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ASD Device Closure in Dextrocardia with Situs Inversus Totalis

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ABSTRACT
Device closure of the atrial septal defect in dextrocardia with situs inversus totalis has been reported previously. Dextrocardia with situs Inversus totalis in ASD makes the procedure complex.With proper imaging ASD device closure can be done in such cases. We present a case of a 19 years old female, who underwent successful ASD device closure for the second time in our center.

Keywords: *atrial septal defect; dextrocardia; transcatheter device closure.*

INTRODUCTION

An atrial septal defect (ASD) is an opening in the atrial septum, excluding a patent foramen ovale.[1] ASD is a common congenital heart defect, with an estimated birth prevalence of 1.6 per 1000 live births and a 97 % probability of survival into adulthood.[2] Dextrocardia with situs inversus is the most common variety of dextrocardia. Situs inversus with Dextrocardia or Situs inversus totalis occurs once in about 6-8,000 live births.[3] Among a subgroup of patients with situs inversus dextrocardia with concordant AV connections, ASD is relatively rare.[4] ASD can be treated with deice closure or with surgical closure. Though both approaches are effective, device closure is associated with lower mortality, complications, and length of hospital stay while surgical closure has a lower rate of residual shunting.[5] There are few case reports[6,-13] of ASD device closure in patients with dextrocardia and situs inversus totalis including one from Nepal.[14] We report a case of ASD device closure in dextrocardia with situs inversus totalis.

CASE REPORT

A 19-year-old female was referred to Shahid Gangalal National Heart Centre with the findings of dextrocardia on chest x-ray and an ASD and dextrocardia on echocardiography for ASD device closure. She complained of shortness of breath on and off, however, she has no history of orthopnea, swelling of legs, sore throat, joint pain, weakness of limb or facial deviation. There was no history of any cardiovascular disease in her family. A routine physical examination revealed a right-sided cardiac apex. Her vitals were stable with blood pressure of 100/70 mmHg and regular pulse of 81/min with normal volume and equally palpable in both limbs. Cardiac auscultation revealed splitting of the second heart sound at the right upper sternal border. The cardiac apex was palpable on the right fifth intercostal space along the mid-clavicular line. ECG showed sinus rhythm with Dextrocardia and the right bundle branch block.

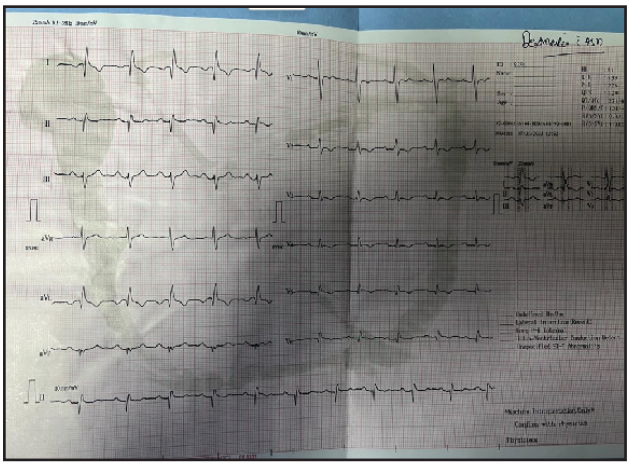

Figure 1. ECG.

Chest radiography confirmed that her heart was in the right chest and that the cardiac apex pointed to the right. The aortic arch was on her left, the mediastinal contours were normal, the lungs were clear, and the gastric bubble was on her right side. Her transthoracic echo findings were: abdominal situs inversus, dextrocardia, Inferior venecava/

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superior venecava draining into left-sided right atrium, pulmonary veins draining into the right-sided left atrium, Atrioventricular/Ventriculoarterial (AV/VA) concordance, great arteries relation normal, atrial septal defect secundum 30 mm in size, with shunt from left atrium to right atrium, mild tricuspid regurgitation (TR) with peak gradient of 30 mmHg with dilated right atrium and right ventricle and intact interventricular septum. A cardiac CT done by the referring physician showed Situs inversus, dextrocardia, ASD with dilated right-sided cardiac chambers, and normal pulmonary venous drainage. She was further evaluated with transesophageal echocardiography (TEE) which revealed dextrocardia with atrial septal defect, 3.0 cm in size with left to right shut. The aortic rim was absent while all other rims were adequate. As there was a dilation of RA and RV and ASD was suitable for device closure, we opted for ASD device closure.

