

Case Report

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Intraoperative 2- and 3-Dimensional Transesophageal Echocardiographic Guidance for Cardiovascular Surgery in a Patient with a Single Papillary Muscle “Parachute Valve”

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Introduction

Parachute Mitral Valve (PMV) is a rare congenital cardiac defect characterized by focalized attachment of the chordae tendinae of both leaflets to a single papillary muscle [1]. It may occur as an isolated lesion or part of a Shone's anomaly in infants and young children [2]. Not much is known about PMV in adults [3]. There have been reports of complete Atrioventricular Septal Defects (AVSD) associated with PMV in children, but a partial or transitional-type AVSD [4]. A combination of 2- (2D) and 3-dimensional (3D) transesophageal echocardiography (TEE) is essential to guide these complex surgical procedures in children and, more rarely, in adults [5].

Case Description

A 55-year-old white male with new onset of atrial fibrillation, left hemiparesis, and a subsequent myocardial infarction presented for elective AVSD repair and coronary artery bypass graft. His medical history was also significant for peripheral neuropathy, Charcot-Marie-Tooth disease, diabetes mellitus, and hyperlipidemia. Transthoracic Echocardiography (TTE) revealed what appeared to be an AVSD with a large ostium primum Type Atrial Septal Defect (ASD) component and a restrictive Ventricular Septal Defect (VSD). The shunt across the ASD was predominantly left to right with a left mitral valve cleft and preserved ventricular systolic function. Cardiac catheterization demonstrated coronary artery disease.

After an uneventful anesthesia induction, the patient was placed on Cardiopulmonary Bypass (CPB) under mild hypothermic conditions. Intraoperative TEE confirmed the preoperative diagnosis of AVSD. 3D TEE further revealed the leaflets to be very primitive and thickened with no evidence of VSD (Figure 1). The right and left Atrioventricular Valves (AVV) were on the same plane, confirming the AVSD anatomy, and the direction of the right AVV regurgitation was toward and through the large primum ASD. The anterior mitral leaflet was larger and the cleft was oriented so that it pointed toward the septum and right ventricle (Figure 2). There was a mild mitral regurgitation through the cleft. The aortic valve was wedged between the left and right AVV. There was no evidence of left heart outflow obstruction. Further examination revealed a single papillary muscle with all the primary chordae originating from and supplying the left AVV structure (Figure 3). The chordae tendinae were short and thick. They were oriented in a posterior-to-anterior direction and converged on a centrally placed, single papillary muscle. The surgeon's direct visual inspection confirmed these findings. The surgical team decided against closing the cleft as planned based on concerns that this could result in mitral stenosis in the future.

The primum ASD was closed with a piece of bovine pericardial patch. Once the coronary bypass grafting was completed, the patient was successfully separated from CPB with milrinone and epinephrine

infusions. TEE performed after CPB revealed complete closure of the ASD. There was trace left AVV and mild right AVV regurgitation. After an uneventful clinical course, the patient was discharged to a rehabilitation center five days after surgery.

Discussion

Pre- and intraoperative anatomical and structural delineation of the mitral valves and subvalvular apparatus are paramount to achieving successful and durable repair. Intraoperative TEE has a significant impact on both the surgical care and anesthetic management of the patient [6]. There are several 2D but few 3D echocardiographic reports

