Case Report

Single atrium - a rare case report

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Received: 14 May 2016
Revised: 06 June 2016
Accepted: 07 June 2016

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ABSTRACT

Single atrium is a rare congenital heart disease in which there is developmental absence of both septum primum and secondum part of atrial septum. Present study reports a case of an adolescent female patient with single atrium without any endocardial cushion defect and other variety of congenital heart disease. Clinically the patient presented with features of congestive heart failure. She underwent successful surgical separation of atrium.

Keywords: Single atrium, Atrial septal defect, Congenital heart disease

INTRODUCTION

Single atrium is a rare cardiac defect seen at birth in which both septum primum and secondum of atrial septum is absent.1 It is anatomically a separate entity from common atrium in which the association of endocardial cushion defect is common.2 The clinical presentation of a large secundum atrial septal defect, single atrium and common atrium is similar though the surgical challenge lies in letter two conditions where atrioventricular conduction bundle is at risk for injury. Present study reports a case of single atrium and describes present preferred safe method of surgical correction.

CASE REPORT

This thirteen years old girl has presented with exertional dyspnoea, easy fatigability, and palpitations for last four years duration. On clinical examination, patient was found to be mildly cyanotic. The chest radiograph revealed mild cardiomegaly and pulmonary plethora. The electrocardiogram showed incomplete right bundle branch block. The echocardiography revealed a large ASD of 44mm size with left to right shunt and mild to moderate pulmonary hypertension with intact interventricular septum. There was no abnormality at atrioventricular valves. Based on the pre-operative findings, patient was planned for reconstructive surgery for closure of the atrial septal defect.

Figure 1: Opened right atriotomy shows complete absence of interatrial septum with visualisation of anterior mitral leaflet (black arrow), tricuspid valve (red arrow), coronary sinus (blue arrow), conduction bundle of his area (yellow star) and imaginary line for attachment of normal atrial septum (green arrow).
Intraoperatively there was complete absence of atrial septum without any ramnent and normal both atrioventricular valves suggesting it were a case of single atrium without any AV cushion (Figure 1).

The defect was reconstructed with autologous pericardial patch. The lower edge of the patch was anchored to the tricuspid and mitral valve annulus, and to the tissues between the two atriovenricular valves by superficial interrupted sutures to avoid injury to the conduction system. The rest of the patch was sutured in a continuous fashion to the atrial wall, thus separating the two atria and leaving the coronary sinus in the right atrial side. Post operatively there was no conduction defects in electrocardiogram and echocardiogram showed no residual shunt across the neoatrial septum. The postoperative period was uneventful and the patient was discharged in stable condition. The patient has been on regular follow-up for last 1 year.

**DISCUSSION**

The terms common atrium and single atrium have been used interchangeably in literature. But single atrium, which is also known as Cortriloculare biventriculare, is one of the rare congenital anomaly in which there is complete absence of the atrial septum without an endocardial cushion defect.1

It is characterized by (i) complete absence of the atrial septum, (ii) absence of malformation of the AV valves, and (iii) absence of interventricular communication.2 On the other hand, the term common atrium (CA) is used to denote the condition where (i) complete absence of the atrial septum or it is represented by a small strand of tissue present at the superior atrial wall of the common chamber (ii) absence of interventricular communication, and (iii) an accompanying atrioventricular cushion defect.3 Atrial septal defects occur in 1 in 1500 live births, accounting for 10% to 15% of congenital heart defects in children and the incidence of common atrium among atrial septal defect patients is 3-4% only.4

The clinical presentation and physical findings of a patient with single atrium are similar to those of a large ASD at the level of the fossa ovalis.5 Effort intolerance, palpitation and recurrent respiratory tract infection are the usual presentations. There is a soft systolic murmur in the pulmonary area, and a fixed wide splitting of the second pulmonary sound.

The mixing of blood at atrial level is notably more in common atrium due to AV valve regurgitations. As a result these patients seem to show a decrease in exercise tolerance early in life, increased fatigability, and shortness of breath, mild cyanosis or obvious heart failure.4 5

A variable degree of cardiomegaly with normal left sided chambers, absence of pulmonary bay and plethora of peripheral branches of the pulmonary vasculature is seen in routine chest radiograph.5 A three-dimensional transesophageal echocardiography is more accurate then transthoracic and transesophageal two-dimensional echocardiography in defining anatomy of single atrium heart to rule out existence of classical partial AV septal defect or cortriatratium.6

In repair, the single atrium needs to be separated into two atria with a patch, such as a Dacron or an autologous pericardial patch. The special surgical consideration is to avoid the injury to the conduction system. Two sutting methods are used in the reconstruction of a new atrial septum.7 In the first method: the patch was sutured to the base of the mitral anterior valve, and then gradually shifted to the posterior wall of the left atrium and the remaining border of the atrial septum via the posterior border of the coronary sinus.

Suture line used in the second method is that the patch was sutured from the middle of the ventricular septal crest, upward to the tricuspid septal valve annulus, and downward to the base of the tricuspid septal valve, to the borderline between the tricuspid septal leaflet and the anterior leaflet, and then via the lateral side or left inner side of the coronary sinus to the wall of the right atrium and the remaining border of the atrial septum.

Some prefers to anchor the pericardial patch by superficial interrupted sutures to the tissues between the two AV valves.7 The stitches are placed so superficially so that they could be visualized through the endocardium.

In present patient part of the patch was anchored with interrupted suture to the tricuspid annulus then via the left inner side of the coronary sinus to the wall of the right atrium and mitral valve annulus, and a few very superficial stich was taken at the tissues between the two atrioventricular valves. T

aking a couple of suture at mitral and tricuspid valve annulus avoids the risk of superficial suture giving away. Also in this method we believe that the suture line remains away from the triangle of Koch which avoids the risk of injury to the any part of bundle of His. This method resulted in freedom from complete block and arrhythmia postoperatively.

**CONCLUSION**

The long-term survival of surgical correction of single atrium is good. One has to be utmost careful to avoid injury to the conduction system by keeping the sutures remote and superficial from the bundle. Mitral valve to be examined visually to rule out the presence of cleft in leaflets.

**Funding:** No funding sources

**Conflict of interest:** None declared

**Ethical approval:** Not required
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