

Septum primum malposition defect: a rare congenital anomaly

Septum primum malpozisyon defekti: Nadir bir doğuştan anomali

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Displacement of the septum primum-leftward in atrial situs solitus or rightward in atrial situs inversus is termed as septum primum malposition defect. It appears to be responsible for the anomalous pulmonary venous drainage. This abnormality occurs predominantly in patients with visceral heterotaxy, usually with polysplenia, or rarely with asplenia or a normally formed spleen. Poor development and absence of septum secundum are considered to be responsible for the malposition of septum primum. Transthoracic echocardiographic recognition of the displacement of septum primum facilitates surgical management. In this article, we present an 18-year-old male case of an echocardiographic diagnosis of septum primum malposition defect and its repair technique.

Key words: Pericardial patch; septum primum; septum secundum; transthoracic echocardiography.

The septum primum malposition (SPM) defect is a very rare congenital anomaly.^[1,2] Displacement of the septum primum-leftward in atrial situs solitus or rightward in atrial situs inversus is known as the SPM defect, and it appears to be responsible for anomalous pulmonary venous drainage. This abnormality occurs predominantly in patients with visceral heterotaxy and is usually accompanied by polysplenia, although it can be rarely seen with asplenia or a normally formed spleen as well. Poor development or absence of septum secundum appeared to be responsible for the malposition of septum primum,^[1-3] but very few cases of SPM defect have been reported in the literature. This anomaly was first reported by Edwards in 1953 and confirmed by Moller et al. in 1967 but it did not attain its deserved recognition in the diagnosis of partial or total

Atrial situs solitusta septum primumun sola doğru veya atriyal situs inversusta septum primumun sağa doğru yer değiştirmesine septum primum malpozisyon defekti adı verilir. Bu defektin pulmoner venöz drenaj anomalisine neden olduğu düşünülmektedir. Bu anormallik, çoğunlukla viseral heterotaksi, genellikle polispleni ve nadiren aspleni veya normal şekilli dalağı olan hastalarda görülür. Septum sekundumun kötü gelişimi ve mevcut olmamasının, septum primum malpozisyonunun nedeni olduğu düşünülmektedir. Septum primumun yer değiştirdiği transtorasik ekokardiyografi ile saptanırsa cerrahi tedavi kolaylaşır. Bu makalede tamir tekniği ile birlikte septum primum malpozisyon defektinin ekokardiyografi ile saptandığı 18 yaşında bir erkek olgu sunuldu.

Anahtar sözcükler: Perikardiyal yama; septum primum; septum sekundum; transtorasik ekokardiyografi.

anomalous pulmonary venous return (TAPVR) into the right atrium (RA) until 1993.^[1-5] Diagnosis can be established by transthoracic echocardiography, and the deviation of the septum primum is best demonstrated with the subxiphoid coronal, apical four-chamber, and parasternal long-axis views.^[4] Herein, we describe the case of an 18-year-old male who was diagnosed with the SPM defect by transthoracic echocardiography and then underwent successful surgical treatment.

CASE REPORT

An 18-year-old male patient who had been categorized as New York Heart Association (NYHA) class II was admitted to our institute with shortness of breath on exertion. A physical examination revealed wide and fixed splitting of the second heart sound along



Available online at
www.tgkdc.dergisi.org
doi: 10.5606/tgkdc.dergisi.2013.6340
QR (Quick Response) Code

Received: December 14, 2011 *Accepted:* February 20, 2012

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Figure 1. A chest X-ray shows plethoric lung fields with prominent upper lobe pulmonary vessels and cardiomegaly with obliteration of the pulmonary bay.

with a grade 2/6 ejection systolic murmur at the pulmonary area. A chest X-ray showed plethoric lung fields with prominent upper lobe pulmonary vessels and cardiomegaly with obliteration of the pulmonary bay (Figure 1). In addition, an electrocardiogram revealed normal sinus rhythm, right axis deviation, and right ventricular hypertrophy with an rsR' pattern in leads V1 and V2. A transthoracic echocardiogram was done in the apical four-chamber and subcostal views (Figure 2), and the SPM defect was revealed. The patient was taken up for surgery based on these findings.

