

Two Cases of Atrial Aneurysms and Pectinate Muscle Dysplasia in Childhood

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Abstract

Primary atrial enlargement is a rare condition that can involve either the right or left atrium or both. Two such cases are described, and pathogenesis and treatment considerations are reported, concluding that surgical treatment does not necessarily exclude the possibility of late recurrence of supraventricular arrhythmias.

Keywords: Atrial aneurysm; Congenital heart disease; Arrhythmia.

Introduction

Primitive atrial enlargement is a rare condition that may involve right, left or both atria, without a universally recognized aetiology [1]. We report two cases detected in pediatric age: one of left atrial appendage aneurysm and one of idiopathic right atrial aneurysm (IDRA) [2], in which we postulate a congenital disarray and dysplasia in pectinate muscles as primary aetiology. Both patients received surgical treatment. Considerations are expressed on the efficacy of surgery in the long-term follow-up.

Case study

The first case, an 18-month boy, weighting 12.5kg, was admitted with the incorrect suspicion of metal object accidental ingestion. The chest X-ray highlighted an enlarged heart profile, ECG was normal.

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The chest X-ray highlighted an enlarged heart profile, ECG was normal. Echocardiography showed an isolated enlarged left atrial appendage (4x4.5cm). In full median sternotomy and extracorporeal circulation, the aneurism was resected at the base and directly sutured side to side in a double layer fashion. Post-operative course was uneventful.

Specimen examination revealed left atrial tissue with mild hypertrophy and severe focal thinning. At 8 years follow up the child is in NYHA class I, sinus rhythm, no new onset of atrial enlargement and normal ventricular contractility.

The second case, 15 years old patient, came initially to our attention for arrhythmic

symptoms. Ultrasound scan revealed an idiopathic extension of the right atrium and normal right ventricular volume, the tricuspid valve was severely incompetent. The standard 12 leads and 24 hour ECG evaluation showed the presence of symptomatic intra-atrial reentry tachycardia episodes with ventricular pre-excitation.

After two years of medical therapy with Sotalol and in presence of a progressive increase in right atrial size, the patient underwent surgery performed in extracorporeal circulation on the beating heart (Figure 1). At surgical inspection, the tricuspid valve presented a normal sized anterior leaflet, a large septal leaflet, a cleft of an hypoplastic posterior leaflet (Figure 2a), and the areas of leakage were the cleft itself and the commissure between the posterior and septal leaflets.

