Juxtaposition of the Atrial Appendages
A Report of Six Necropsied Cases

By Henry R. Wagner, M.D., Luis E. Alday, M.D., and Peter Vlad, M.D.

SUMMARY
Six cases of juxtaposition of the atrial appendages are documented by necropsied specimens and analyzed. A high incidence of conotruncal anomalies, especially transposition of the great arteries, further A-V valve abnormalities, and pulmonary valve atresia, is found. The recognition of juxtaposition of the atrial appendages during cardiac catheterization should alert the investigator to the presence of a complex cardiac anomaly. A case with a rare type of right juxtaposition of the atrial appendages and bizarre spatial orientation of the atria, ventricles, and great arteries is presented.

Additional Indexing Words:
Conotruncal malformation
Transposition of great arteries
Pulmonary valve atresia
Malrotation of cardiac loop

IN THE frontal view of the heart the right and left atrial appendages are normally separated by the great arteries. As the name indicates, juxtaposition of the atrial appendages may be defined as their side-by-side location, not separated by the great arteries (juxta [Latin] = close by, near to).

There are two types of juxtaposition, left and right, depending on whether the side-by-side atrial appendages both lie to the left or to the right of the great arteries (fig. 1A and B).

Juxtaposition of the atrial appendages has not been reported as an isolated congenital cardiac malformation but is accompanied by other almost predictable structural anomalies of the heart, usually of a severe nature and resulting in clinical cyanosis. This coexistence of a specific group of anomalies with juxtaposition of the atrial appendages constitutes a characteristic syndrome comparable to the asplenia syndrome. Since juxtaposition is easily demonstrated by angiocardiography (fig. 2), its presence suggests a complex cardiac malformation with frequently predictable components. Melhuish and Van Praagh recently reviewed 21 cases from the literature and added 21 new observations. The present report analyzes six further patients, one of them being the fourth hitherto reported case with the rare type of right juxtaposition.

Methods
Six specimens with juxtaposition of the atrial appendages were found among 800 hearts obtained at necropsy collected during the years 1955 to 1968. All patients died at the Children’s Hospital of Buffalo, New York. The segmental approach and terminology as proposed by Van Praagh and associates were used to describe and classify the anatomic characteristics of the specimens.

Results
The clinical and anatomic data are presented in tables and photographs. Table 1 gives the clinical data in our six patients. Table 2 presents the anatomic findings. Case 6 has unusual and hitherto undescribed anatomic features which are illustrated in figure 3.
Clinical Data

There were three males and three females. The patients survived from 1 day to 12 years of age (mean 4.5 years).

All patients had signs and symptoms of severe cyanotic heart disease since early life such as cyanosis, polycythemia, congestive heart failure, heart murmur, and abnormal electrocardiogram and chest x-ray. The pertinent data are listed in table 1. Figure 2 shows the typical angiographic appearance of left juxtaposition (case 3).

Anatomic Data

There were five cases with left juxtaposition (cases 1 to 5) and one case (case 6) with right juxtaposition. The anatomic features of the case with right juxtaposition will be described in detail while the ones with left juxtaposition will be summarized.

Abnormal position of the heart within the chest was present in two cases, one with dextrocardia and one with mesocardia.

The most frequently found segmental type of cardiac structure, found five times in our series, is S-D-D (situs solitus of viscera and atria, D-loop of the ventricles or non-inverted ventricles, and D-transposition of great arteries). Case 6 exhibits a malrotated D-loop, with situs solitus of the atria and abnormally related great arteries.

An anomaly of the conotruncus producing absence of mitral-aortic fibrous continuity is present in all six cases. In five cases (cases 1 to 5) this conotruncal malformation effected what is known as D-transposition of the great arteries. Case 6 exhibits a malrotated D-loop, with situs solitus of the atria and abnormally related great arteries.
Table 1

Clinical Findings in Six Patients with Juxtaposition of the Atrial Appendages

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Sex</th>
<th>Age at death</th>
<th>Auscultatory findings</th>
<th>ECG</th>
<th>Chest x-ray</th>
<th>Cardiac catheterization and angiography</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>3 mo</td>
<td>2/6 syst. M. LSB</td>
<td>RVH</td>
<td>CTR 61%</td>
<td>Left JAA, TGA, PS, VSD</td>
<td>Died in CHF</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>7 wk</td>
<td>2/6 syst. M. LSB</td>
<td>RVH</td>
<td>CTR 50%</td>
<td></td>
<td>Died postop. Blalock-Hanlon septectomy</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>12 yr</td>
<td>3/6 syst. M. LSB</td>
<td>RVH</td>
<td>CTR 50%</td>
<td>Left JAA, TGA, PA, ASD, VSD, LSVC</td>
<td>Died postop. AARPA shunt</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>2 yr</td>
<td>3/6 syst. M. LSB</td>
<td>RVH</td>
<td>CTR 53%</td>
<td>TGA, TS, PS, VSD, right aortic arch, PDA</td>
<td>Died after cardiac catheterization</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>12 yr</td>
<td>4/6 syst. M. RUSB</td>
<td>RVH</td>
<td>CTR 62%</td>
<td>Isol. dextrocardia</td>
<td>Died postop. right Blalock-Taussig shunt</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>1 day</td>
<td>3/6 syst. M. LSB</td>
<td>RVH</td>
<td>PV increased</td>
<td>TGA, PA, TA, VSD, PDA, LSVC</td>
<td>Died in CHF</td>
</tr>
</tbody>
</table>