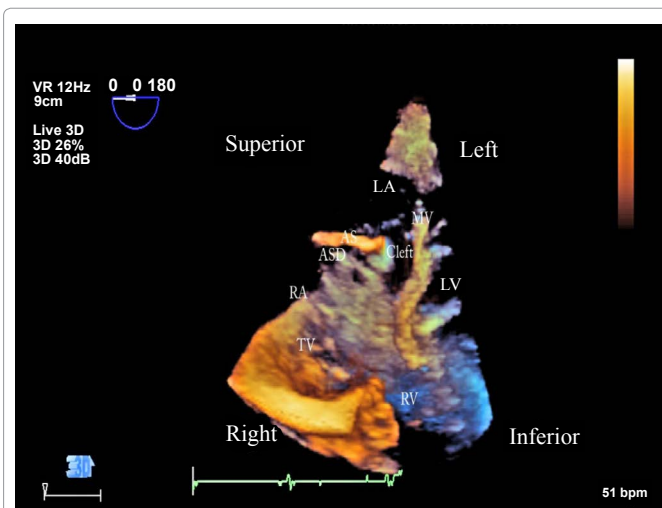


Figure 1: Midesophageal 4-chamber view of 3D transesophageal echocardiography showing primitive and thickened atrioventricular valves. The mitral and tricuspid valves are on the same plane. Most of the atrial septal defect (ASD) and the superior aspect of the atrial septum (AS) are included in this 3D image. The mitral cleft is visible through the ASD. Right Atrium (RA); Right Ventricle (RV); Left Atrium (LA); Left Ventricle (LV); Mitral Valve (MV); Tricuspid Valve (TV).

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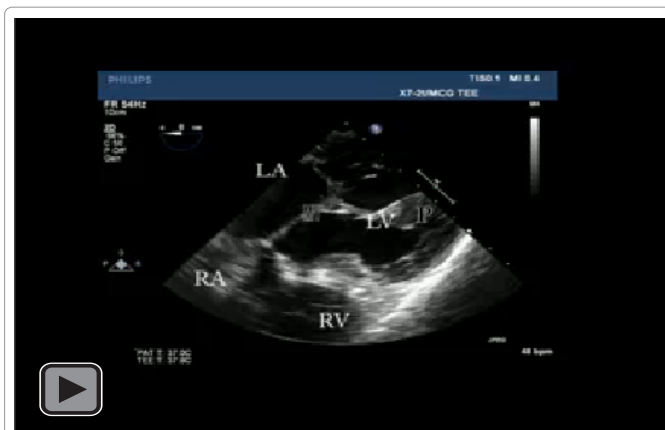
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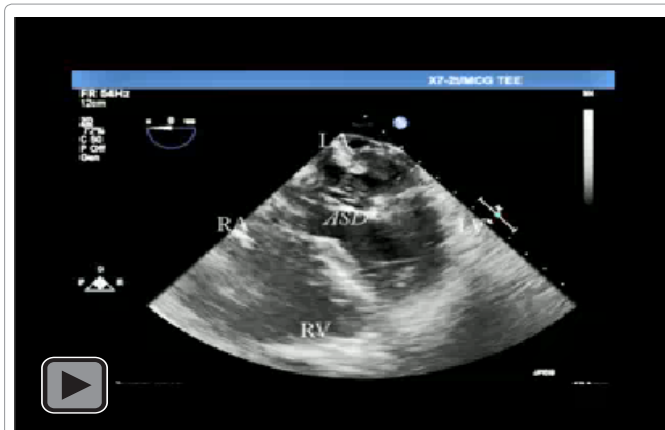
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of AVSD [7-8]. While 2D echocardiography provides clearer pictures, 3D TEE enables better visualization to determine the mechanism and site of AVV regurgitation. The combination of 2D and 3D real-time TEE has been shown to improve diagnostic accuracy from 40.4% to 65.4% [5]. In our case, the intraoperative TEE findings changed the surgical plan, potentially averting the adverse effects of mitral stenosis including heart failure, stroke, arrhythmia, and endocarditis.