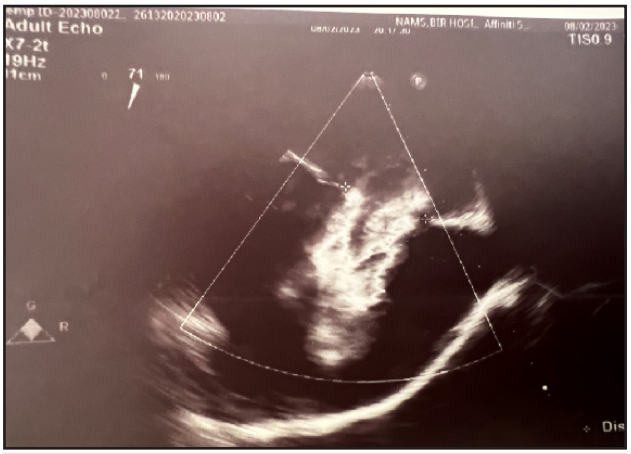

Figure 2. TEE bicaval view.

The procedure

The device closure procedure was done under local anesthesia with transthoracic echocardiogram guidance. The right femoral vein was cannulated, and after adequate heparinization, Judkins right (JR) catheter with Terumo wire was used to cross the ASD and JR was attempted to park in the pulmonary vein. Probing the pulmonary vein during the procedure was the most challenging because of the abnormal orientation compared to a normal heart. With difficulty, a JR catheter was placed in the right upper pulmonary vein and a 36mm Amplatzer septal occluder could be successfully deployed through the right upper pulmonary vein technique.

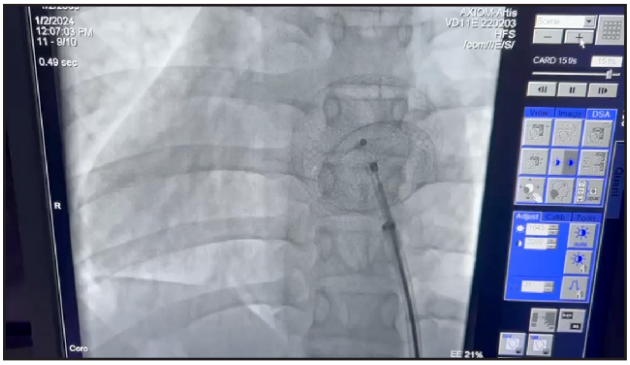

Figure 3. Device with delivery cable.

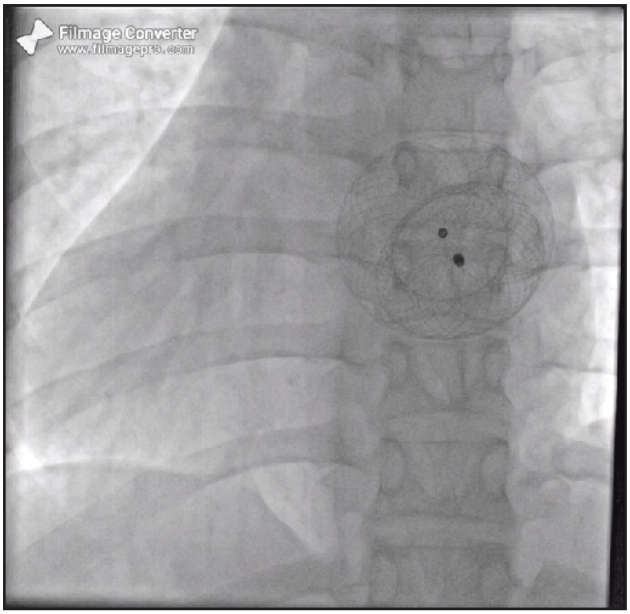

Figure 4. Device without cable.

Post-catheterization X-ray of the chest showed the device in a good position. The echocardiogram showed a good position with no residual shunt. Echocardiography 24 hours after the procedure showed the device in a good position with no residual shunt. Chest X-ray showed the device in good position at 3 months and 6 months after device closure. Echocardiography 3 months and 6 months after the procedure showed the device in a good position with no residual shunt.

