

Right juxtaposition of atrial appendages with isolated ventricular septal defect

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SUMMARY

Juxtaposition of atrial appendages is a very rare congenital cardiac anomaly. It is not of much importance per se, but its identification pre-operatively usually indicates the presence of complex cardiac or extracardiac malformations. Left juxtaposition of atrial appendages is more common than right juxtaposition. Juxtaposition of atrial appendages is mostly associated with cyanotic congenital heart diseases and very rarely with acyanotic cardiac anomalies. We here present a case of right juxtaposition of atrial appendages in an infant associated with isolated ventricular septal defect.

Key words: Juxtaposition of atrial appendages – Left atrial juxtaposition – Right atrial juxtaposition – Ventricular septal defect

INTRODUCTION

Juxtaposition of atrial appendages is a rare congenital cardiac anomaly characterized by the presence of the two atrial appendages on the same side of the great arteries lying adjacent to each other with reported incidence of 0.28-1.1% in various pathological series (Van Praagh et al., 1996; Lai et al., 2001). It was initially classified by Dixon as left juxtaposition if both the atrial appendages were present on the left side of the great arteries; and right juxtaposition when the appendages are present on the right side of the great arteries

(Dixon, 1954). However, Van Praagh et al. (1996) classified the condition on the basis of the morphology of the appendages as follows: right atrial appendage juxtaposition, if the broad, triangular and trabeculated morphological right atrial appendage lies over the finger-like, slender and thin morphological left atrial appendage; and vice-versa with left atrial appendage. Van Praagh's classification becomes more advantageous in cases of heterotaxy and situs inversus. Right juxtaposition of atrial appendages is very rare and more commonly associated with cyanotic congenital heart disease (Van Praagh et al., 1996; Lai et al., 2001; Dixon, 1954; Frescura and Thiene., 2012), and very rarely with acyanotic congenital heart disease (Mathew et al., 1975; Boutayeb et al., 2017). We here present an infant with right juxtaposition of atrial appendages, which was operated for isolated muscular ventricular septal defect.

CASE SUMMARY

A 10-month-old male child presented with isolated mid-muscular ventricular septal defect (VSD), with inlet extension with moderate pulmonary hypertension for VSD closure. Intraoperatively, the child had situs solitus, levocardia, D-looped ventricles and normally related great arteries. The child was accidentally found to have the morphological left atrial appendage lying superior to the morphological right atrial appendage on the right side of the great arteries (Fig. 1). A well-defined large, mid-muscular VSD with inlet extension was present. There was no other VSD, atrial septal defect (ASD), patent ductus arteriosus (PDA) or pulmonary stenosis. VSD was closed with polytetrafluoroethylene patch uneventfully. The juxtaposed atrial appendages were recorded on the post-

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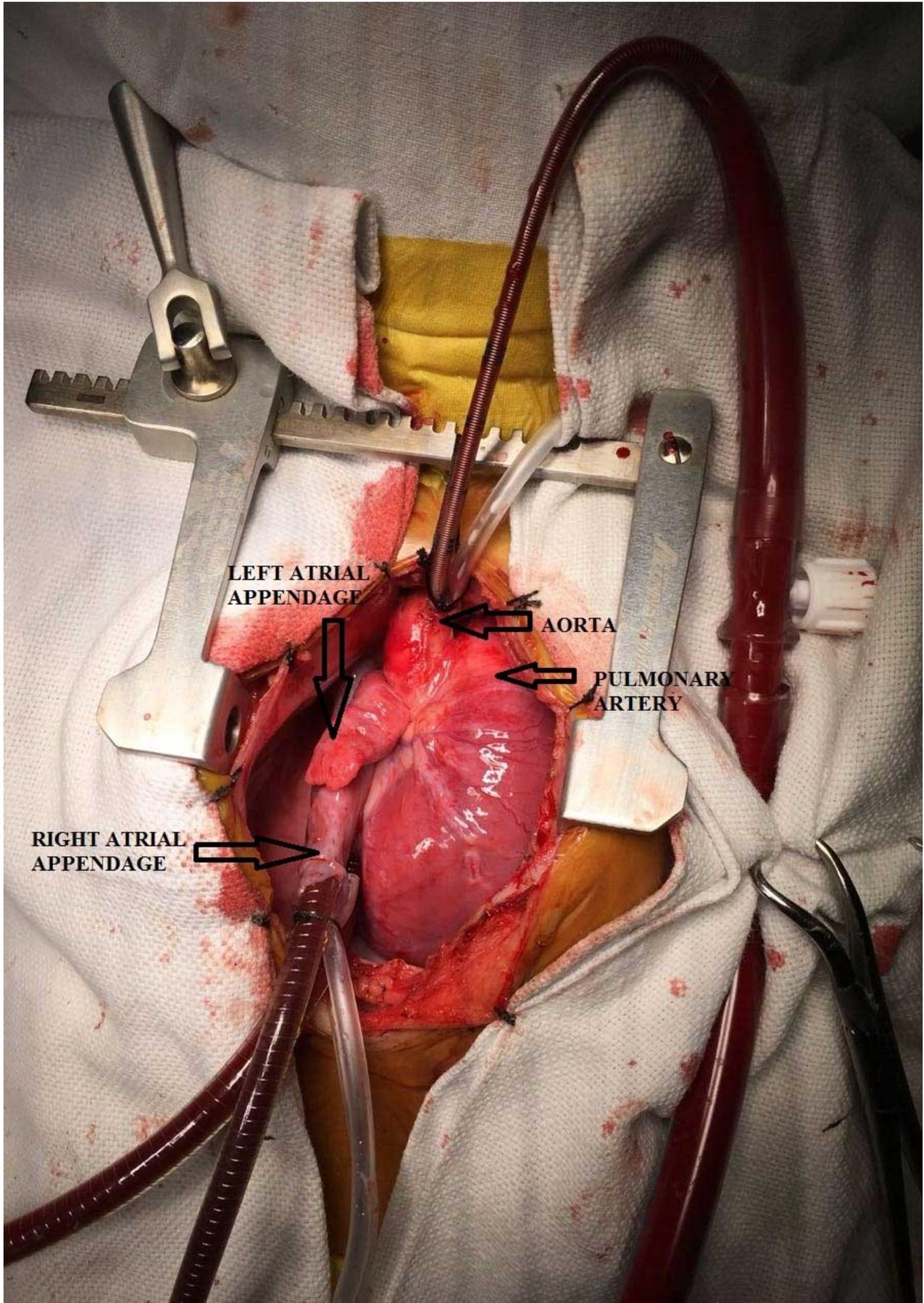