Abbreviations: syst. M. = systolic murmur; cont. M. = continuous murmur; LSB = left sternal border; RUSB = right upper sternal border; RVH = right ventricular hypertrophy; BVH = biventricular hypertrophy; LAH = left atrial hypertrophy; BAH = bialtrial hypertrophy; CTR = cardiothoracic ratio; PV = pulmonary vascularity; JAA = juxtaposition of atrial appendages; TGA = transposition of great arteries; TA = tricuspid atresia; PA = pulmonary valve atresia; PS = pulmonary stenosis; ASD = atrial septal defect; VSD = ventricular septal defect; TS = tricuspid stenosis; PDA = patent ductus arteriosus; LSVC = left superior vena cava; CHF = congestive heart failure; AARPA shunt = ascending aorta-right pulmonary artery shunt.
Anatomic Findings in Six Patients with Juxtaposition of the Atrial Appendages

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</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>3 mo</td>
<td>L</td>
<td>S-D-D</td>
<td>ASD II</td>
<td>N</td>
<td>N</td>
<td>H + E</td>
<td>H + E</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>3 × 5 mm</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>7 wk</td>
<td>L</td>
<td>S-D-D</td>
<td>PFO</td>
<td>N</td>
<td>N</td>
<td>H + E</td>
<td>TW + SC</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>12 yr</td>
<td>L</td>
<td>S-D-D</td>
<td>PFO</td>
<td>N</td>
<td>N</td>
<td>H + E</td>
<td>H</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>2 yr</td>
<td>L</td>
<td>S-D-D</td>
<td>ASD II</td>
<td>TS</td>
<td>N</td>
<td>H + E</td>
<td>TW + SC</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>12 yr</td>
<td>D</td>
<td>S-D-D</td>
<td>ASD II</td>
<td>TAT</td>
<td>N</td>
<td>Hypoplastic RV situs</td>
<td>H + E</td>
</tr>
</tbody>
</table>

| 6        | F   | 1 day        | M              | Malrotated D-loop | ASD II        | N MV         | Abnormal TV | Small (L) | H + E (R) |
|          |     |              |                | 8 × 8 mm         |              |              |              |             |             |

**Left juxtaposition of atrial appendages**

**Right juxtaposition of atrial appendages**

Abbreviations: O = absent; + = present; H = hypertrophy; E = enlargement; N = normal for age; L = levocardia; D = dextrocardia; M = mesocardia; S = situs solitus of atria; -D = D-loop (non-inverted ventricles); -D = D-transposition of great arteries; ASD II = secundum type of atrial septal defect; PFO = patent foramen ovale; N = normal; TS = tricuspid stenosis; TAT = tricuspid atresia; Morph. = morphologically; TW + SC = thick-walled and small chambered; (R) = right sided; (L) = left sided; VSD = ventricular septal defect; A = subaortic; B = bilateral; SLV-AVV continuity = semilunar valve-atrioventricular valve continuity; PV = pulmonary valve; MV = mitral valve; PAT = pulmonary valve atresia; PS = pulmonary stenosis; inf. = infundibular; L = left; R = right.

arteries (aortic valve anterior and to the right of the pulmonary artery). One of these five (case 1) has a subaortic conus only (permitting pulmonary valve-mitral valve continuity), while four specimens are characterized by a bilateral conus (no semilunar valve-atrioventricular valve continuity). Case 6 also exhibits a conotruncal malformation but here transposition of the great arteries is not produced. A bilateral conus (subpulmonary and subaortic) is present. The aorta is related to the anatomic left ventricle, and the pulmonary artery takes off from the right ventricle.

In four specimens an atrial septal defect of the secundum type is found; two have a patent foramen ovale-type opening. A high ventricular septal defect, usually of moderate size, is present in all six hearts.

Tricuspid valve atresia is present in one case (case 5). Case 6 also presents a tricuspid valve anomaly, as described separately.