PMV is caused by the defective delamination of the anterior and posterior parts of the trabecular ridge between the 5th and 19th weeks of gestation [9]. The underdeveloped chordae tendinae decrease mobility of the valve leaflets and reduce the mitral orifice. The narrowing of the interchordal spaces results in a smaller secondary mitral orifice, thus causing mitral inflow obstruction. The degree of stenosis is progressive and dependent on the tethering of the leaflets and reduction of the distal orifice. Most patients present during infancy with mitral stenosis of varying severity, or, less frequently, mitral regurgitation [10]. The



Video 1:



Video 2:

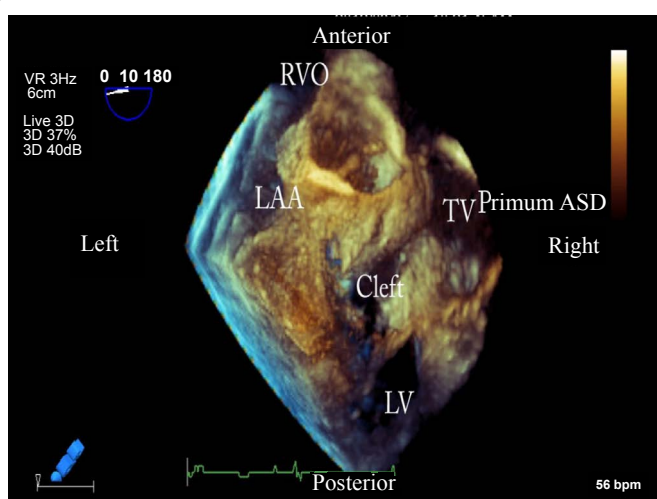


Figure 2: 3D view of mitral valve cleft and wedge aortic valve. It is a surgeon's view. Tricuspid Valve (TV) can partially visible through the primum ASD. Right ventricular outflow (RVO) track is behind the aortic valve (AoV). Parts of Left atrial appendage (LAA) and left ventricle (LV) are visible.

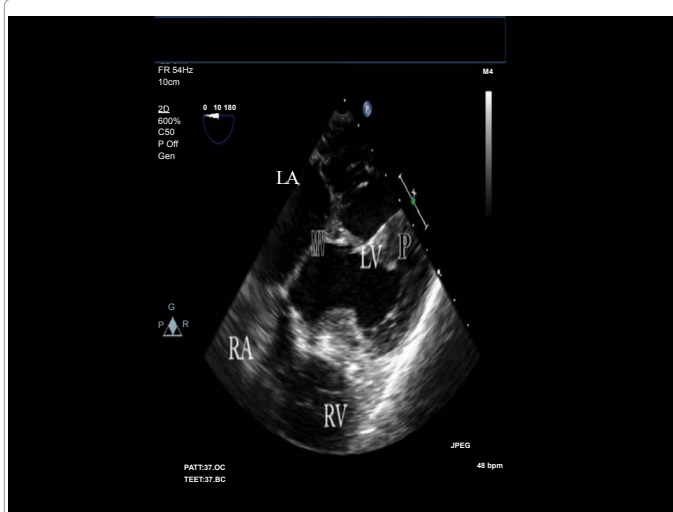


Figure 3: Modified deep transgastric long-axis view demonstrating the relationship between the single papillary muscle (P) and mitral valve. The TEE probe was positioned between the deep transgastric and 4-chamber views.

outcome is generally poor because of the multiple surgeries required to attain hemodynamic stability.

Adult PMV is an exceedingly uncommon condition. Only nine cases were identified in a nearly 50-year period (1960-2008) [3]. Of those nine, seven were male and five had isolated PMV. In contrast to PMV in the pediatric population, concomitant cardiac abnormalities are uncommon in adults.

Ours may be the first report of a transitional-type AVSD with an adult PMV and a cleft mitral valve. This is perhaps also one of the oldest patients with primum ASD to remain asymptomatic for so long. We speculate that the anomalous right AVV regurgitation through the primum ASD balanced the left-to-right shunt. Further, the cleft of the mitral valve provided additional space for the mitral orifice, thus preventing mitral stenosis in this adult PMV. Had we not discovered the single papillary muscle, the mitral valve cleft would have been closed as planned, eventually causing stenosis, because the only remaining blood flow from the left atrium to the left ventricle would have been through the interchordal spaces.

This case demonstrates the importance of 2D and 3D TEE imaging in helping to select the proper surgical intervention of adult patients requiring complex congenital cardiac repairs.

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