Fig 1. Intraoperative image showing left atrial appendage lying superior to the right atrial appendage (cannulated).

operative transesophageal echocardiography (Fig. 2).

An informed consent has been given by the patient for publication of the case report.

DISCUSSION

Juxtaposition of the atrial appendages was first reported by Birmingham in 1893 (Van Praagh et al., 1996), and was classified first by Dixon (1954), and then by Van Praagh et al. (1996). It is a very uncommon congenital cardiac malformation with left juxtaposition of the atrial appendages outnumbering right juxtaposition by 6:1 (Dixon, 1954; Frescura and Thiene, 2012). Van Praagh et al. (1996) reviewed post mortem cases of JAA, with 35 cases being juxtaposition of right atrial appendage (JRAA), and 18 being of juxtaposition of left atrial appendage (JLAA). Out of the 18 cases of JLAA, 2 had situs inversus and 16 were associated with situs solitus (i.e., left atrial appendage was present on the right side in situs solitus and on the left in situs inversus). They found striking differences between the two groups in terms of associated congenital cardiac anomalies. JRAA was more commonly associated with conotruncal and right ventricular outflow tract anomalies with hypo-

plastic right ventricle, unlike JLAA, in which the conus was almost always normal but had anomalies of left inflow and outflow tracts with left ventricle hypoplasia. Similarly, Lai et al. (2001) also found association of JRAA with conotruncal anomalies, and JLAA with complex atrioventricular anatomy in their clinical series of 22 patients. The rarity of JLAA can be gauged by the study of Frescura and Thiene (2012), who reviewed 1,526 specimens out of which only 2 cases were found to have right juxtaposition of atrial appendages, which were also associated with complex left side cardiac lesions. Also, Charuzi et al. (1973) found only 1 case of right-sided juxtaposition out of the 16 specimens studied for determination of congenital heart defects associated with juxtaposition of atrial appendages. None of the patients in these large series had isolated simple cardiac lesion associated with JAA.

Our case is unique due to the presence of rare combination of right sided juxtaposition of left atrial appendage with a simple, isolated ventricular septal defect without any other complex cardiac or extracardiac malformations in an infant. There are only five case reports of JAA with simple, isolated congenital cardiac defects as depicted in Table 1.

