Percutaneous Closure of the Atrial Septal Defect in a Patient with Dextrocardia and Situs Inversus Totalis

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ABSTRACT

Situs inversus totalis with dextrocardia and atrial septal defect is extremely rare. Different position and inversion types are required different closure techniques. We present a dextrocardia patient whose atrial septal defect was closed percutaneously. A 47-year-old female patient was referred for cardiological evaluation due to the diagnosis of situs inversus totalis and dextrocardia while preparing for gall bladder surgery. The patient's heart was located on the right side, right heart chambers were dilated and a secundum-type atrial septal defect of 15 mm in width was detected. A 20 mm atrial septal defect closure device (Occlutech, GmbH) was placed and the defect was completely covered. In this case report, we defined technical differences and issues that should be considered when planning interventions in dextrocardia patients.

Key Words: Atrial septal defect; dextrocardia; percutaneous closure; situs inversus totalis.

Situs İnversus Totalis ve Dekstrokardili Hastada Perkütan Yolla Atriyal Septal Defekt Kapatılması

ÖZ

Situs inversus totalis, dekstrokardi ve atriyal septal defekt ile birlikteliği oldukça nadir görülen bir patolojidir. Bu tür pozisyon yerleşim anomalisi olan hastalarda, defektlerin onarımı için farklı yaklaşımların kullanılması gerekir. Bu olgu sunumunda, atriyal septal defekti perkütan olarak kapatılmış bir dekstrokardi hastası sunulmuştur. Kırk yedi yaşında kadın hasta safra kesesi operasyonu hazırlığı sırasında situs inversus totalis ve dekstrokardi tanısı olması nedeniyle kardiyolojik değerlendirme için tarafımıza yönlendirilmiştir. Hastanın kalbi sağ tarafta ve sağ kalp boşluklarında dilatasyon izlenmiş, 15 mm genişliğinde sekundum tipi atriyal septal defekt saptanmıştır. Defekte 20 mm atriyal septal defekt kapatma cihazı (Occlutech, GmbH) yerleştirilmiş ve defekt tamamen kapatılmıştır. Bu olgu sunumuında, dekstrokardi hastalarında müdahaleler planlanırken hangi konulara dikkat edilmesi gerektiği ve teknik farklılıkları tanımlanmıştır.

Anahtar Kelimeler: Atriyal septal defekt; dekstrokardi; situs inversus totalis; perkütan kapatma.

INTRODUCTION

Atrial septal defect (ASD) is direct communication between the two atrial chambers which allows shunting of blood and is the most common pathology among congenital heart disease in adults. Epidemiologically, secundum (60%) defects are the most common type of ASD followed by primum (20%), and sinus venosus (15%), and lastly the coronary sinus (CS) (< 1%) defects[1]. Secundum type ASD is positioned by the fossa ovalis, primum ASD inferiorly as part of the spectrum of endocardial cushion defects, and sinus venosus ASD near the superior or inferior vena-cava entry[2]. The occurrence of this congenital anomaly has been reported to range between 1/6000 to 1/35000 live births and affects males and females equally[3]. The diagnosis is made by showing the defect in the atrial septum and shunt in transthoracic (TTE) and/or transoesophageal (TEE) echocardiography. When there are signs of significant volume overload of the right heart (i.e. a dilated right ventricle or shunt ratio > 1.5:1) the defect is usually closed[4].

Congenital heart diseases can be a single pathology or may be accompanied by more than one anomaly. Situs inversus totalis (SIT) and dextrocardia are extremely rare, and their association with ASD has been reported in few cases in the literature. In this case report, we present a dextrocardia patient whose ASD was closed percutaneously.

