) a characteristic abnormal spatial orientation of the atrial septum (Fig. 1B and C). The parasternal short-axis scan at or just above the level of the semilunar valves was the most diagnostic view for visualization of the malpositioned right atrial appendage. Normally, the right atrial appendage cannot be visualized and the left and right atria are separated by an anteroposteriorly (vertical on the two-dimensional image) oriented atrial septum (Fig. 2A). Anteriorly, the atrial septum normally inserts beneath the posterior wall of a great artery. In normal subjects, this is the aorta; however, in the presence of transposed great arteries, it would be the pulmonary artery.

Abnormal spatial orientation of atrial septum. In all 10 patients with left juxtaposed atrial appendages, the major plane of the anterior atrial septum was abnormal and oriented horizontally from right to left as visualized from the parasternal short-axis scan at the level of the great arteries (Fig. 2B and 3). The major plane of the anterior portion of the atrial septum was transverse and parallel to the anterior chest wall. The horizontal portion of the atrial septum visualized is the floor of the juxtaposed right atrial appendage (Fig. 1B). The right atrial appendage was visualized to the patient’s left, posterior to the great arteries and anterior to the normally positioned left atrium and left atrial appendage (that is, filling the pericardial transverse sinus). In this projection, the posterior portion of the atrial septum inserted in a normal manner and separated the lower third of the right and left atrial cavities. In eight patients, an associated
Table 1. Clinical Data in 10 Patients

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yr) &amp; Sex</th>
<th>Cardiac Diagnosis</th>
<th>Diagnosis of Left Juxtaposed Atrial Appendages</th>
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<td>Prospective Echocardiographic Diagnosis (four patients)</td>
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<tr>
<td>1</td>
<td>14M</td>
<td>TA, TGA, PS, Glenn anastomosis, left SVC, left BT shunt</td>
<td>+</td>
</tr>
<tr>
<td>2</td>
<td>11F</td>
<td>DORV, VSD, PA, hypoplastic TV, Glenn anastomosis, ASD</td>
<td>+</td>
</tr>
<tr>
<td>3</td>
<td>9M</td>
<td>Dextrocardia, TGA, VSD, PS, ASD, right BT shunt</td>
<td>+</td>
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<td>4</td>
<td>15M</td>
<td>DORV, VSD, PA, ASD, right BT shunt, Waterston shunt, LAD off RCA</td>
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Retrospective Echocardiographic Diagnosis (six patients)

<table>
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<th>Case</th>
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<th>Cardiac Diagnosis</th>
<th>Diagnosis of Left Juxtaposed Atrial Appendages</th>
</tr>
</thead>
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<td>Dextrocardia, TGA, small VSD, ASD</td>
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<tr>
<td>7</td>
<td>17F</td>
<td>Dextrocardia, TA, ASD, PS, Glenn anastomosis, PDA</td>
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<td>TA, double-outlet outlet chamber, PS, left SVC</td>
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<td>16F</td>
<td>Postop Rastelli, complete TGA, obstructed RV to pulmonary artery conduit, small residual VSD</td>
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<tr>
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<td>13M</td>
<td>DORV, subaortic VSD, PA, postop Blalock-Hanlon + Waterston, midmuscular VSD, left SVC</td>
<td>+</td>
</tr>
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*Prospective angiographic diagnosis; ASD = atrial septal defect; AV = atroventricular valve; BT = Blalock-Taussig. Cath = cardiac catheterization. DORV = double outlet right ventricle; Echo = echocardiography, F = female, LAD = left anterior descending coronary artery, M = male, PA = pulmonary atresia, PDA = patent ductus arteriosus; Postop = postoperative; PS = pulmonary stenosis, RCA = right coronary artery, RV = right ventricle, SVC = superior vena cava, TA = tricuspid atresia, TGA = transposition of the great arteries; TV = tricuspid valve; VSD = ventricular septal defect, + = performed, – = not performed, – – = nondiagnostic.

secundum atrial septal defect was located either in the normally oriented posterior portion of the atrial septum or in the transition zone from the normal to the horizontal portion of the atrial septum (Fig. 2B).

A second consistent but nondiagnostic alteration of atrial septal orientation was observed in the four chamber plane as viewed from an apical or subcostal transducer position (Fig. 1C and 4). Superiorly, the atrial septum appeared to wrap around the posterior aspect of the right atrium (Fig. 4); this made the right atrium appear slightly smaller than the left. Inferiorly, the atrial septum inserted in a normal manner at the crest of the ventricular septum. In each patient with left juxtaposition of the atrial appendages, the atrial septum had this distinctly abnormal curvature toward the right as visualized in the four chamber projection. This alteration of atrial septal anatomy has been seen only in other complex congenital lesions, especially patients with isolated dextrocardia, and is not similar to the features seen with right atrial hypertension (for example, tricuspid regurgitation).

