

RIGHT JUXTAPOSITION OF THE ATRIAL APPENDAGES

Sohail Khan Bangash¹, Iqbal Hussain Pathan²,
Saad Bader Zaki³, Toufik Ahmed⁴

¹⁻⁴ Department of Paediatric Cardiac Surgery National Institute of Cardiovascular Disease, Karachi, Pakistan

Address for Correspondence:

Sohail Khan Bangash,

Department of Paediatric Cardiac Surgery National Institute of Cardiovascular Disease, Karachi, Pakistan

E-Mail: sohailbangash@yahoo.com

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Contribution

SKB conceived the idea, planned the study. IH did literature review and final drafting. SBZ reviewed the case report. TA helped in acquiring photographs. All authors contributed significantly to the submitted manuscript.

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ABSTRACT

We are presenting extremely rare congenital anomaly of atrial appendage. By itself atrial juxtaposition is a benign condition. Nevertheless its strong association with other congenital heart disease well established. In addition its presence may change the surgical plan for certain conditions like maze procedure, senning as well as change in position of conduction tissue in left juxtaposition

Key Words: Atrial Appendage, Congenital Hear Disease, Maze Procedure, Senning

INTRODUCTION

Juxtaposition of atrial appendages is a rare congenital anomaly. The atrial appendages lying side by side on either side of great arteries is a one of rare presentation of cardiac anomalies. We are presenting a case report of a child with extremely rare congenital anomaly of right-sided juxtaposition of atrial appendages. Melhuish and Van Praagh reviewed 42 cases of atrial Juxtaposition and out of these only 3 were of the right juxtaposition type.¹ They observed high association of transposition of the great arteries in the case of left juxtaposition (95 %) and the ventriclular inversion with right juxtaposition of atrial appendage. Beside these anomalies atrial juxtaposition is found to be associated with ventricular septal defect (88%); atrial septal defect (78%); hypoplastic right ventricle (71%) and pulmonary or aortic outflow obstruction (52 %and 38%) respectively.¹

CASE REPORT

A 6 year old baby girl presented with shortness of breath and recurrent chest infection along with failure to thrive. She was a diagnosed case of ventricular septal defect. Her antenatal history was unremarkable. She was born with normal vaginal delivery at full-term. She was delayed for proper intervention with frequent palliative non invasive interventions. At age of 6 years, she was hospitalized for congestive heart failure. On physical examination, she had peripheral arterial pulses with 111 beats per minute with regular rhythm, no cyanosis, respiratory rate 22 breadth per minute. Examination of precordium revealed left parasternal heave with laterally displaced apex. S1 and S2 ware audible in all four areas with loud pulmonary component of second heart sound

with grade 4/6 holosystolic murmur along left parasternal area. The electrocardiogram revealed sinus rhythm, and right axis deviation with signs of right ventricular hypertrophy. The chest X-ray showed cardiomegaly. Echocardiography revealed basal muscular VSD of 13mm with left to right shunt and pulmonary artery systolic pressure of 60 mm of Hg.

PROCEDURE

Median sternotomy was done. After pericardiotomy, the presence of a left atrial appendage juxtaposed to the right atrial appendage and the right atrium was noted (Figure 1,2). Bicaval cardiopulmonary bypass with hypothermia at 28°C was established. Myocardial protection was achieved with intermittent hypothermic antegrade blood cardioplegia. Right atriotomy was done and trans septal vent was used to get clear filed and for deairing of heart. The basal muscular ventricular septal defect was closed by the treated pericardial patch (0.2% gluteraldehyde) via right atrial approach with single suture continues technique. The total cardiopulmonary bypass time and aortic cross clamp (ischemia) times were 55 and 22 minutes respectively. Patient shifted to ICU on ventilator on adrenaline and GTN infusion. The child was discharged on the 5th postoperative day. A follow-up echocardiogram after one week demonstrated good ventricular functioning; without any

residual shunt

DISCUSSION

Juxtaposition atrial appendages itself is a benign condition, but it may be associated with severe congenital heart defects with or without cyanosis as well as it may influence the technique and choice of therapeutic interventions. Wennerit suggested that left-sided juxtaposition is due to underdevelopment of torsion of the primitive cardiac tube and is a marker for the presence of cyanotic congenital heart disease while right-sided juxtaposition is due to over development of the torsion process of the primitive tube.^{2,3} A revision of 1,526 specimens of hearts with congenital heart defect, incidence of juxtaposition atrial appendages, was found 17 (1.1%) cases of atrial appendages juxtaposition with left juxtaposition in 15 (88%) and right juxtaposition in 2 (12%). Complete form was present in 11 cases and partial form in 6 cases.⁴ Left-sided juxtaposition atrial appendages is more common than right-sided juxtaposition atrial appendages by a ratio of 6: 1 to 8: 1.^{13,14} The detection of atrial juxtaposition is very difficult with imaging and more commonly made at the time of surgery or autopsy. While certain nonspecific finding on plain film x ray and electrocardiographic findings may suggest for it.⁵⁻⁷ Nevertheless angiography has been the only reliable means of preoperative recognition.⁸⁻¹⁰ Role of echocardiography is

