CLINICOPATHOLOGIC CORRELATIONS

Juxtaposition of the Atrial Appendages

By Yzhar Charuzi, M.D., Panayotis K. Spanos, M.D., Kurt Amplatz, M.D., and Jesse E. Edwards, M.D.

SUMMARY
Juxtaposition of the atrial appendages refers to that condition in which both atrial appendages or one and part of the other lie beside each other and to one side of the great arterial vessels. The appendages lying to the left of the great vessels (left-sided juxtaposition) is considerably more common than the state in which the appendages lie to the right of the great vessels (right-sided juxtaposition) by a ratio of about 6:1. Among left-sided juxtaposition it is common that the right atrial appendage be bifid and that only its left unit be “juxtaposed.” Such a state is termed “bifid atrial appendage with partial juxtaposition.” Juxtaposition almost always is associated with significant congenital heart disease in which transposition of the great vessels is commonly a feature. Tricuspid atresia with transposition or complete transposition are the two conditions most commonly associated with juxtaposition. Recognition of juxtaposition as an entity may serve, under certain circumstances, as an aid in the evaluation of an angiocardiogram.

Additional Indexing Words:
Transposition of great vessels  Tricuspid atresia

In patients with major types of congenital heart disease, one may encounter details which, while they attract attention, do not contribute to the functional disturbance. One such condition is juxtaposition of the atrial appendages.

Normally, each atrial appendage lies to the respective side of the two great arterial vessels (fig. 1). Uncommonly, both or one and part of the other lie to one side of these vessels, yielding a condition named juxtaposition of the atrial appendages by Dixon. The condition may cause unusual shadows in angiocardiograms (fig. 2). Two basic types may be identified, the more common wherein both appendages lie to the left of the great vessels (left-sided juxtaposition) (figs. 3, 4) and much less common the type in which the atrial appendages lie to the right of the great vessels (right-sided juxtaposition) (fig. 5).

To our knowledge, 70 cases of juxtaposition of the atrial appendages have been reported, the left-sided being more common than the right-sided by a ratio of 6:1. Classically, each type is associated with significant congenital heart disease in which some type of transposition of the great vessels is usually manifest. For example, among the nine cases of right-sided juxtaposition reviewed by Becker and Becker, seven showed some form of transposition. In their case, representing the tenth reported example of right-sided juxtaposition, the great vessels were normally related as in the case previously described by Wagner and associates.

Existing reports suggest that when juxtaposition occurs all of the involved atrial appendage is malpositioned. On the contrary, our studies have shown that in some cases of left-sided juxtaposition, the right atrial appendage may be bifid and one element is juxtaposed while the other lies in a normal position. One may, therefore, speak of complete or partial juxtaposition.

We reviewed the 16 specimens with juxtaposition in our files so as (1) to define the nature and to

From the Department of Pathology, United Hospitals-Miller Division, St. Paul, Minnesota and the Departments of Pathology and Radiology, University of Minnesota, Minneapolis, Minnesota.

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Address for reprints: Dr. Jesse E. Edwards, Department of Pathology, United Hospitals, Inc.-Miller Division, 125 West College Ave., St. Paul, Minnesota 55102.
determine the incidence of bifid right atrial appendage with partial juxtaposition and (2) to determine the types of congenital heart disease which were associated. We also studied all available specimens with those types of congenital heart disease that are most frequently associated with juxtaposition of the atrial appendages in order to learn the incidence of juxtaposition in these conditions.

Bifid Right Atrial Appendage with Partial Juxtaposition

In our studies, we commonly observed a variant of left-sided juxtaposition. This is characterized by the right atrial appendage exhibiting a bifid state. The left unit lies to the left of the great vessels, beside the left atrial appendage, while the right unit lies to the right of the great vessels in a normal position. Pectinate muscles are present in each unit of the bifid right atrial appendage while, between these, pectinate muscles are absent (figs. 6, 7). We have called this variant bifid right atrial appendage with partial left-sided juxtaposition of the atrial appendages. It was seen in nine specimens among 15 cases of left-sided juxtaposition.

Figure 1

Normal atrial appendages. (a.) The great vessels have been reflected forward to allow a view from in front and above of the right (R.A.A.) and left (L.A.A.) atrial appendages. The probe is in the superior vena cava (S.V.C.). (b.) Frontal section has been made through the atria following which the upper segment was removed to expose the interior of the right (R.A.) and left (L.A.) atria. A.S. = atrial septum. The right (R.A.A.) and left (L.A.A.) atrial appendages are in normal positions.
Figure 3

Complete left-sided juxtaposition of the atrial appendages in a case of complete transposition of the great vessels. The right (R.A.) and the left (L.A.) atrial appendages lie side by side and to the left of the great vessels. A = aorta; P = pulmonary trunk.

Of the 15 cases with left-sided juxtaposition, nine were examples of the partial type, and six were of the complete type. Among the 15 cases, the most common associated condition was complete transposition (eight cases). In three of these, pulmonary or subpulmonary stenosis was associated. In five other of the 15 cases of left-sided juxtaposition, atresia of the right atrioventricular valve was present. In four of these, this involved the tricuspid valve. In the fifth instance, there was inversion of the ventricles and of the atrioventricular valvular mechanism so that the right-sided atretic valve would be interpreted as having been the anatomic mitral valve. In this case, the great vessels were transposed as they were in three of the four cases of classical tricuspid atresia. Pulmonary or subpulmonary stenosis was present in two of the five cases with atresia of the right atrioventricular valve.

In one of the two remaining cases of left-sided juxtaposition, the associated condition was double-outlet right ventricle and, in the other, corrected transposition of the great vessels.

