A Case of Primum Type Atrial Septal Defect, Unroofed Coronary Sinus, Left Superior Persistent Vena Cava and Cor Triatrium. Surgical Intervention and Postoperative Findings

BİR PRİMUM TİP ATRİAL SEPTAL DEFEKT, UNROOFED KORONER SİNÜS, SOL SÜPERİOR PERZİSTAN VENA KAVA VE COR TRİATRİUM VAKASI. CERRAHİ MÜDAHALE VE POSTOPERATİF BULGULAR

Barlas N. AYTAÇOĞLU*, Mehmet ÖZKAN**, Tuncay AYVAZ**, Alp ASLAN**, Yaman ZORLUTUNA***

* Op.Dr., Dept. of Thorax and Cardiovascular Surgery, Medical School of Mersin Unıversity, Mersin
** Op.Dr., City Hospital, Dept. of Cardiovascular Surgery,
*** Doç.Dr., Dept. of Thorax and Cardiovascular Surgery, Ankara Numune Hospital, Ankara, TURKEY

Summary

Objective: Thirty four year old male patient was operated upon in our clinic with the diagnosis of a primum type atrial septal defect, unroofed coronary sinus, left superior persistent vena cava and cor triatrium.

Methods: Left superior persistent vena cava was ligated, the atrial septal defect was closed with a knitted dacron patch leaving the coronary sinus to drain into the left atrium. There were no complications detected in the early or late periods.

Conclusion: The right atrial approach to correct such anomalies is recommended. This report is presented to review the subject because such anomalies are very rare.

Key Words: Atrial septal defect, Cor triatrium, Unroofed coronary sinus, Left superior persistent vena cava

T Klin J Cardivascular Surgery 2003, 4:105-108

Özet

Olgu: Otuz dört yaşında erkek hasta kliniğimizde primum tip atrial septal defekt, unroofed koroner sinüs, sol superior perzistan vena kava ve cor triatrium tanılarıyla ameliyat edildi.

Metod: Sol superior perzistan vena kava bağlandı, atrial septal defekt knitted dakron yama kullanılarak koroner sinüs solda bırakılarak kapatıldı. Erken ve geç dönemlerde herhangi bir komplikasyon saptanmadı.

Sonuç: Bu tür anomalilerin tedavisinde sağ atrial yaklaşımlar önerilmektedir. Bu makale bu tür anomalilerin son derece nadir olmaları nedeniyle konunun yeniden gözden geçirilmesi amacıyla hazırlanmıştır.

Anahtar Kelimeler: Atrial septal defekt, Cor triatrium, Unroofed koroner sinüs, Sol süperior perzistan vena kava

T Klin Kalp-Damar Cerrahisi 2003, 4:105-108

Case Presentation

A thirty four year old male patient admitted to our outpatient ward. In his history he had vertigo, weakness, shortness of breath and rarely palpitation. His physical examination revealed almost normal findings but a second degree systolic murmur at the pulmonary and mesocardiac zones of the heart. His electrocardiography showed a sinusal rhythm with a first degree A-V block. Cardiothoracic ratio was normal in his telecardiography but the pulmonary conus was prominent and there was an increase in bronchopulmonary arborisation. Echocardiographic findings consisted of enlarged left and right ventricles, big atrial septal defect and pulmonary hypertension approximately 61 mm.Hg. The patient underwent left and right heart catheterization and oxygen saturation pressures and blood pressures were recorded (Table 1). The Q_p:Q_s was 3:1.

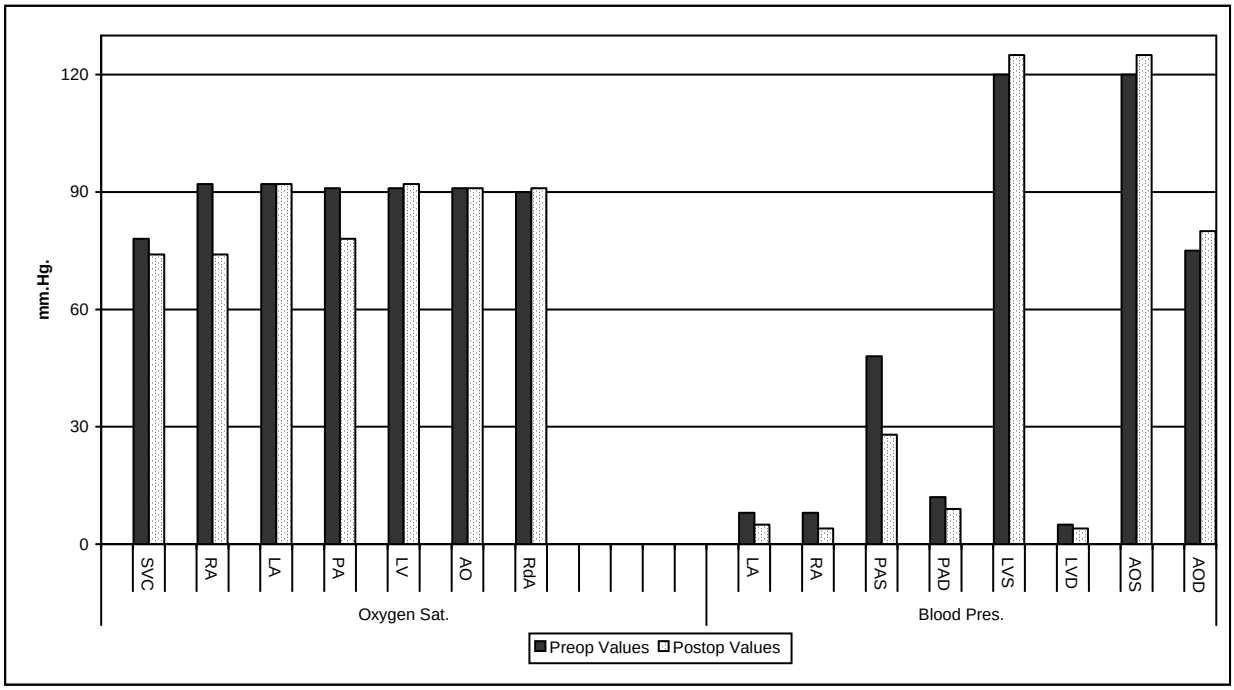
The patient was prepared for an operation aiming a total correction. Under neurolept aenesthesia a standart median sternetomy was performed.