In four hearts the pulmonary valve is atretic, and one other has infundibular pulmonic obstruction (case 6). Case 1 shows a narrowed subpulmonic fibrous ring and a bicuspid pulmonic valve.

Case 4 exhibits a right aortic arch and a right-sided patent ductus arteriosus. The spleen was present in all cases.

Anatomic Description of Case 6 (Fig. 3)
The heart is located in the mid chest but the apex points toward the left. Situs solitus of the viscera including lungs and atria is present. There is right-sided juxtaposition of the atrial appendages (fig. 3A). The right atrial appendage is extremely to the right and somewhat posterior to the left atrial appendage. The right-sided morphologically right atrium receives the superior vena cava, the inferior vena cava, and the coronary sinus normally. The septal surface displays a secundum type atrial septal defect. The right atrium opens into a morphologically right ventricle which is left-sided and lies posterior to the other ventricle. The tricuspid valve ring is relatively small, and multiple wartlike polyps along the free margin of the valve leaflets are present.
All chordae tendineae insert directly into the wall of the right ventricle with no evidence of true papillary muscles (fig. 3B). The pulmonary artery originates above the morphologically right ventricle (fig. 3C). The subpulmonary conus is well developed, resulting in fibrous discontinuity between the pulmonary valve and both atrioventricular valves below. The morphologically left atrium which is left sided and anterior to the right atrium but completely to the right of the great arteries receives the pulmonary veins (fig. 3A). It opens into the morphologically left ventricle via a structurally normal mitral valve (atrioventricular concordance). The left ventricle, however, is anterior and to the right. The aorta is related to the left ventricle, but the presence of a subaortic conus separates the aortic valve above from the mitral valve below.

The semilunar valves are approximately side by side, the aortic valve is to the right, and the pulmonary valve is to the left. The pulmonary valve is somewhat more superior than the aortic valve. Both semilunar valves are tricuspid. The aortic valve is distinctly smaller than the pulmonary valve. Viewed from below, a subaortic conus is present due to the subaortic infundibulum. The ascending aorta and the left-sided aortic arch are hypoplastic. A large patent ductus arteriosus with a considerably wrinkled intima leads to the descending aorta.

Discussion

Melhuish and Van Praagh\(^1\) noted the rarity of right-sided juxtaposition as compared to left (less than 10% of all juxtapositions, or only three cases reported so far), and this is related to the fact that inverted ventricles (L-loops) are much less frequent than non-inverted ventricles (D-loops), since all three had L-loops. In our case with right juxtaposition, however, ventricular inversion is simulated by malrotation of a D-loop. This case clearly does not have a discordant ventricular loop because of the presence of atrioventricular concordance. The anatomic left ventricle, however, is anterior to the anatomic right ventricle, which is posterior. The rule of coexistence of juxtaposition of the atrial appendages only with L-loops is broken by our case 6.

All cases have conotruncal abnormalities. The combined juxtaposition-conotruncal malformation appears almost uniform, since only three previously reported patients had normally related great arteries.\(^1\)\(^,\)\(^5\)\(^,\)\(^6\) Our case 6 has a distinct conotruncal anomaly but no transposition. The aortic valve and ascending aorta are abnormally related to the mitral valve, although they take off from the left ventricle.
This case illustrates that not all conotruncal anomalies (such as a subaortic conus) lead to transposition of the great arteries.

D-transposition in the S-D-D segmental combination is found most frequently in our collection as well as in the review by Melhuish and Van Praagh.\(^1\)

Other salient features in our series such as atrial and ventricular septal defects, A-V valve abnormalities, outflow tract obstruction, pulmonary valve atresia, right aortic arch, and patent ductus arteriosus occur in about as similar a frequency as in the recent review from the literature.\(^1\) We have not, however,
encountered double outlet right ventricle in our small series. Five specimens (cases 1 to 5) have similar anatomy and correspond to the most frequently described segmental combination and concomitant anomalies. Case 6 with right juxtaposition, situs solitus of the atria, a malrotated D-loop, and a subaortic conus but no transposition of the great arteries is distinctly different from any previously reported specimen with juxtaposition.

Morphogenetic considerations raised by Melhuish and Van Praagh merit mention. In their opinion, juxtaposition of the atrial appendages appears to be a secondary effect produced by one or more associated anomalies, not a primary cardiac malformation per se. Our series shows that in left juxtaposition only the right atrial appendage is placed to the left of the great arteries while the rest of the right atrium remains to the right. In our case with right juxtaposition, however, the entire left atrium is placed to the right side of the great arteries. Here, the concomitant malrotation of the primitive cardiac loop may have contributed to this unusual spatial arrangement. This suggests that several factors may be responsible for the pathogenesis of juxtaposition of atrial appendages.

Acknowledgment
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