Case Report

Single Atrium- A Case Report

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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. this report of rare case is to share our experience in surgery for Single atrium. Patient with single atrium with persistant left superior venacava (PLSVC) and left sided Inferior venacava (IVC) underwent gluteryldehye treated autologous pericardial patch closure, separating the two atria and leaving the coronary sinus, both PLSVC and Left IVC in the right atrial side and drainage of all four pulmonary vein into Left atrium. Post operatively there was no conduction defects in electrocardiogram and echocardiogram showed no residual shunt across the neoatrial septum. The long-term survival of surgical correction of single atrium is good. To avoid injury to the conduction system one has to be utmost careful by keeping the sutures remote and superficial from the bundle.

Key words: *Single atrium,Congenital heart disease, Conduction defect.*

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Introduction

Single atrium is a rare congenital heart disease in which there is developmental absence of both septum primum and secondum part of atrial septum.[1] Anatomically it is a separate entity from common atrium in which the association of endocardial cushion defect is common.[2] The clinical presentation of a single atrium , a large secundum atrial septal defect and common atrium is similar . There is risk of injury of atrioventricular conduction bundle during surgery. We report a case of single atrium and describes present preferred safe method of surgical correction.

Case Report

This eleven years old boy has presented with exertional dyspnoea and palpitations for last four years duration. He had history of recurrent respiratory tract infection since birth. On clinical examination, patient was found mildly cyanotic. There is a soft systolic murmur in the pulmonary area and a fixed wide splitting of the second heart sound. The chest radiograph revealed mild cardiomegaly and plethoric lung field. The electrocardiogram showed incomplete right bundle branch block. The echocardiography revealed a large ASD with absent of interatrial septum, mitral regurgitation GrI, Aortic regurgitation GrI, pulmonary hypertension (PASP=35). with intact interventricular septum. There was no abnormality at atrioventricular valves. (Figure 1) CT angiogram revealed large interatrial septal deffect with smaller right atrium simulating single atrium. Bilateral SVC, right sieded SVC draining into right side of common atrium and left sieded

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SVC draining into left side of common atrium. Bilateral IVC hepatic vein forms right sieded infra diaphragmatic IVC and drains into right side of common atrium.Both renal veins and abdominal organs (except liver) drains into left sieded IVC and drains into left side of common atrium.Adequate size pulmonary arteries. No major aortopulmonary collateral arteries (MAPCA).Bilobed lungs with left sieded liver,right sieded polysplenia and right sieded stomach (Figure 2)

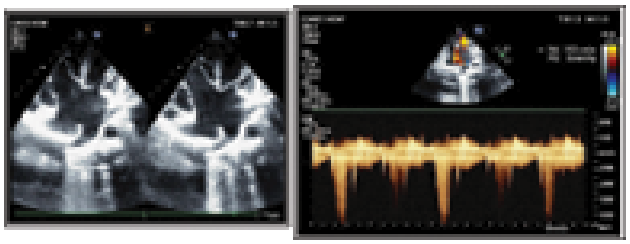

Fig 1: Echocardiography demonstrate Single atrium

Patient was planned for reconstructive surgery for closure of the atrial septal defect. CPB was established with aortic and venous cannulation (separately SVC, PLSVC, IVC and Left IVC cannulation done and connected with single venous tubing system by multiple Y connectors). Vent was placed at right superior pulmonary vein. Heart was arrested with intermittent antegrade cold bood cardioplezia.

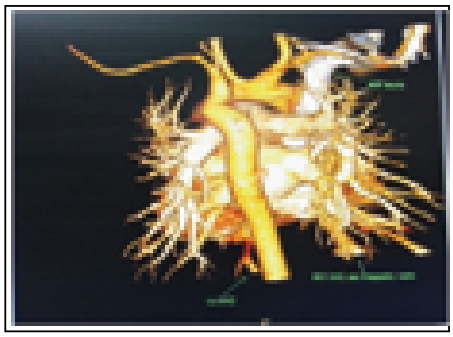

Figure 2: CT angiogram showing SVC, PLSVC and left sided IVC.