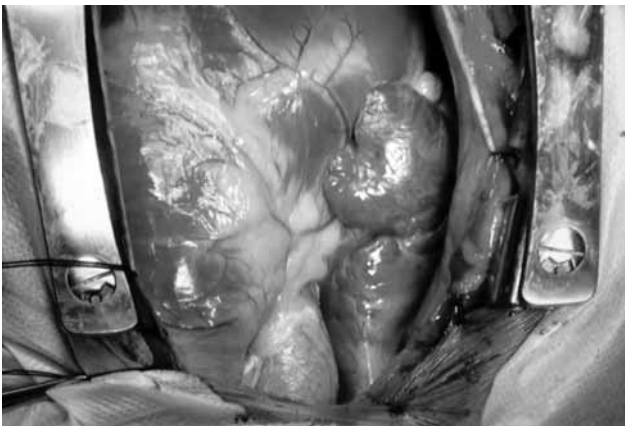


Figure 3. The external appearance shows the left atrial isomerism and the dilated right ventricle.

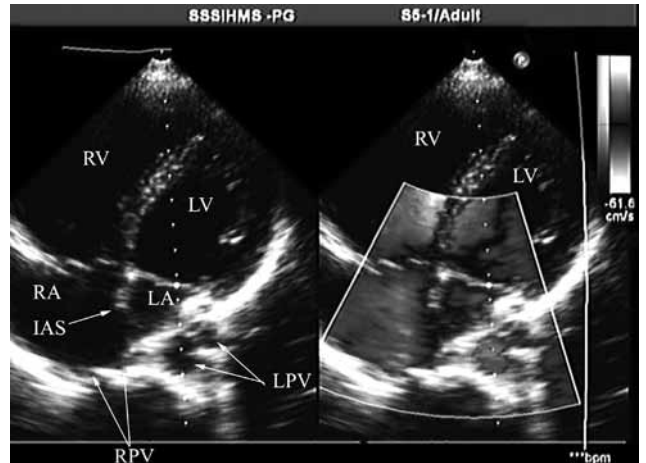


Figure 2. A transthoracic echocardiogram in apical four-chamber view shows the dilated right atrium and right ventricle, the small size of the left atrium due to the abnormally leftward-deviated interatrial septum, the left pulmonary veins draining into the left atrium, and the right pulmonary veins draining into the right atrium. 2D Echocardiography with color Doppler shows the pulmonary venous flow (grey) in the right and left atriums.

Surgical technique

Under general anesthesia, the conventional median sternotomy approach was used, and cardiopulmonary bypass (CPB) was established with aortic and bicaval cannulation. Under moderate hypothermia, the aorta was cross-clamped, and antegrade cold blood cardioplegia was applied to the aortic root. The intraoperative findings showed the left atrium (LA) isomerism, absence of an interatrial groove (Sondergaard's/Waterston's groove), and right pulmonary veins (RPVs) connected to the RA (external appearance) (Figures 3 and 4). The RA was opened



Figure 4. The absence of Sondergaard's/Waterston's interatrial groove is shown along with the right pulmonary veins connected to the right atrium.

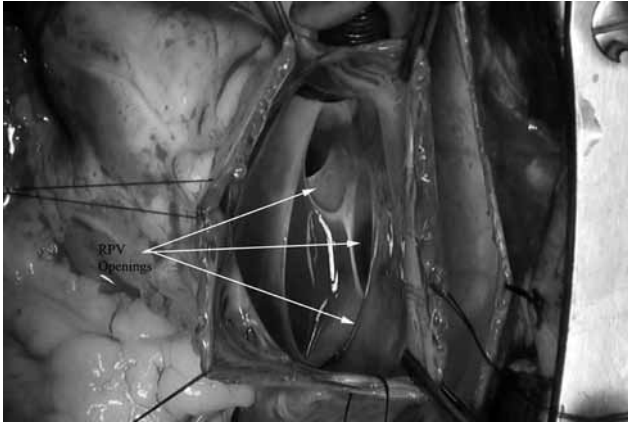


Figure 5. The right pulmonary veins are opening onto the posterior wall of the right atrium.

with a conventional oblique incision, and the anatomy showed that the RPVs were opening into the posterior wall of the RA (Figures 5 and 6). In addition, the atrial septum was displaced posteriorly and leftward with a small opening of about 1 cm in diameter at the upper end. This opening was the only communication between the right and left side of the heart. The tricuspid valve was normal, and the coronary sinus was intact and in its normal position. The posteriorly displaced interatrial septum was cut and the edges were endothelialized using 4-0 polypropylene sutures (Figures 7-10). An appropriately measured, autologous, untreated pericardial patch was used to reroute the RPVs into the LA. A new septum was created by suturing this pericardial patch in the RA using 4-0 polypropylene sutures (Figures 11-14). The patient was then rewarmed and weaned from the CPB in the usual way. The postoperative course was uneventful, and postoperative echocardiography showed unobstructed pulmonary venous return into the LA, no residual shunt, and good biventricular function (Figure 15).