Less commonly from the subcostal or apical four chamber view and with anterior tilt of the transducer, the anteriorly located right atrial appendage could be visualized coursing beneath the great arteries and leftward over the left atrial cavity (Fig. 5).

From suprasternal or high parasternal long-axis projections of the great arteries, the juxtaposed right atrial appendage and normally positioned left atrial cavity were both
Figure 2. Case 4. Patient with double-outlet right ventricle and left juxtaposed atrial appendages. Sequential parasternal short-axis scans were used. A. At the base of the heart, the two transposed great arteries are visualized and the atrial septum (AS) is normal in orientation (that is, oriented anteriorly toward the posterior wall of the pulmonary artery [PA]). B. Scanning further superiorly, the plane of the atrial septum becomes nearly horizontal, coursing toward the left posterior to the great arteries (refer to inset in Fig. 1B). This distinctly abnormally oriented portion of the atrial septum represents the floor of the juxtaposed right atrial appendage. The juxtaposed right atrial appendage (RAA) is imaged posterior and to the left of the great arteries. The left atrium (LA) lies posterior to the right atrial appendage. Note that a secundum atrial septal defect is located just below the floor of the juxtaposed right atrial appendage (arrow). A = anterior; Ao = aorta; AV = aortic valve; L = left; P = posterior; R = right; RA = right atrium.

Figure 3. Case 3. Patient with dextrocardia, situs solitus of the atria and viscera, complete transposition of the great arteries and left juxtaposition of the atrial appendages. A high parasternal short-axis scan at the midatrial septum shows the diagnostic orientation of the atrial septum (AS, small arrows) seen in left juxtaposition of the atrial appendages. The right atrial appendage (RAA) lies posterior and to the left of the transposed arteries (Ao = aorta; PA = pulmonary artery). The body of the left atrium (LA) lies posterior to the right atrial appendage. Abbreviations as in Figure 1.

imaged posterior to the great arteries (Fig. 6). Normally, only the left atrium is visible posterior to the great arteries. This particular observation was obtainable in 4 of the 10 patients.

Angiography and catheterization. In four patients, sufficient contrast medium in the atria permitted a prospective diagnosis of juxtaposed atrial appendages. In an additional five patients, the diagnosis was made retrospec-

tively, at times with the use of roentgenographic subtraction techniques. One patient had insufficient contrast medium in the atria to permit a radiographic diagnosis. In all catheterized patients, simultaneous contrast echocardiography was performed to help confirm the various anatomic features described.

Surgery and autopsy. Nine patients underwent surgery and had sufficient exposure to confirm the diagnosis of left juxtaposition of the atrial appendages. In the 10th patient (Case 4), a juxtaposed right atrial appendage was not adequately visualized at surgery but could not be seen in its usual location. Two of the 10 patients died, and in both the diagnosis of left juxtaposition of the atrial appendages was confirmed at autopsy. The right atrium appeared somewhat smaller than normal but was normally located. The abnormal plane of the atrial septum, as observed by two-dimensional echocardiography, was confirmed at autopsy (Fig. 7). Anteriorly, the right atrial appendage was located behind and to the left of the great arteries and anterior and superior to the left atrial appendage (Fig. 7A). Posteriorly, the left atrium partially encircled the superior aspect of the right atrium (Fig. 7B).

Discussion

Left juxtaposition of the atrial appendages is almost always associated with complex cardiac malformations. Common associations include transposition of the great arteries
Figure 4. Apical four chamber scan in the presence of left juxtaposition of the atrial appendages. A, Case 4 with levocardia. Note the unusual rightward angulation of the superior atrial septum (AS, dashed line and arrow). The left atrium (LA) and right pulmonary veins (PV) are superior to the right atrium (RA). B, Case 5 with isolated dextrocardia and left juxtaposition of the atrial appendages. Note a similar abnormal rightward angulation of the atrial septum (AS, arrows). I = inferior; mv = mitral valve; S = superior; tv = tricuspid valve, VS = ventricular septum; other abbreviations as in Figure 1.

(more than 90% of cases), atrial septal defect (78%), ventricular septal defect (65%) and hypoplastic right heart with outflow obstruction (50%) or inflow obstruction (41%) (1,2,5,7–13,16,18–25).