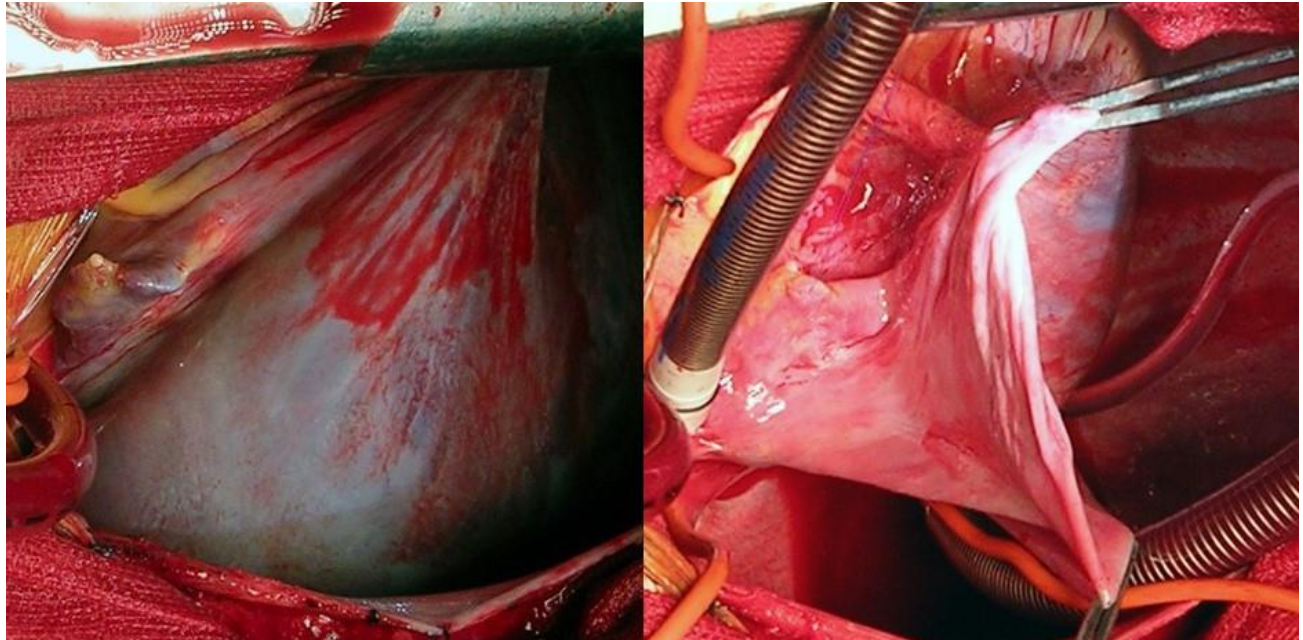


Figure 1: Enlarged and thin-walled right atrium before (a) and after (b) the institution of cardiopulmonary bypass.

“Bicuspidalization technique” consisted in suture closure of the cleft of the posterior leaflet and the commissure between anterior and posterior leaflets (Figure 2b). Moreover, a 30mm biodegradable annuloplasty ring was implanted and an EmiMaze right atrial ablation procedure was performed by

creating two isolation two lines, one between inferior vena cava and tricuspid annulus and the other between inferior and superior vena cava. Finally, a wide reduction of the right atrium was performed by 15x9cm appendage removal with pericardial lateral wall reinforcement (Figure 3).

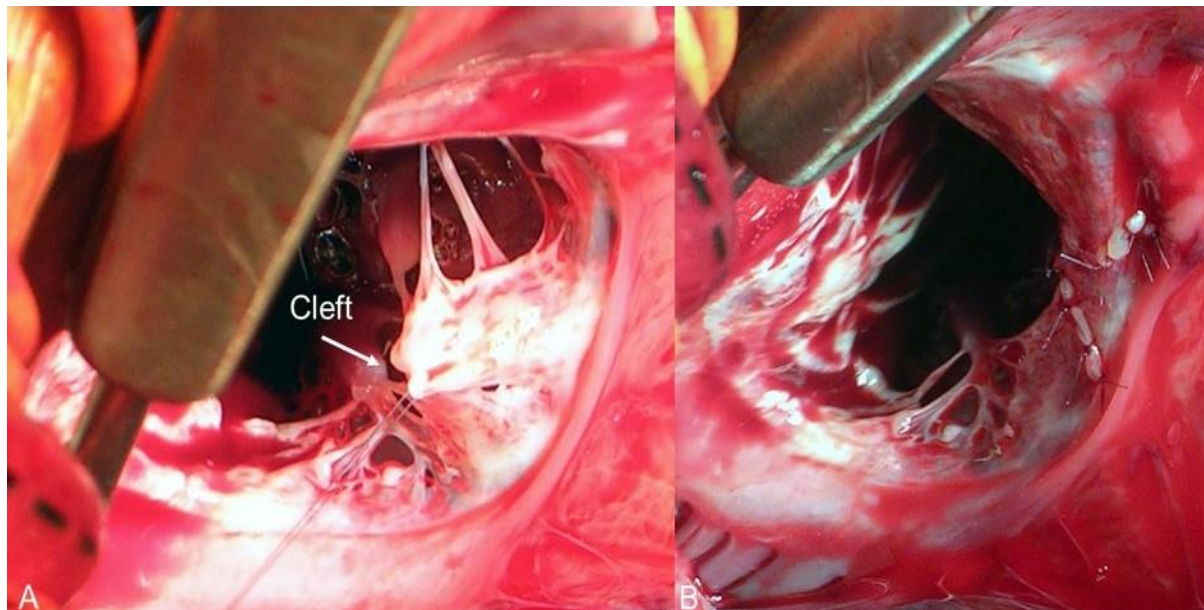


Figure 2: Appearance of the tricuspid valve before (a) and after (b) repair.