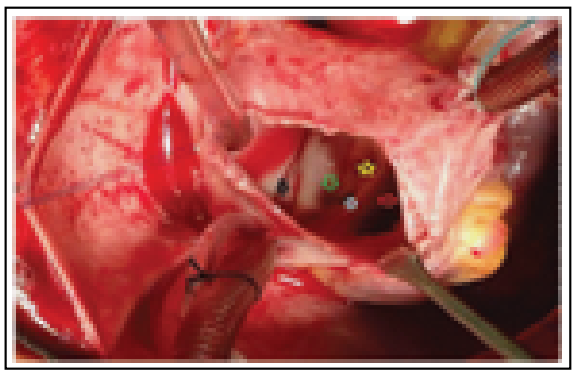

Figure 3: Opened right atriotomy shows complete absence of interatrial septum with visualisation of anterior mitral leaflet (black arrow), tricuspid valve (red arrow), coronary sinus (blue arrow), conduction bundle of his area(yellow star) and imaginary line for attachment of normal atrial septum (green arrow).

Intraoperatively there was complete absence of atrial septum without any ramnent , normal both atrioventricular valves suggesting it was a case of single atrium without any AV cushion defect, persistant left SVC (PLSVC) and left IVC. (Figure 3).

The defect was reconstructed with gluteryldehye treated 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 atrio-ventricular 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, both PLSVC and Left IVC in the right atrial side and drainage of all four pulmonary vein into Left atrium. 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. Postoperative follow up echocardiogram revealed inter atrial septum was intact with paradoxic IVS motion. Left atrium diameter was 41mm.Right atrium smallere size. PASP=27mm Hg. (Figure 3)

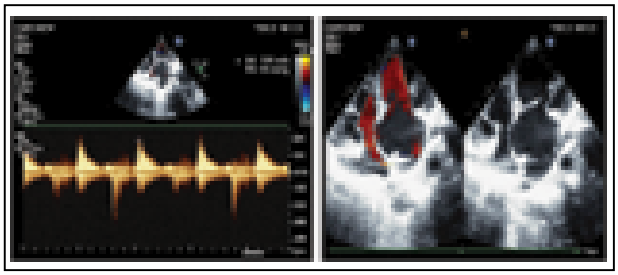

Figure 3: Echocardiography demonstrate intact inter atrial septum.

Discussion

Single atrium, also known as Cortriloculare biventriculare, is one of the rare congenital anomaly . 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.[1] On the other hand, the term common atrium (CA) is used to denote the condition where (i) complete absence of the atrial septum or 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.[2] Atrial septal defects occur 1 in 1500 live births. It is accounting for 10% to 15% of congenital heart

defects in children . The incidence of common atrium among atrial septal defect patients is 3-4% only.[3] 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.[3] The usual symptoms are palpitation, effort intolerance and recurrent respiratory tract infection. On examination, there is a soft systolic murmur in the pulmonary area and a fixed wide splitting of the second heart sound.The mixing of blood at atial level is more in common atrium due to AV valve regurgitaions. 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]

Chest radiograph findings are a variable degree of cardiomegaly with normal left sided chambers, absence of pulmonary bay and plethora of peripheral branches of the pulmonary vasculature.[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 cortriatriatum.[6] Echocardigram need to define anatomy of single atrium heart and to rule out existence of partial AV septal defect or cortriatriatum. The single atrium need to be separated into two atria with a patch, such as a Dacron or an autologous pericardial patch during repair. It is need to avoid the injury to the conduction system. For reconstruction of a new atrial septum two suturing methods are used.[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.[4] The stitches are placed so superficially so that they could be visualized through the endocardium. To avoid the risk of superficial suture giving away we take a couple of suture at mitral and tricuspid valve annulus. To avoid the risk of injury to the any part of bundle of His the suture line remains away from the triangle of Koch. There is no complete heart block and arrhythmia postoperatively.

Conclusion

The long-term survival of surgical correction of single atrium is good. To avoid injury to the conduction system one has to be utmost careful by keeping the sutures remote and superficial from the bundle.

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