

## Intramural Ventricular Septal Defect

Sedigheh Saedi<sup>1</sup>, Mozhgan Parsaee<sup>2,\*</sup>

<sup>1</sup> Assistant Professor, Department of Cardiology, Rajaei Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran

<sup>2</sup> Associate Professor, Department of Cardiology, Echocardiography Research Center, Rajaei Cardiovascular, Medical and Research Center, Tehran, Iran

\* Corresponding author: Mozhgan Parsaee, Rajaie Cardiovascular Medical and Research Center, Niayesh Highway, adjacent to Mellat Park, Tehran, Iran. Tel: 009821-23921. E-mail: Parsaemozhgan@yahoo.com

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### Abstract

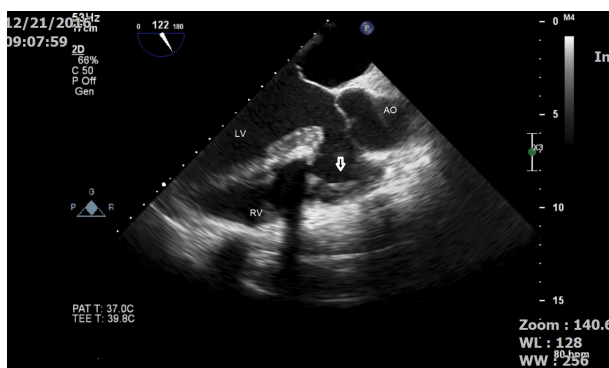
Intramural ventricular septal defects (VSDs) are less frequently encountered but clinically significant type of residual interventricular communications, seen after complex congenital heart surgeries. Hemodynamically significant intramural VSDs can lead to higher postoperative morbidity and mortality. This case highlights the clinical challenges including the need for multiple interventions faced in affected patients.

## INTRODUCTION

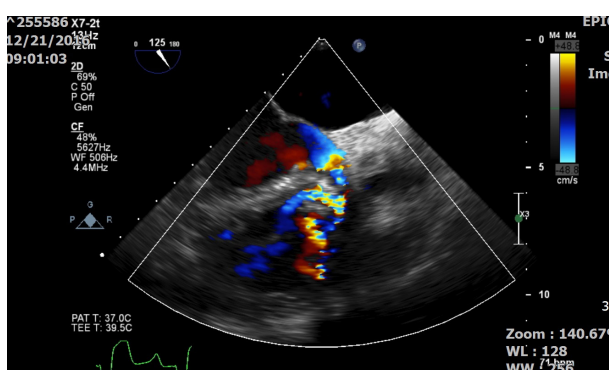
The number of patients with congenital heart disease surviving into adulthood has markedly increased over the past years, mainly due to substantial improvements in surgical techniques for correction or palliation of complex congenital heart diseases. As the result, medical professionals caring for this population constantly face new challenges regarding management of residual defects commonly seen early or late after the surgery. Intramural ventricular septal defects (VSDs) are less frequently encountered but clinically significant type of interventricular communication seen after complex congenital heart surgeries including corrective procedures for tetralogy of Fallot, transposition of great arteries and double outlet right ventricle. In fact, these defects are an unusual type of residual VSDs and result from attaching the aortic side of the VSD closure patch to the right ventricular (RV) trabeculae instead of the RV free wall. Technically, the patch must be inserted as close to the aortic valve as possible. If the patch is inserted somewhere within the hyper trabeculated RV wall far from the aortic valve, leakage of blood through the inter-trabecular recesses and the insertion site leads to left to right shunting. During repair of conotruncal abnormalities, this could happen more commonly due to characteristic multiple hypertrophied muscle bundles and coarse trabeculations in the right ventricle [1-3]. Here we present a typical case with a history of frequent surgeries and intramural VSDs. We also review the deficient existing literature in this regard.

## CASE REPORT

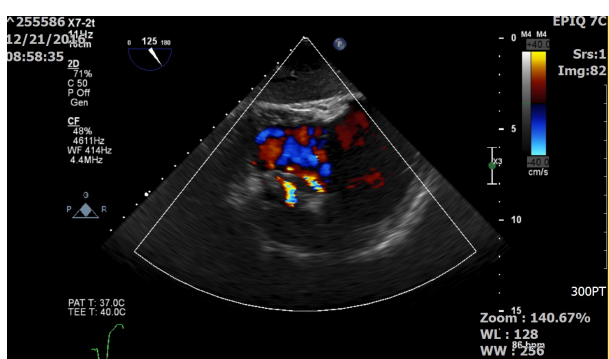
A 30-year-old male presented to our clinic with a history of repaired tetralogy of Fallot and later pulmonary valve replacement. He had a history of four previous surgeries. He had undergone Blalock-Taussig (BT) palliative shunt surgery as an infant. At age 8 he had surgery for total correction consisting of VSD closure; pulmonary commissurotomy and shunt take down. Early post-op the VSD repair patch was realized to be partially detached, for which the patient had another reparative operation. Follow-up transthoracic echocardiography showed that the residual VSD still persisted. At age 21 he had to undergo another surgery for pulmonary valve replacement (PVR) with a mechanical valve due to severe pulmonary regurgitation. At the time of PVR, repair of the residual VSD was also recommended. Now at age 27, our transthoracic echocardiography revealed the cause of failed past surgical efforts to repair the VSD. There was a large intramural VSD with multiple jets and outlet orifices to right ventricle. The VSD was not suitable for transcatheter closure (Figs 1-3). Cardiac catheterization was performed in which the estimated left to right shunting was calculated as  $Q_p/Q_s$ : 1.4:1 and a RV systolic pressure of 65mmHg. (Fig 4). Regarding a peak pressure gradient of 30mmHg across the prosthetic valve in echocardiography, the systolic pulmonary arterial pressure was estimated to be mildly elevated. As the patient had undergone multiple cardiac surgeries and the left to right shunting was not significant, redo surgery was postponed and the patient was decided to be followed closely in clinic.