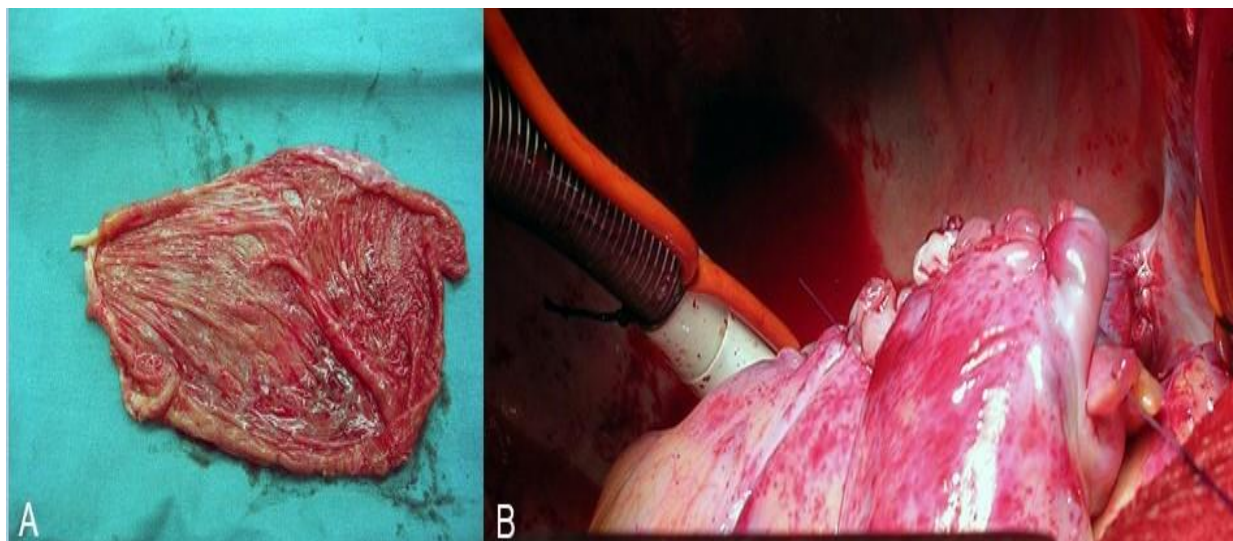


Figure 3: Right atrial wall excised (a) and (b) postoperative right atrial appearance.

Post-operative course was uneventful. No arrhythmias were recorded (Figure 4a) for 6 years after surgery, but subsequent onset of episodes of atrial tachycardia/atrial flutter (Figure 4b) occurred and were treated with radiofrequency ablation. Subsequent rhythm

control and non-inducibility were obtained with the Flecainide-Nadolol association. Mild to moderate stable tricuspid incompetence was recorded at eight years follow up. No new arrhythmic events were recorded since.