Figure 1: Left Atrial Appendage Appears Pink While Right Atrial Appendage Appears Darker



still not well defined. However some clues may help in detection of this malformation on echocardiographic observation like horizontal orientation atrial septum with right atrial appendage displaced behind the great arteries and traveled toward the left over the left atrium. The left and right atrial appendages are both to the left of the great arteries and may be viewed from multiple transducer positions.¹¹ While failure to locate left appendage on echocardiographic study may guide for the presence of right juxtaposition.¹² The importance of diagnosis of this malformation in the pre intervention period is to avoid mistaking the atrial septal defect with opening of the right atrial appendage during surgery or in the patent foramen ovale during balloon atrial septostomy. It also helps in preoperative planning in Senning; difficult to perform and may even be contraindicated in patients with left juxtaposition like Mustard or Fontan procedures as well.¹⁵ Juxtaposition atrial appendages with stenosis at the site of the tricuspid valve can still be neglected in preoperative evaluation in patients with transposition of great arteries. In such a circumstance one may plan for switch without appreciating the tricuspid lesion. Therefore it is suggested that if Juxtaposition atrial appendages is detected during operation in case of a complete transposition planned for biventricular repair, the tricuspid orifice should be inspected first so that total cavopulmonary connection should be considered.¹⁶

CONCLUSION

It is important to diagnose above mentioned malformation in the pre intervention period to avoid mistaking the atrial septal defect with opening of the right atrial appendage during surgery or in the patent foramen ovale during balloon atrial septostomy.

REFERENCES:

1. Melhuish BP, Van Praagh R. Juxtaposition of the atrial appendages: a sign of severe cyanotic congenital heart disease. *Br Heart J* 1968;30:269-84.
2. Wenner O. Beitrage zur Lehre der Herzmissbildungen. *Virchows Arch Pathol Anat* 1909;196:127-68.
3. Dixon AS. Juxtaposition of the atrial appendages: two cases of an unusual congenital cardiac deformity. *Br Heart J* 1954;16:153-64.
4. Frescura C, Thiene G. Juxtaposition of the atrial

appendages. *Cardiovasc Pathol* 2012;21:169-79.

5. Freedom RM, Harrington DP. Anatomically corrected malposition of the great arteries. Report of 2 cases, one with congenital asplenia; frequent association with juxtaposition of atrial appendages. *Br Heart J* 1974;36:207-15.
6. Bream PR, Elliott LP, Barger LM Jr. Plain film findings of anatomically corrected malposition: its association with juxtaposition of the atrial appendages and right aortic arch. *Radiology* 1978;126:589-95.
7. Ho SY, Monro JL, Anderson RH. Disposition of the sinus node in left-sided juxtaposition of the atrial appendages. *Br Heart J* 1979;41:129-32.
8. Hunter AS, Henderson CB, Urquhart W, Farmer MB. Left-sided juxtaposition of the atrial appendages: report of 4 cases diagnosed by cardiac catheterization and angiocardiology. *Br Heart J* 1973;35:1184-9.
9. Deutsch V, Shem-Tov A, Yahmi JH, Neufeld HN. Juxtaposition of atrial appendages: angiocardiology observations. *Am J Cardiol* 1974;34:240-4.
10. Park MK, Chang CHJ, Vaseenon T. Congenital levojuxtaposition of the right atrial appendage. Association with persistent truncus arteriosus, type 4. *Chest* 1976;69:550-2.
12. Ghori MA, Alessandro S. Congenital absence of left atrial appendage: a case report and literature review. *J Saudi Heart Assoc* 2015;27:132-4.
13. Vidne BA, Subramanian S. Complete correction of transposition of the great arteries with left juxtaposition of the atrial appendages. *Thorax* 1976;31:178-80.
14. Becker AE, Becker MJ. Juxtaposition of atrial appendages associated with normally oriented ventricles and great arteries. *Circulation* 1970;41:685-8.
15. Anjos RT, Ho SY, Anderson RH. Surgical implications of juxtaposition of the atrial appendages. A review of forty-nine autopsied hearts. *J Thorac Cardiovasc Surg* 1990;99:897-904.
16. Leu MR, Chiu IS, Hung CR, Wu MH. Surgical implications of Juxtaposed atrial appendages and the associated anomalies. *Ann Thorac Surg* 1992;54:134-6.