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CASE REPORT

A 47-year-old female patient was referred for cardiological evaluation due to the diagnosis of SIT and dextrocardia while preparing for gall bladder surgery. In her history, the patient described New York Heart Association (NYHA) class II exertional dyspnea and palpitations. Physical examination revealed a 2/6 systolic murmur at the right sternal border and a fixed splitting at S2. Normal sinus rhythm was observed in electrocardiography, no pathological finding was found. In TTE, the patient's heart was located on the right side, right heart chambers were dilated, and secundum type ASD was detected. Secundum type ASD of 15 mm in width in the interatrial septum was detected in TEE (Figure 1). The posteroinferior rim was measured as 12 mm, the posterosuperior rim as 6 mm, the anteroinferior rim as 5 mm, and the aortic rim as 6 mm. Qp/Qs was calculated noninvasively from echocardiographic flow parameters and resulting in 1.6 and percutaneous closure was planned. The procedure was started under sedation. A 7F sheath was placed in the right femoral vein. A Multipurpose (MP) 5F catheter was advanced into the right atrium with a 0.035 hydrophilic wire. The projection process and maneuvers were made as a mirror image of a standard procedure. Accompanied by TEE imaging, the ASD was passed from the right to the left atrium in fluoroscopy at LAO 20° and cranial 30° position. The hydrophilic wire was removed, and a 0.035 stiff wire was advanced through the MP catheter and settled in the pulmonary vein. The catheter and sheath were removed and the 12F delivery system was advanced to the left atrium over the wire. A 20 mm ASD closure device (Occlutech, GmbH) was loaded into the system and sent to the left atrium via a delivery catheter. The left atrial disc was opened in 130° short-axis viewing accompanied by TEE and the device was placed. Subsequently, the right atrial disc was opened in the anteroposterior position on the angiography (Figure 2). It was observed that there was no passage through the defect in TEE. The Minnesota maneuver was performed angiographically at the left anterior oblique 25°, caudal 20°, and in the TEE at 30° long axis, 90° bi-caval, 130° short-axis angulations with imaging guidance (Figure 3). After the device was well-placed and the defect was completely covered, the device

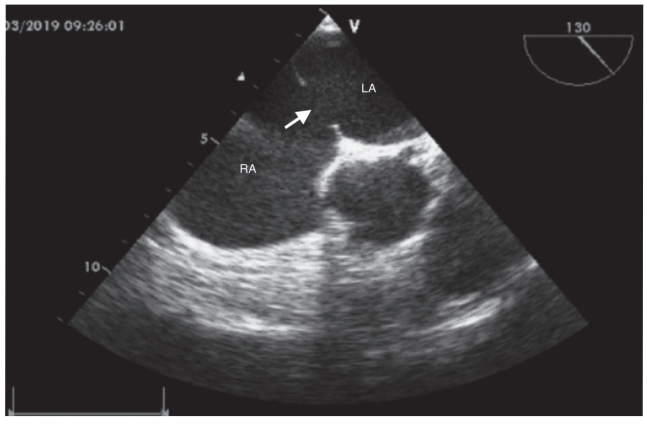

Figure 1. Transoesophageal short-axis view of atrial septal defect at 1300 (arrow) (RA: Right atrium, LA: Left atrium).

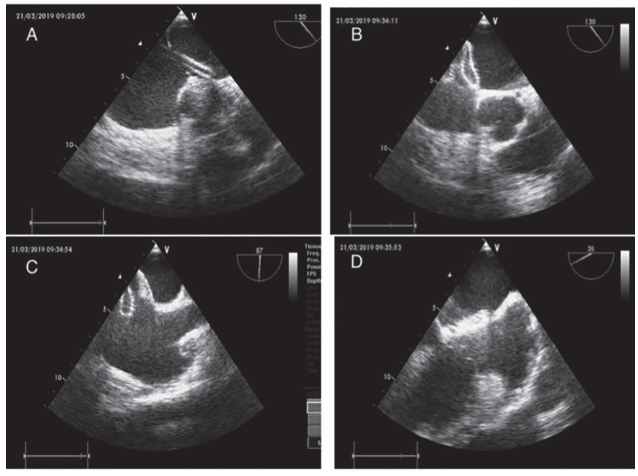

Figure 3. Transoesophageal views of placement of closure device and Minnesota maneuvers. A: Crossing of the delivery system from right to left in short-axis view, B: Left atrial disk opening of the closure device in short-axis view, C: Right atrial disk opening of the closure device in bi-caval view, D: Closure device in the long-axis view.

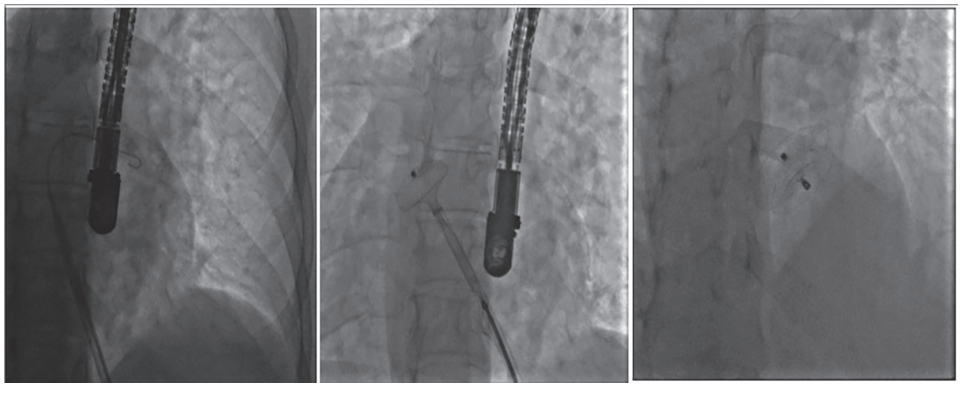

Figure 2. Fluoroscopic images of the placement of closure device.