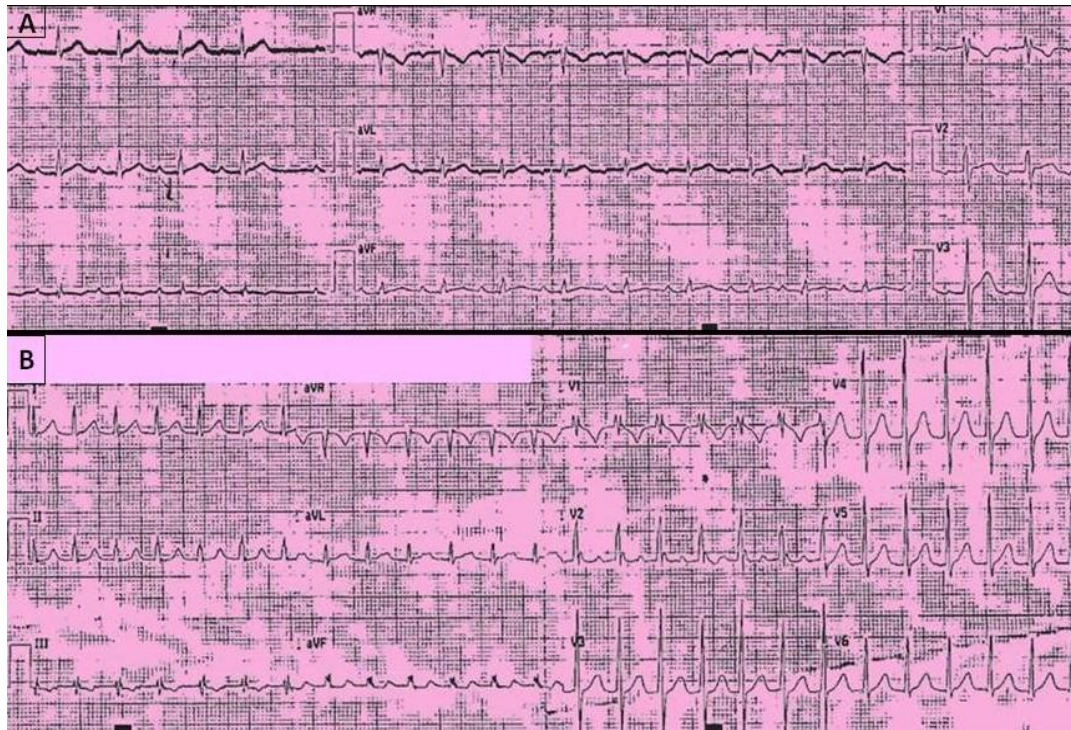


Figure 4: (a) Basic EKG trace without arrhythmia and (b) EKG trace with tachycardia.

Discussion

Left and especially right congenital atrial enlargements are rare in childhood [1].

The right atrium is characterized by an extensive array of pectinate muscles reaching the appendage, whereas the left atrium is relatively smooth walled. A crowd of embryogenesis of the primitive atria could result in the degeneration of the pectineal musculature exiting in marked thinning and bulging of the atrial wall. Right atrial aneurysm seems to be more prone to develop supra-ventricular arrhythmias.

Blondheim, et al. hypothesized that two types of IDRA may exist: one is secondary to a degenerative process of unknown etiology affecting the atrial myocardium (apoptotic process), the other consisting in a congenital absence of atrial myocardium and conduction tissue [2].

Interestingly, in our first case we depicted antenatally an abnormal L-R shunt over the foramen ovale, leading us to postulate a 'functional' partial abnormal pulmonary venous connection. This means that the pulmonary veins are normally connected to

the left atrium. However, there might be an uncommon angle of pulmonary veins insertion leading to a blood flow through the fossa ovalis.

Clinical presentation varies from asymptomatic forms to atrial arrhythmias including atrial fibrillation, atrial flutter, sustained supra-ventricular tachycardia [3].

The mechanism is the conduction tissue irritation caused by the cardiac chamber enlargement.

Symptoms range from palpitations, progressive dyspnoea, postural hypotension, atypical chest pain (due to enlarged left atrium compressing left coronary branches) and cerebral stroke for a thrombus in the abnormally dilated atrial cavity.

The acquired form of the left side is due to pathologies leading to high left atrial pressure as in mitral incompetence; otherwise, the congenital forms are probably caused by dysplasia of the pectinate muscles [4].

This hypothesis was confirmed by the second case specimen where was present thinned and disarrayed pectinate muscles, sometimes absent, giving to the atrial wall a fibrotic and translucent aspect. Somewhere the normal

three layers pattern was lost in favour of a fibro-cells monolayer.

Some cases reported were managed without extracorporeal circulation [5], and other centres prefer to perform surgery in cardiac arrest to be more precise in mass excision and thus avoiding left superior pulmonary venous obstruction.

In our second case the enlargement of right atria, as well as arrhythmia were still present after several years from surgery, probably due to an incomplete excision in the first procedure or a diffuse myocytes alteration which persists during the years.

Conclusion

Surgical treatment of atrial aneurysm aims to prevent the systemic embolic risk related to the atrial dilatation of the left auricle itself and/or to the arrhythmias that any type of atrial aneurysm can cause. However, the treatment of arrhythmias does not appear definitive, probably due to underlying structural alterations.

Declaration

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