Intraoperative 2D and 3D Transesophageal Echocardiographic Assessment of Cor Triatriatum with Ostium Secundum Atrial Septal Defect in an Adult Patient

Abstract
Cor triatriatum is a rare congenital cardiac anomaly. Majority of the cases present in the childhood with an incidence of 0.4%. However, we report a case of cor triatrium sinister with ostium secundum atrial septal defect (ASD) in a 39-year-old female. The intraoperative 3D transesophageal echocardiography (TEE) offers an advantage over 2D TEE in visualizing the interatrial septum and the attachments of the fibromuscular accessory membrane in the left atrium (LA), which could help in surgical decision-making in this patient.

Keywords: Cor triatriatum, ostium secundum ASD, transesophageal echocardiography

Case
A 39-year-old female presented to the cardiology clinic with complaints of gradually increasing dyspnea. A 2D transthoracic echocardiography (2D TTE) revealed nonobstructive cor triatriatum sinister with ostium secundum atrial septal defect (OS-ASD) for which surgical repair was planned. After standard anesthesia induction, intraoperative transesophageal echocardiography (TEE) was performed using a commercial scanner (EPIQ7, Philips, Andover, MA) with fully equipped matrix array transducer X7-2t.

Initial scanning with 2D mode showed a large OS-ASD and membrane across the left atrium (LA) [Figure 1]. 2D color flow Doppler (2D CFD) imaging demonstrated free shunting of blood from left to right across the ASD. The 2D CFD across the membrane in LA did not reveal any turbulence [Video 1]. The 3D TEE zoomed data set of interatrial septum obtained from bicaval view demonstrated OS-ASD defect measuring 3.4 cm craniocaudal and 2.7 cm anteroposterior. The rims of the ASD were visualized well [Figure 2]. The 2D scanning of LA demonstrated a membranous band across the LA. A color flow Doppler (CFD) across the membrane did not reveal any turbulence. Left atrial biplane mode 0° and 90° demonstrated an extent of membrane across LA [Figure 3].

3D LA en-face view demonstrated its attachment and the fenestrations within the membrane as well [Figures 4 and 5, Video 2]. The right ventricle was dilated causing flattening of the interventricular septum. A CFD across tricuspid valve demonstrated mild tricuspid regurgitation. The calculated right ventricular systolic pressure was 35 mmHg.

Discussion
Triatrial heart is one of the rare congenital anomalies of the heart. Church first described it and the name “Cor triatriatum” was given by Borst. This rare abnormality of the heart is found in less than 0.1% of clinically diagnosed cardiopathies and 0.4% at autopsies.[1]

Left atrial involvement (Cor triatriatum sinister) is common than the right atrium (triatriatum dexter). It is thought to be because of failure of common pulmonary vein to incorporate into LA while another theory suggests it as an abnormal growth of the septum primum.[2]

The diagnosis of cor triatriatum is established by diagnostic modalities like echocardiography, cardiac computed tomography (CT), or magnetic resonance imaging. However, an accurate preoperative assessment is critical in planning surgical repair, and TEE showed its advantage over 2D TTE.

Rahul Shabadi, Pushkar Desai, Suresh Chengode
Department of Anesthesia and Intensive Care, Sultan Qaboos University Hospital, Muscat, Sultanate of Oman

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Address for correspondence:
Dr. Rahul Shabadi,
Department of Anesthesia and Intensive Care, Sultan Qaboos University Hospital, Muscat, Sultanate of Oman.
E-mail: drrahool@yahoo.com

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imaging (MRI) scan. Echocardiography determines the extent of the membrane in the LA, identifies its attachment, and assesses any obstruction of blood flow across it. It also helps diagnose associated cardiac anomalies. Thus, echocardiography is pivotal in determining surgical intervention.

However, the evaluation of an accessory membrane might be challenging with 2D TEE alone. In this regard, 3D echocardiography with an improvised spatial orientation helps to assess the size and the number of fenestrations in the partition membrane and delineates the anatomical features of the membrane in different planes.1,4

There are three types of cor triatriatum based on the number and size of fenestrations in the fibro-muscular membrane.2

- Type 1: No connection between two chambers
- Type 2: One or a few small openings (fenestrations) in the fibro-muscular membrane
- Type 3: Large single opening causing free communication between two chambers.
This congenital anomaly is often associated with ASD and anomalous pulmonary venous drainage into the right atrium. Uncommon associations like tetralogy of Fallot (TOF), double outlet right ventricle, and bicuspid aortic valve have also been reported. Type 1 and 2 presents with symptoms similar to mitral stenosis.\textsuperscript{[1]} Our patient had associated secundum ASD and was diagnosed as Type 2 cor triatriatum. On the first look, the surgeon mistook the large defect in the left atrial membrane for the ASD because of nearly absent interatrial septum, but based on the 3D TEE findings, a thorough examination later revealed the exact cardiac anatomy of the patient.

**Conclusion**

Combined 2D and 3D echocardiography provides superior scanning of the cor triatriatum, and it plays a key role in surgical decision-making.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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