Diagnostic and surgical implications. Recognition of juxtaposed atrial appendages is important for planning certain diagnostic and therapeutic procedures. Because of the abnormal plane of the atrial septum and position of the atrial appendages, an altered surgical or diagnostic approach may be required. Maneuvering a catheter across the abnormally oriented atrial septum can be difficult because of the more horizontal orientation of an atrial septal defect or patent foramen ovale (Fig. 2B). The orifice of the juxtaposed atrial appendage may be mistaken for an atrial septal defect at the time of surgery. The leftward position of the right atrial appendage might also lead to incorrect catheter positioning, thereby making balloon atrial septostomy a more risky procedure (1,5,13,16). The usual approach to the surgical Blalock-Hanlon atrial septostomy can be more difficult and risky because of the smaller right atrial size and rightward position of the left atrium. However, if juxtaposed atrial appendages are anticipated, a simple anastomosis of the atrial appendages is often preferred (12,13). Placement of an atrial septal baffle in the Mustard or Senning operation can be compli-

Figure 5. Case 10. Subcostal four chamber view with anterior scanning demonstrating the right atrial appendage (RAA) projecting to the left (arrow) beneath the great arteries. Abbreviations as before.

Figure 6. Case 4. High parasternal long-axis projection in a patient with transposed great arteries. Posteriorly, the juxtaposed right atrial appendage (white arrow) is visualized posterior to the transposed main pulmonary artery (MPA) (normally only the left atrium is visualized). The floor of the juxtaposed right atrial appendage (AS) separates the two atrial cavities. Small black arrows represent the position of a pulmonary artery band. Abbreviations as before.
Figure 7. Case 3. Pathologic specimen cut at two levels in the four chamber plane in a patient with transposition of the great arteries and dextrocardia. A, The anterosuperior atrial septum, forming the floor of the juxtaposed right atrial appendage, sweeps to the left (arrows) anterior to the body of the left atrium (LA). This section corresponds to the echocardiographic anatomy seen in Figure 5. B, Posteriorly, the atrial septum curves rightward (arrows). The left atrium partially encircles the superior aspect of the right atrium (RA), corresponding to the echocardiographic anatomy seen in Figure 4. Abbreviations as before.

cated by the smaller right atrium and atypical position of the orifice of the juxtaposed right atrial appendage (12–14). A Fontan procedure (anastomosis of the right atrium to the pulmonary artery) can be altered with anastomosis of the juxtaposed right atrial appendage to the left pulmonary artery (14,17).

The diagnosis of left juxtaposition of the atrial appendages is usually an incidental finding at surgery or autopsy. Only with a high degree of alertness and astute angiographic observation has prospective diagnosis been made. Chest roentgenography (3) and electrocardiography (4) have been of little help.

Echocardiographic diagnosis. This report describes the noninvasive recognition of left juxtaposition of the atrial appendages by two-dimensional echocardiography. The observations were based on standard echocardiographic projections (that is, parasternal short-axis views at the level of the great arteries and apical or subcostal four chamber views of the atrial septum). Recognition of left juxtaposition of the atrial appendages was based on characteristic alterations of the plane of the atrial septum and visualization of the leftward positioned right atrial appendage, which courses posterior to the great arteries. In the four chamber plane, the atrial septum curves characteristically to the right. Although consistently seen in patients with left juxtaposed atrial appendages, a similar configuration has been observed in the presence of other complex lesions, particularly isolated dextrocardia (situs solitus of the atria and viscera with cardiac apex to the right); therefore, in the presence of dextrocardia, this particular observation is less specific. However, in the presence of levocardia, this observation should consistently alert the echocardiographer to the possibility of left juxtaposed atrial appendages. This marked alteration of the posterior septal anatomy has not been seen in more than 10,000 examinations of lesions other than those associated with complex congenital heart disease, such as dextrocardia, extreme septal malalignment and left juxtaposition of the atrial appendages.

The most diagnostic echocardiographic observation is the horizontal orientation of the anterior atrial septum, which represents the floor of the left juxtaposed right atrial appendage (Fig. 1C). The left and right atrial appendages are both to the left of the great arteries and may be viewed from multiple transducer positions (parasternal, suprasternal, subcostal and apical). The right atrial appendage (cavity) is posterior to the great arteries and courses toward the left over the left atrium. The floor of the abnormally positioned atrial appendage accounts for the diagnostic image. In the normal heart, the right atrial appendage is not imaged as it lies anterior and to the right of the great arteries. The plane of the anterior atrial septum is normally vertical, with insertion on to the posterior wall of a great artery. The observations described in this report suggest that two-dimensional echocardiography is an excellent noninvasive means for making a definitive diagnosis of left juxtaposed atrial appendages. The sensitivity of this observation awaits further studies.

References