Figure 7. A crescent-shaped communication at the upper end of the malpositioned septum primum.

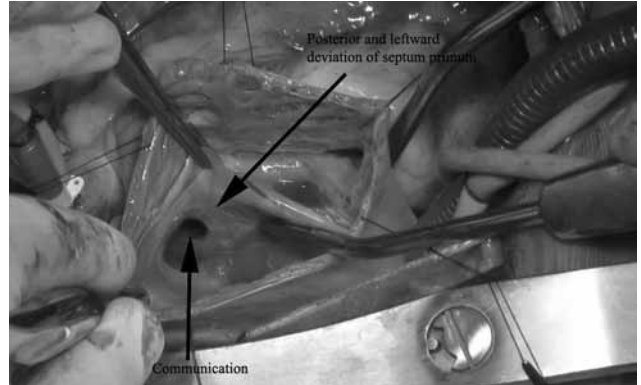


Figure 6. The interatrial septum is displaced posteriorly and leftward with about a 1 cm-sized crescent-shaped opening at the upper end of the septum communicating between the right and left atrium.

DISCUSSION

Normally, the septum primum grows from sinus venous tissue adjacent to the inferior vena cava (IVC)-RA junction and parallel to the left venous valve. When it completes its normal growth, it is attached on the LA side of the superior limbic band. It is the valve of the foramen ovale for fetal circulation and it forms part of the interatrial septum for the postnatal heart. The normal growth and attachment of the septum primum on the septum secundum are essential for the alignment of the common pulmonary vein with the cavity of the LA. If the superior limbic band of the septum secundum fails to develop, the cephalad crescent border of the septum primum remains unattached and could be carried leftward in cases of atrial situs solitus or rightward in cases of atrial situs inversus by the bloodstream of the fetal circulation which proceeds from the RA toward the LA. Depending on the degree of septum primum displacement toward the LA, half or all of the pulmonary veins may drain into the cavity of the



Figure 8. The septum primum is getting cut to enlarge the opening, which should be approximately the size of the mitral valve.



Figure 9. The raw cut edge of the septum primum is being endothelialized with 4-0 polypropylene suture.

anatomic RA, despite their normal connection to the posterior wall of the LA.^[1-3]

Van Praagh et al.^[1-3] speculated that the reason for the malposition of the septum primum might be poor development or the absence of the superior limbic band of the septum secundum. The high incidence of visceral heterotaxy with polysplenia in affected patients is consistent because polysplenia is known to be associated with the absence of this band.^[1-3] In our case, this absence was observed both via a transthoracic echocardiogram and during the operation. Our findings appear to confirm this theory by Van Praagh et al.,^[1] although the polysplenia in our patient was not diagnosed. Transthoracic echocardiography with Doppler interrogation is a reliable method for diagnosing this malformation, and the subxiphoid coronal, apical four-chamber, and parasternal long-axis views clearly demonstrated the deviation of the septum primum.^[4] Preoperative recognition of the true

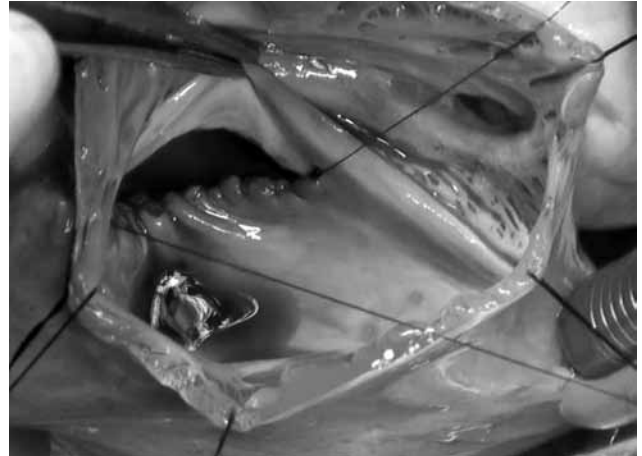


Figure 10. The final look of the reendothelialized margin and the size of the communication.