Examination revealed a left superior persistenet vena cava. Measurement from this vessel showed a pressure of 9 mm.Hg. The patient was connected to the extra corporeal circulation with an aortic and bicaval cannulation and standard cardiopulmonary bypass instituted. A right atriotomy was performed under inflow occlusion. A quick exploration revealed a primum type atrial septal defect, unroofed coronary sinus and cor triatrium. There was a fenestrated membrane between the true left atrium and the chamber to which the pulmonary veins were draining. This membrane was excised. Left superior persistent vena cava was ligated externally. Atrial septal defect was closed with knitted dacron patch and the coronary sinus was left to drain into the left atrium.

Initially a cold crystalloid cardioplegia (St. Thomas II) and a totall of two blood cardioplegia (one cold and one as hot shot) were given. After the cardiopulmonary bypass period no positive inotropic support was needed. The patient was discharged from the hospital on the postoperative fifth day.

One month after discharge we found out that his vital findings were absolutely in normal ranges. In his physical examination there was no audible murmur and no clinical pathologic finding due to the ligation of left persistent superior vena cava. The patient was taken to the catheter laboratory and left and right cardiac catheterization was performed for a control. The pressures and oxygen saturations were recorded (Table 1). When the table is examined it is found out that there is no detectable

Table 1. Preoperative and Postoperative Oxygen Saturations and Blood Pressures in Different Chambers of The Heart.



SVC: Spureior Vena Cava
RA: Right Atrium
LA: Left Atrium
PA: Pulmonary Artery Pressure
LV: Left Ventricle
AO: Aorta
RdA: Radial Artery

LA: Left Atrium
RA: Right Atrium
PAS: Pulmonary Artery Systolic Pressure
PAD: Pulmonary Artery Diastolic Pressure
LVS: Left Ventricle Systolic Pressure
LVD: Left Ventricle Diastolic Pressure
AOS: Aortic Systolic Pressure
AOD: Aortic Diastolic Pressure

LA: Left Atrium
LV: Left Ventricle

Figure 1. Postoperative Control Catheterization Showing No Right Atrial Passage and The Third Chamber

complication due to leaving coronary sinus to drain into the left atrium. An opaque injection into the pulmonary artery showed that the third chamber into which the pulmonary veins are drained and the true left atrium are visualized but there is no opaque passage to the right atrium (Figure 1). The differences in the pressures and oxygen saturations after the operation are summarized in Table 1.

The patient was followed up to two years after the operation. In his echocardiography after two years there was no pathologic flow pattern and he had an ejection fraction of 64%. His vital findings were in normal limits.

Comment

Cor triatrium is a very rare anomaly (1-4). It is encountered in 0.1% - 0.4% among all of the congenital anomalies (1, 2). In its classical form the accessory left atrial chamber is the place where all of the pulmonary veins are drained into and the chamber is opened into the true atrium through a fenestrated membrane (1-4). This classical form of cor triatrium was first described by Church in 1968 (1, 2). Pulmonary venous congestion ensues since the membranous septum forms an obstruction to the flow of blood and causes supravalvular stenosis. The result is pulmonary hypertension, right ventricular hypertrophy and insufficiency. The symptoms arise due to this increase in pressures (1-5).

Cor triatrium is frequently encountered with accompanying anomalies. The most frequently seen anomalies are atrial septal defect (1) and left superior persistent vena cava (6,).

Persistent left superior vena cava is the most common anomaly encountered in the vena caval system (1, 2, 7). It is an accompanying pathology in 2 % of the patients with atrial septal defects (8).

Unroofed coronary sinus was first described by Raghip et al in 1965 (1, 2, 9). This is a very rare pathology (1, 9). It is frequently seen together with left superior persistent vena cava (1) and coronary sinus type atrial septal defect. It's diagnosis can be made with transthoracic and transoesophagial echocardiography (9). Furthermore cardiac catheterization can identify the pathology as it gives rise to a right to left shunt at the left atrial level (2).

In this case primum type atrial septal defect, left superior persistent vena cava, unroofed coronary sinus and cor triatrium were found all together. The clinical symptoms occurred due to the increase in pulmonary pressure caused by the left to right shunt and the fenestrated membrane between the true left atrium and the third atrial chamber.

Result

The treatment of these pathologies is surgical (2). In surgery right atrial and transseptal procedures are offered (2, 4, 10). We preferred this method of surgery in this case. The fenestrated membrane between the third atrial chamber and true left atrium was successfully excised. As left superior persistent vena cava was ligated externally there has been no technical difficulty in doing so. Leaving the coronary sinus to drain into the left atrium did not give rise to any clinical problems.

As a result, we think that such complex pathologies can be managed rapidly and effectively through right atrial and transseptal approach.

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Geliş Tarihi: 04.03.2002

Yazışma Adresi:. Dr. Barlas N. AYTAÇOĞLU
Mersin Üniversitesi Tıp Fakültesi
Araştırma ve Uygulama Hastanesi
Göğüs Kalp ve Damar Cerrahisi AD,
Zeytinlibahçe Caddesi,
33070 MERSIN
baytacoglu@isbank.net.tr