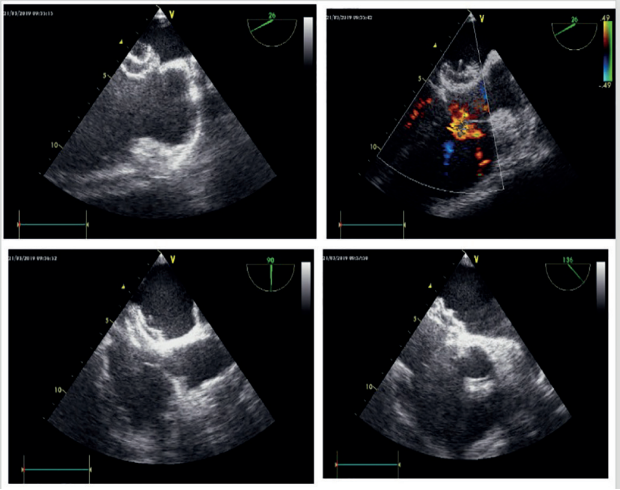

Figure 4. Transoesophageal views of closure device after placement.

was released (Figure 4). The carrier sheath and system were removed and a 10F sheath was placed in the femoral vein and the procedure was successfully completed.

DISCUSSION

In this case report, we present a successful percutaneously ASD closure procedure in a patient with dextrocardia and SIT. Dextrocardia is defined as a cardiac position anomaly that is a mirror image of normal anatomy. Dextrocardia with SIT is a rare congenital anomaly whereby the position of the abdominal and thoracic viscera is reversed.

In SIT, there is a complete right to a left reversal of all of the viscera including dextrocardia; the morphological right atrium is on the left and the left atrium is on the right. The normal pulmonary anatomy is reversed such that the left lung has three lobes, and the right lung has two[5]. Situs inversus with a left thoracic heart is associated with complex congenital heart disease. The most common subtype of this group is the situs inversus of the viscera and atria (I), L-loop ventricles (L), and normally related great arteries of the inverted type (I)[6]. Situs inversus with a right thoracic heart usually occurs with a structurally normal heart. Once the malposition has been identified, situs, ventricular loop and arterial connections should be evaluated. The interatrial septum plane is oblique in cases of levocardia, with the left atrium more posterior than the right atrium. In some dextrocardia cases, the interatrial septum is directed anteriorly and to the right, with the morphologic right atrium situated to the right and slightly posteriorly, and the morphologic left atrium to the left and slightly anteriorly that called dextroversion[7]. But in our case atrial and ventricular positions were placed mirror image to normal heart, the right atrium was placed anterior and the left atrium was placed posterior and superior. Therefore, we used mirror image planes in TEE and fluoroscopic views.

A few numbers of cases different closure techniques in child and young patients with dextrocardia and SIT were reported in the literature[8-12]. Galal et al. reported technical difficulties and solves in case situs solitus and dextrocardia that dextroversion[8]. In another case report that dextrocardia with interrupted inferior vena cava and ASD, left internal jugular vein was used. They reported that it is hard to fixation of the stiff guidewire because of dextrocardia[13]. Therefore, the classical femoral approach may be reasonable in dextrocardia patients if available.

Congenital heart defects usually can be diagnosed early because the SIT and ASD could be together with other congenital anomalies. In our case, ASD could not be detected until older age although the presence of SIT was known. Therefore, congenital anomalies should be investigated in patients with cardiac position anomalies whatever the patient's age.

CONCLUSION

In this case, it was shown that mirror images of standard procedures should be used when planning the interventions in dextrocardia patients and that the procedures can be performed successfully in this way.

Informed Consent: Informed consent form was obtained from patient.

Peer-review: Externally peer-reviewed.

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Conflict of Interest: The authors have no conflicts of interest to declare

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