Echocardiography is the mainstay of pre-

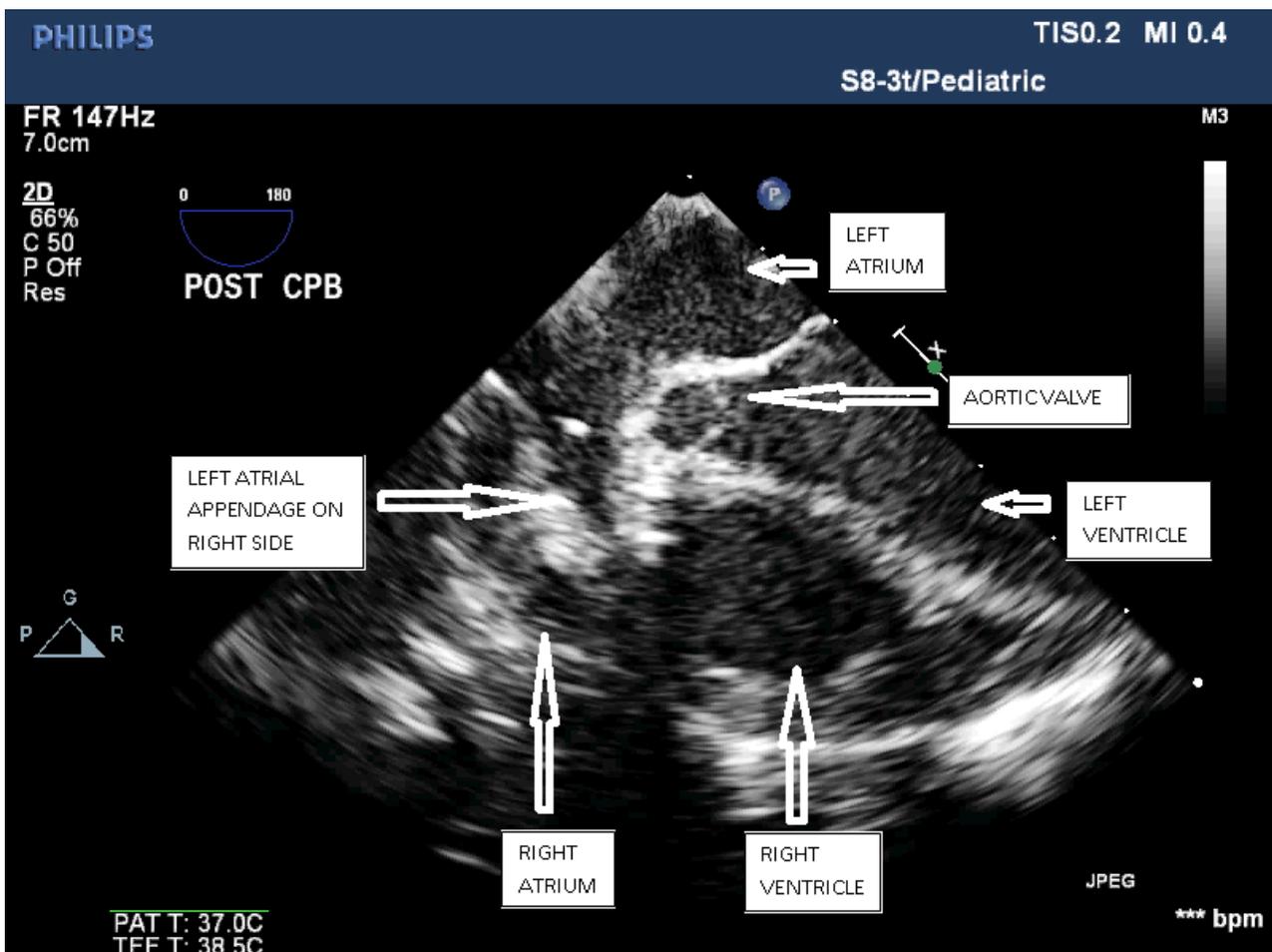


Fig 2. Post-operative mid-esophagus (0° view) transesophageal echocardiographic view showing left atrial appendage on the right side.

Table 1. Case reports of juxtaposition of atrial appendages with isolated, simple congenital cardiac defects.

S.NO.	AUTHOR	YEAR	ASSOCIATED CARDIAC LESION
1.	Becker et al.	1970	Atrial septal defect with bicuspid pulmonary valve; but it was a stillborn infant with lethal extracardiac malformations
2.	Mathew et al.	1975	Ventricular septal defect and patent ductus arteriosus
3.	Anderson et al.	1976	Atrial septal defect and persistent left superior vena cava
4.	Singhi et al.	2016	Ventricular septal defect
5.	Boutayeb et al.	2017	Bicuspid aortic valve with severe aortic stenosis
6.	Present case	2018	Ventricular septal defect

operative diagnosis of JAA characterized by the horizontal orientation of interatrial septum and abnormal location of atrial appendages on the same side (Singhi et al., 2016). Pre-operative diagnosis of right juxtaposition of atrial appendages was missed in our case on transthoracic echocardiography, mainly because of very low suspicion of its presence in an isolated, non-complex cardiac lesion. Pre-operative diagnosis of JAA is important, as its presence usually signifies the presence of complex congenital cardiac malformations, and also it may present some difficulties in some surgical procedures. In left juxtaposition of atrial appendages, atrial septum is transversely oriented and is displaced postero-inferiorly, which poses difficulty during balloon atrial septotomy (Tyrell et al., 1971), and also, due to small size of the right atrium, Senning procedure becomes very difficult and thus Mustard operation is recommended in such cases with transposition of great arteries (Leu et al., 1992). Right atrial cannulation in left juxtaposition of atrial appendages becomes impractical, and the superior vena cava has to be cannulated directly. Leu et al. (1992) advises evaluation of tricuspid valve for its stenosis in case of transposition of great arteries with JAA planned for biventricular repair.

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