nature of this anomaly facilitates its successful surgical management. There are two other malformations in which the RPVs drain into the RA:^[1,2] (i) sinus venosus defects of the RA type with unroofing of the RPVs and (ii) large ostium secundum defects extending into the posterior border of the atrial septum that allow the RPVs to drain into the RA, although they normally are connected with the LA. These defects should be differentiated from atrial septal malposition because they require a different surgical treatment. In the case of the sinus venosus defects, a patch should be sutured between the upper border of the septum primum and the right border of the pulmonary veins. For the large atrial septal secundum defects with posterior extension, a patch should be used to close the defect, thus rerouting the pulmonary veins into the LA. Various surgical

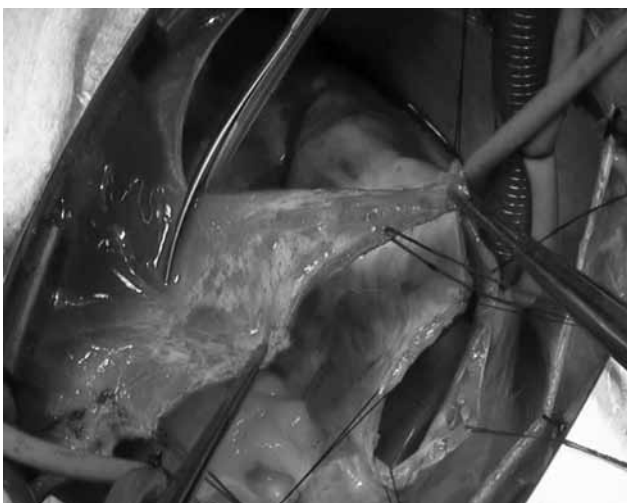


Figure 11. The appropriate size of the pericardial patch is being cut to create the neo-interatrial septum.



Figure 12. The suturing of the pericardial patch is started at the four o'clock position in the posterior wall of the right atrium. The pericardial patch is lowered, and the suturing is continued along the margin of the inferior vena cavae and right pulmonary vein openings towards the lateral wall of the right atrium.



Figure 13. The suturing is continued on the lateral wall of the right atrium up to the eleven o'clock position. The completed suturing of the pericardial patch on one side and the other end of the suture is continued on the cut edge of the enlarged septum primum margin.

techniques have been reported in the literature. Van Praagh et al.^[1] described a technique involving septum primum excision and the rerouting of the PVs to the LA using the pericardium or prosthetic material. Additionally, Hiramatsu et al.^[2] described in their series the use of the native septal tissue for rerouting the RPVs to the LA. This was done in order to avoid the use of the pericardium or prosthetic material so as to prevent pulmonary vein stenosis secondary to the thickness of whichever connective material was used. However, this technique is only useful in neonates or small children. It is necessary to use the pericardium or prosthetic material in adult patients as the septal tissue is inadequate to bridge the defect. This was seen in our case.

In conclusion, the SPM defect is a very rare congenital anomaly. Transthoracic echocardiography with Doppler interrogation provides a reliable method

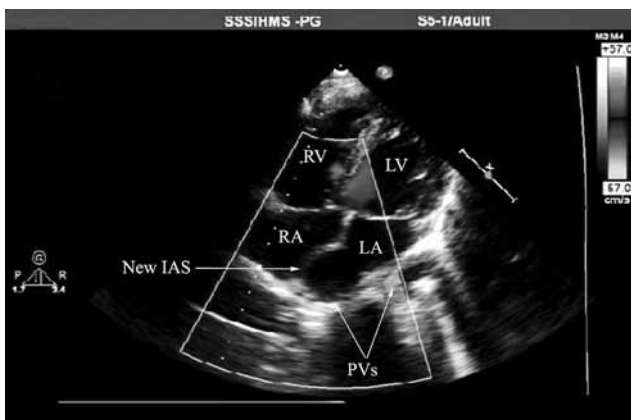


Figure 15. A postoperative transthoracic echocardiogram in the apical four-chamber view shows an unobstructed pulmonary venous return into the left atrium and no residual shunt.



Figure 14. A final look at the completed patch, or the neo-interatrial septum as it was being checked for a leak.

for the diagnosis of this malformation. The use of native septal tissue is feasible for rerouting the pulmonary veins to the left atrium in neonates and children, but the pericardium or prosthetic material should be used in adult patients.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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