DISCUSSION

This is the second case report of ASD device closure in dextrocardia with situs inversus totalis from our center. Experience gained in the previous case was instrumental in the successful device closure of this case. Dextrocardia is a rare congenital cardiac malformation with an incidence of 0.83/10,000

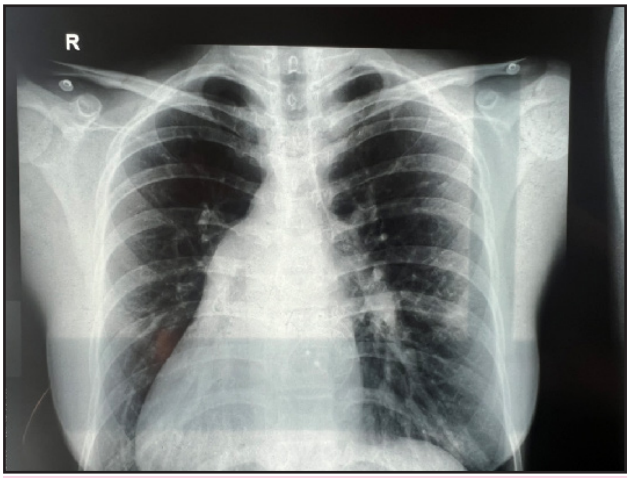

Figure 5. Chest X-ray AP view at 3 months.

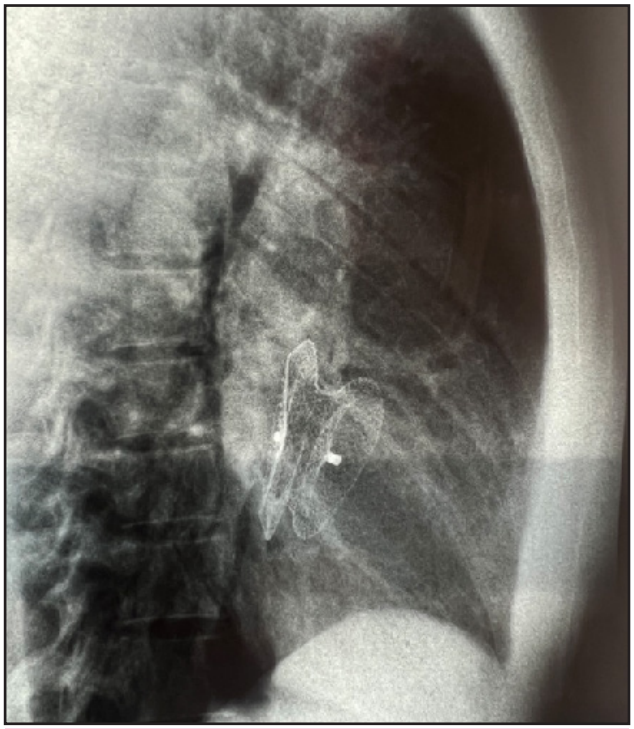

Figure 6. Chest Xray Lateral view at 3 months follow up.

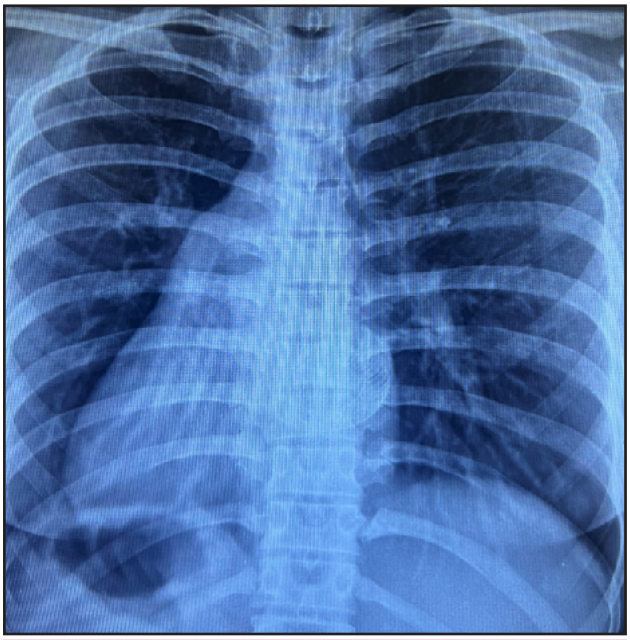

Figure 7. Chest X-ray AP at 6months of follow up.

