Juxtaposition of Atrial Appendages Associated with Normally Oriented Ventricles and Great Arteries

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SUMMARY

A case is reported of a stillborn infant in whom both atrial appendages lay to one side of the great arteries, so-called juxtaposition of the atrial appendages.

The unusual features of the case reported are twofold. First, both atrial appendages lay to the right of the great vessels while, more commonly, juxtaposed atrial appendages lie to the left of the aorta and pulmonary trunk.

Second, in our case the great vessels were normally related and no major intracardiac malformations were present (only an atrial septal defect and a bicuspid pulmonary valve), while, more commonly, the great vessels are transposed and major intracardiac malformations are associated.

Additional Indexing Words:
Right-sided juxtaposition of atrial appendages

Juxtaposition of atrial appendages, a condition in which both atrial appendages lie to one side of the great arteries, is an uncommon congenital malformation. The situation in which both atrial appendages lie to the left side of the great vessels (left-sided juxtaposition) is more common than that in which the appendages lie to the right of the arteries (right-sided juxtaposition). As far as we are aware, 59 cases of left-sided juxtaposition have been reported,1-4 whereas only nine reported cases of right-sided juxtaposition are known to us. In most cases reported, juxtaposition was accompanied by major deviations from normal in cardiac structure including transposition of the great vessels and ventricular inversion. Atresia or stenosis of the right atrioventricular valve and pulmonary stenosis are also frequently associated conditions.

The rarity of right-sided juxtaposition, especially in the normally oriented hearts, prompts us to record a new case.

Report of Case

The subject was a stillborn, anencephalic, female infant with a probable gestational age of 36 weeks. Apart from anencephaly with complete rachischisis and hypoplasia of the adrenals, there was a large, left-sided diaphragmatic hernia. Several abdominal organs were present in the left hemithorax. These included the spleen, the left adrenal gland and kidney, the left lobe of the liver, the stomach, and the greater part of the small intestine. The thoracic organs were displaced into the right hemithorax. Each lung, normally lobulated, was hypoplastic (total weight, 4 g).

The cardiac apex was directed toward the left, and normal relationships existed between the ascending aorta and pulmonary trunk and between these vessels and the heart. The aortic arch passed over the left main bronchus. No atrial

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appendage was present to the left of the great arteries, while two appendages were present to the right of these structures (fig. 1). The superior one, located immediately adjacent to the ascending aorta, was slender. It was the appendage of the left atrium. The inferior and more laterally located appendage was large and massive. The latter communicated with the right atrium with which the superior and inferior caval veins and the coronary sinus were connected. In the septal wall of the right atrium, there was a distinct limbus of the fossa ovalis covered on the left side by a fenestrated valve of the foramen ovale.

Each of the four pulmonary veins joined into the left atrium. The ostium of the left atrial appendage was wide, measuring about 0.5 cm in diameter. It was located just anterior to the anterior free edge of the valve of the foramen ovale (fig. 2) and led into the superior of the two appendages which were seen from the exterior of the heart.

The right and left atrioventricular ostia possessed valves with characteristics of tricuspid and mitral valves, respectively. Numerous pouchlike protrusions directed upward were present in each of the three tricuspid leaflets.

The ventricles were not inverted as evidenced by the fact that the right-sided and left-sided ventricles showed morphologic characteristics of the anatomic right and left ventricles, respectively. The great arteries arose normally from the ventricles. There was a bicuspid pulmonary valve, with left and right cusps. The latter was the larger and a low raphe extended along its arterial aspect. The aortic valve was normal. The coronary arteries arose normally from the aorta.

The pericardial reflection line around the vessels was normal. A few pericardial adhesions were found over the ventral surface of each atrial appendage.

**Figure 1**

**Figure 2**
Interior of left atrium (L.A.) and left ventricle (L.V.). Each chamber, as well as the atrioventricular valve, exhibits the morphologic characteristics for the left side of the heart. Anterior to the fenestrated valve of the foramen ovale is the orifice (O.) of the left atrial appendage. L.L.V. = orifice of left lower pulmonary vein.
JUXTAPOSITION OF ATRIAL APPENDAGES

Comment

As far as could be judged from the literature, the present case is rare in exhibiting no severe disturbance of cardiac structure in combination with juxtaposition of the atrial appendages and is of further interest in being right-sided. So common is the association of juxtaposition with major forms of congenital heart disease that the presence of juxtaposition is usually a reliable index as to the presence of major intracardiac malformations.2

Among a total of 68 reported cases of juxtaposition of atrial appendages which we were able to find in the literature, only nine showed the right-sided type.

The following is a summary of these nine cases of right-sided juxtaposition of the atrial appendages. The earliest reported case was that of Abbott.5 From a man, 20 years old, the necropsy exhibited complete transposition of the great vessels, rudimentary ventricular septum, stenosis of the infundibulum of the right ventricle, hypoplasia of transposed aorta, and ventricular septal defect. In 1941, Saphir and Lev6 presented a case of double outlet right ventricle in a male, 21 years of age, with this condition. In 1954, Dixon,7 who introduced the term of juxtaposition of the atrial appendages, described this condition in a newborn infant with partial transposition of the great vessels, common pulmonary vein, hypoplasia of right ventricle and tricuspid valve, atrial septal defect, coarctation of the aorta, and patent ductus arteriosus.

The heart of a 4-week-old male infant with right-sided juxtaposition of atrial appendages, described by Meitner and Slugene8 showed complicated malformations as well. In this case, transposition of great vessels with ventricular inversion, ventricular and atrial septal defects, and a bicuspid pulmonary valve were also present. According to Meitner,1 two similar cases were reported by Dušek.9 In 1968, Melhuish and Van Praagh2 described two cases of right-sided juxtaposition. Each was a newborn infant in whom ventricular inversion was associated with atrial and ventricular septal defects. In one of these cases, the great vessels were normally related, but the left atrioventricular valve was atretic. In the other, the great vessels were transposed and inverted, and the left atrioventricular valve was hypoplastic.

The ninth reported case of right-sided juxtaposition was an exception. This case, reported by Edwards,10 showed the basic structure of the heart and great vessels to be normal. The only associated abnormality was a ventricular septal defect.

Explanation of the developmental basis of left-sided juxtaposition was given by Wenner in 1909.11 This author considered the condition to represent underdevelopment of torsion of the primitive cardiac tube. Dixon,7 accepting this explanation, proposed that right-sided juxtaposition was caused by “overdevelopment” of the process of torsion. In a treatise on embryology of the heart, Los12 accepted these explanations.

From a functional point of view, juxtaposition appears to be of no significance, although it may give rise to a confusing situation in diagnostic studies.

Knowledge that juxtaposition may accompany other cardiac malformations serves to make understandable certain peculiarities that may be encountered during diagnostic studies.

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