

LEFT JUXTAPOSED ATRIAL APPENDAGES IN A PATIENT WITH DEXTROCARDIA AND TRICUSPID ATRESIA - SURGICAL IMPLICATIONS

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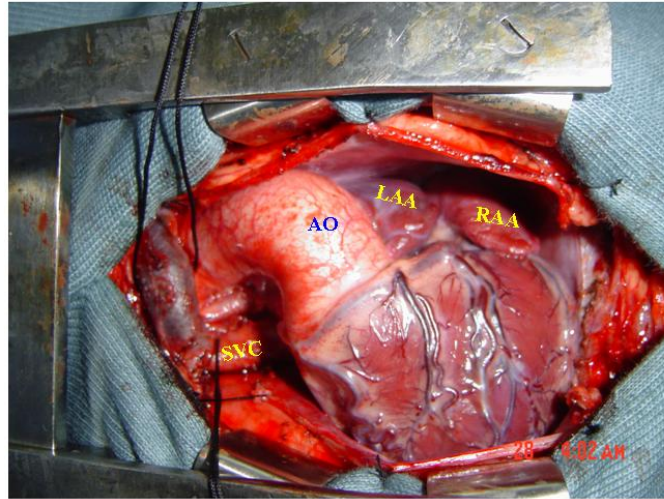
Introduction:

The frequent association of complex congenital heart defects and left juxtaposition of atrial appendages (LJAA) is well known. The incidence of LJAA is seen in approximately 10% of those with tricuspid atresia, and in up to 30% of those with tricuspid atresia with ventriculo-arterial discordance. We report a case of LJAA in a patient with tricuspid atresia and dextrocardia.

Case Report:

A one-and-a-half year old male child, the fourth born of consanguineous parents was brought to Frontier Lifeline with a history of cyanosis since birth. The child had four episodes of cyanotic spells since birth. There was no history of forehead sweating or failure to thrive or recurrent respiratory tract infections. On examination, the child was cyanosed with an oxygen saturation of 75% by pulse oximetry. Clubbing was present. Cardiovascular examination revealed a systolic murmur at the left lower sternal border and apex. There was no splitting of the second heart sound. Hemoglobin was 18gm and hematocrit was 58%. All other hematological and biochemical investigations were normal. Electrocardiogram showed sinus rhythm, right axis deviation and right atrial enlargement. Chest radiograph showed dextrocardia, cardiothoracic ratio of 65%, right ventricular apex and pulmonary oligemia. Two dimensional echocardiography demonstrated tricuspid atresia with a large secundum atrial septal defect. The right ventricle was hypoplastic with severe pulmonary stenosis and good sized branch pulmonary arteries. Cardiac catheterization confirmed the echocardiography findings. Additionally, one MAPCA was seen to arise from the descending aorta, which was coil embolised.

The child underwent a bi-directional Glenn procedure through a median sternotomy using cardiopulmonary bypass. Intraoperatively, there was dextrocardia. Both atrial appendages were juxtaposed on the left side with distinct morphological characteristics (Figure 1). The aorta was seen to arise anterior to the pulmonary trunk. The superior vena cava was divided and the atrial end was oversewn. The proximal end was anastomosed to an arteriotomy made on the superior surface of the right pulmonary artery. The child was weaned off cardiopulmonary bypass and the post-operative recovery was uneventful.



INTRA-OPERATIVE PHOTOGRAPH SHOWING JUXTAPOSED ATRIAL APPENDAGES (Fig 1)

AO – Aorta

PA – Pulmonary Artery

LAA – Left Atrial Appendage

RAA – Right Atrial Appendage

Discussion:

Developmentally, juxtaposed atrial appendages is a consequence of ectopic origin of the atrial appendage rudiment, and is a marker for the presence of cyanotic congenital heart disease¹. More morbid anomalies are seen in those with left juxtaposition than in those with right juxtaposition². Left juxtaposed atrial appendages are most commonly associated with transposition of great vessels, and these patients are likely to have co-existent right ventricular inflow stenosis or atresia². However, association of juxtaposed atrial appendages with isolated tricuspid atresia and dextrocardia is uncommon.

The presence of juxtaposed atrial appendages in a univentricular situation has got certain surgical implications. First, it is easy to create an inter-atrial communication by performing appendage-to-appendage anastomoses. This obviates the need for cardiopulmonary bypass and cardioplegia to do a formal atrial septectomy. The second implication relates to the construction of direct total cavopulmonary anastomoses. Direct anastomosis between the inferior vena cava and main pulmonary artery is advantageous since it avoids the thrombogenic potential, peel formation and infectious complications associated with the use of a prosthetic conduit⁴. Further, growth potential is maintained and costs are lowered⁴. It is better than an intra-atrial tunnel which has a high incidence of atrial arrhythmias, atrial thrombus formation and atrially induced pulmonary vein obstruction. The presence of L-malposed great vessels and left juxtaposed atrial appendages is the ideal situation for the construction of direct anastomoses without tension^{4, 5}. However, this can be performed only in the presence of levocardia. In a situation where dextrocardia exists, as in our patient, a bi-directional Glenn shunt is the logical option.

References:

1. Munoz Castellanos L, de la Cueva R, Zavaleta D, Kuri Nivon M. Juxtaposition of the atria. Arch Inst Cardiol Mex.1989 Jul-Aug; 59 (4):375-382.
2. Leu M R, Chiu I S, Hung C R, Wu M H. Surgical implications of juxtaposed atrial appendages and the associated anomalies. The Ann Thorac Surg 1992; 54: 134-136.
3. Kaneko Y, Okabe H, Nagata N, Kobayashi J, Kanemoto S. Anastomosis of the left juxtaposed atrial appendages in a patient with tricuspid atresia. Ann Thorac Surg 1998; 65 : 1783-1784.
4. van Son J A M, Reddy M V, Hanley F L. Extracardiac modification of the Fontan operation without the use of prosthetic material. J Thorac Cardiovasc Surg 1995; 110: 1766-1768.
5. Cariotti A, Amodeo A, Giamberti A, Marianeschi S, Nava S, De Simone G, di Carlo D C, Marcelletti C. Total cavopulmonary direct anastomosis: a logical approach in selected patients. Ann Thorac Surg 1993; 56: 963-964.