The distribution of associated conditions was about the same for the nine cases of the partial left-sided type as among the six cases of complete left-sided juxtaposition.
Figure 4

Complete left-sided juxtaposition of the atrial appendages. (a.) The great vessels have been reflected forward allowing a view from above of the right (R.A.A.) and left (L.A.A.) atrial appendages. These lie beside each other and to the left of the great vessels. S.V.C. = superior vena cava. (b.) View from above after frontal section of the atria has been done and the roof of the atria removed. The entire right atrial appendage (R.A.A.) lies to the left of the great vessels and beside the left atrial appendage (L.A.A.). A.S. = atrial septum.

Among the entire series of 16 cases with juxtaposition, only one, a case of tricuspid atresia, did not show some form of transposition. In addition to other malformations, isolated dextrocardia was observed in four instances of our 16 cases with juxtaposition. In three, the juxtaposition was of
Figure 5

Complete type of right-sided juxtaposition in a case of single ventricle with transposition of the great vessels. (a.) View from above. The right (R.A.A.) and left (L.A.A.) atrial appendages lie beside each other and to the right of the great vessels. A. = transposed aorta. (b.) Interior of atria viewed from above. The left atrial appendage (L.A.A.) lies to the right of the great vessels and beside the right atrial appendage (R.A.A.). Probe in superior vena cava. A.S. = atrial septum.

the complete left-sided type and, in the fourth case, it was of the partial left-sided variety.

Incidence of Juxtaposition in Several Types of Congenital Heart Disease

The usual report on juxtaposition relates the various types of cardiovascular malformations which are associated with observed malposition of the atrial appendages. It is also of interest to determine the incidence of juxtaposition among those conditions with which a recurring association with juxtaposition is recognized.
Partial left-sided juxtaposition of the atrial appendages in a case of complete transposition of the great vessels. A. = aorta; P. = pulmonary trunk. One portion of a bifid right atrial appendage (R.A.A.I) lies to the left of the great vessels and beside the left atrial appendage (L.A.A.). Part of the right atrial appendage (R.A.A.II) lies to the right of the great vessels.

We therefore reviewed all specimens in our files of tricuspid atresia with transposition, complete transposition, and corrected transposition for the incidence of juxtaposition.

Among our 13 cases of tricuspid atresia with transposition of the great vessels but without ventricular inversion, there were three cases of juxtaposition (23%), each left-sided. In one of the cases, pulmonary stenosis was associated. These three cases, each associated with tricuspid atresia and transposition, were among five cases in our series in which atresia of the right atrioventricular valve was associated. Of the other two, one was an example of tricuspid atresia without transposition and the other displayed inversion of the ventricles and of the left atrioventricular valve. The atretic right atrioventricular valve was therefore to be considered an atretic inverted mitral valve.

Among the 136 specimens with complete transposition of the great vessels, there were eight instances of juxtaposition (6%), each of the left-sided type. Since among these cases there were variations both as to size of the right ventricle and as to the caliber of the pulmonary arterial tract, analysis was made upon the incidence of juxtaposition considering these two conditions.

Among the 14 cases of complete transposition with pulmonary stenosis but with a normally developed right ventricle, the incidence of juxtaposition was highest among the subgroups considered, this condition being observed in two cases (15%). One case (8%) of juxtaposition was found among the eight cases of complete transposition with hypoplastic right ventricle and with pulmonary stenosis. No examples of juxtaposition were found in the four cases of complete transposition with hypoplasia of the right ventricle but without pulmonary stenosis. Five cases (4.5%) were observed in the 110 cases of complete transposition without pulmonary stenosis or hypoplasia of the right ventricle.

One case (4%) of juxtaposition was found among the 28 cases of corrected transposition reviewed. Although pulmonary stenosis was present in 19 of these cases, the one case of juxtaposition associated with corrected transposition did not show this condition.

A potential source of misinterpretation of juxtaposition in angiograms is the uncommon situation in which both atrial appendages are in normal positions, while the left atrial appendage...
Partial left-sided juxtaposition of atrial appendages. (a) The great vessels have been reflected forward allowing a view of the atrial appendages from above and in front. The right atrial appendage is bifid. Its left half (R.A.A.I) lies to the left of the great vessels and beside the left atrial appendage (L.A.A.). The right half of the right atrial appendage (R.A.A.II) lies in a normal position. Probe in superior vena cava. (b) Interior of the atria viewed from above. The right half of the right atrial appendage (R.A.A.II) shows pectinate muscles as does the left half (R.A.A.I), the latter lying beside the left atrial appendage (L.A.A.).

A.S. = atrial septum.

Figure 7

exhibits an uncommon variant of normal in the form of a bifid state (fig. 8). While it is hypothetically possible for one of the two units to be juxtaposed to the right of the great vessels, we have not observed an example of right-sided juxtaposition which was of the partial form.

Juxtaposition of the atrial appendages does not contribute to malfunction of the circulation, yet it is of value to the radiologist and clinician to know of this entity as a basis for the understanding of certain peculiarities that may be present in given cases. As a potentially practical point, anastomosis of the two appendages would constitute a simple way of increasing communication between the two sides of the heart, as might be desired in complete transposition or in tricuspid atresia with a narrow foramen ovale. Even more simple, however, for this purpose is the closed Rashkind procedure.
References


Figure 8

Bifid left atrial appendage without juxtaposition, in a case of complete transposition in which the right atrial appendage was in a normal position. The left atrial appendage is bifid, represented by two structures (L.A.A.I; L.A.A.II). These lie to the left of the great vessels. L.A. = left atrium.
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YZHAR CHARUZI, PANAYIOTIS K. SPANOS, KURT AMPLATZ and JESSE E. EDWARDS

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