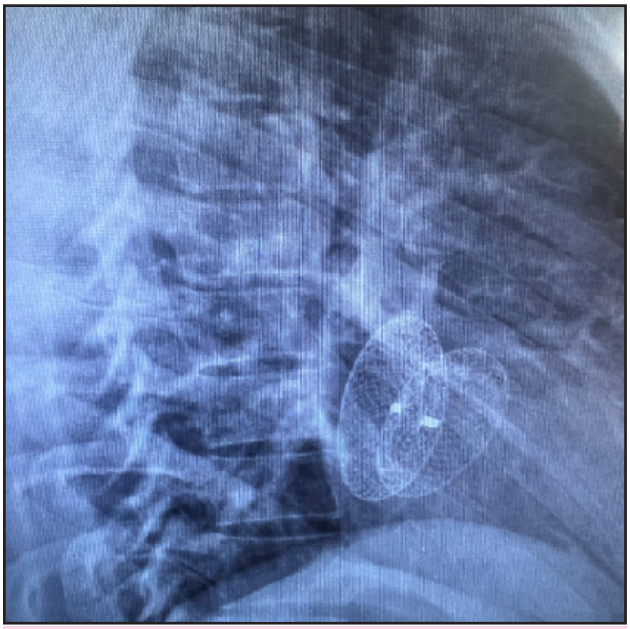

Figure 8. Chest Xray lateral at 6 months of follow up.

births, characterized by displacement of the cardiac apex to the right of the thoracic midline.[8,15] Dextrocardia with Situs inversus, also known as mirror-image dextrocardia, accounts for nearly 40% of all dextrocardia cases and is characterized by heart chambers opposite their normal positions. Dextrocardia with Situs inversus is the third most common type of dextrocardia in a series of autopsied cases.[16] Hakim et al. in 1998 described a successful ASD device closure in a dextrocardia patient by ASO device.[10] The authors suggested that difficulties encountered were due to heart position which was a mirror image of normal, therefore, the operators had to get used to this situation during the procedure.[10] In Situs inversus totalis, there is a complete right-to-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.[17] 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) .[16] 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.[18] 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 called dextroversion.[19] In dextrocardia with situs inversus totalis, ASD device closure is technically difficult due to unfamiliarity with the different radiological orientations of the heart and the need for modified views.[20] Difficulty lies in imaging and placement of the JR catheter in the pulmonary vein. Difficulty in imaging and placement of the catheter is due to abnormal heart orientation. Interventions in dextrocardia require modifications of imaging angles and in the catheter maneuvering techniques. Due to the unusual orientation of the heart (difference in morphology) and consequent different orientation of the common defects and the conduction system, there are unique challenges during interventions in the catheterization laboratory.[20] The "mirror image technique" which involves reversing the required right anterior oblique (RAO)/left anterior oblique (LAO) angles keeping the cranial/caudal tilts unchanged was used for ease of approach in such patients. The catheters can be passed using the standard technique, except that the catheters are rotated in the opposite direction compared to the patients with normal cardiac anatomy. The image inversion technique involves right-left image inversion as seen in a plain mirror. However, it still requires counter-directional torquing techniques and also a reversal of angiographic angles which leads to difficulty in interpretation of angiographic structures. To overcome this, a double inversion technique is used which involves right/left image reversal using a horizontal sweep reverse button on the machine and reversal of angiographic angle. As a result, the final images obtained resemble the normal angiographic images obtained in a levocardia patient. In the "double inversion technique,"[21]there is an artificial reversal of visualization of responses of catheters and wires to normal manipulation. A combination of right–left reversal of image on the monitor using the "horizontal sweep reverse" function during acquisition and a reversed RAO/LAO angle selection can help in better localization. The images acquired simulate normal cardiac anatomy and make the interpretation easy and avoid error. The advantage of the "double inversion technique" is a correction of unfamiliar angulated pictures in dextrocardia into the familiar conventional angiographic pictures of a normally located heart. This technique requires specialized software to be integrated.[20]

Muhammad Arif Khan et al suggest that device closure of ASD secundum in dextrocardia with situs inversus totalis is doable, but attention should be paid to the abnormal orientation of the interatrial septum and guidance by hand injections as well as the pressure tracing are occasionally more important than TEE.[7]

CONCLUSION

Our case highlights device closure of ASD secundum in dextrocardia with situs inversus totalis can be performed. Abnormal orientation of the interatrial septum should be taken care of with the proper use of imaging modalities like echocardiography during the procedure.

Conflict of interest: None

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