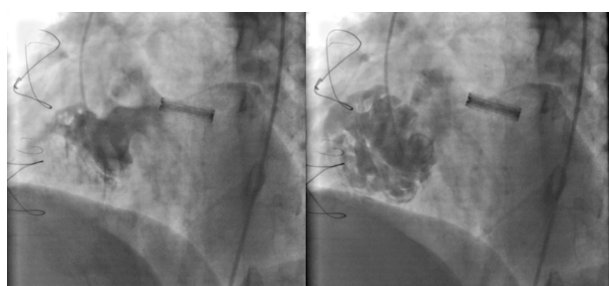
**Figure 1:** Multiple Intramural Ventricular Septal Defects in Transe-sophageal Echocardiography after Total Correction of Tetralogy of Fallot due to Attachment of Repair Patch to Right Ventricular Trabeculae



**Figure 2:** Color Doppler Images of the Same Patient Showing Multiple Exit Sites of the Ventricular Septal Defect



**Figure 3:** Transgastric View of the Intramural Ventricular Septal Defect during Trans-Esophageal Echocardiography



**Figure 4:** Left Ventricular Angiography in Left Anterior Oblique (LAO) Projection Showing Residual Ventricular Septal Defect through the Repair Path that is Attached to RV Trabeculations with Multiple Exit Sites.

**DISCUSSION**

Our case is a typical case of intramural VSD as faced in clinical practice. The reason for frequent unsuccessful attempts to close the VSD is the abnormal course and underlying pathology of intramural VSDs. The clinical impact of intramural VSDs is greater than simple residual VSDs commonly seen after repair of conotruncal anomalies and post-operative diagnosis is of higher importance. Children with hemodynamically significant intramural VSDs are reported to have a substantially higher rate of postoperative morbidity, mortality, and hospital stays. Intramural VSDs are also reported to be larger than other residual VSDs that might lead to increased clinical sequel [1, 2]. The prevalence, risk factors and outcomes of these unusual residual defects are not well defined in the literature. Intramural VSDs were first described by Preminger et al in eight patients about 22 years ago. They reported residual channels around patch attachment to RV. Multiple surgical and interventional procedures were unsuccessful mostly due to failure to localize the defect and multiplicity of defects [3]. In the study by Patel et al, an 11% of studied population were found to have intramural VSDs after repair of conotruncal abnormalities accounting for 19% of residual VSDs. Most of the affected patients had undergone total correction for tetralogy of Fallot as it is the most common type of conotruncal abnormality but a higher incidence in the more complex conotruncal pathologies should be expected [1]. Diagnosis could be far more challenging than other types of residual VSDs. The initial diagnosis is often made by transthoracic and sometimes transesophageal echocardiography. Intramural VSDs often have a complex, serpiginous tunnel shape course through intertrabecular spaces of the RV wall. The left ventricular end is commonly found under the aortic cusps, but the outlet on the RV side could be through multiple orifices [3]. These defects are not visualized at the expected location of the repair patch edges and the complex exit site might be found in the RV chamber with no direct relation to the repair patch. Therefore unconventional, non-standard echocardiographic views are needed for detection. RV inflow and high parasternal views in transthoracic echocardiography could be recommended for visualization. Deep transgastric views with smooth short axis rotations may better envision the defects during transesophageal echocardiography [4, 5]. Even in intramural VSDs that seem hemodynamically insignificant early after operation, close follow up with transthoracic echocardiography is recommended, as there is a possibility for gradual enlargement and increase in the ventricular level blood shunting, with a common cause being consistent regression of RV hypertrophy following corrective surgery for RV outflow tract stenosis [1-3]. There are no large-scale studies on preferred method of repair of hemodynamically significant intramural VSDs. In the study by Belli et al consisting of 4 patients, trans-aortic approach was reported to have more satisfactory results than closure through right atrium or the right ventricle. If transatrial method is used, the patch should be partially resected and edges reassessed, as attempts to close multiple intertrabecular defects often fail. Transcatheter closure might rarely be an alternative in carefully selected cases [2, 5]. Special attention should be paid to detection of intramural VSDs as a distinct and highly missed sub-group of residual VSDs in post-operative follow-up of complex congenital heart disease patients. We also aimed to bring light to the point that before proceeding with re-intervention for residual congenital defects, one should have their complex nature in mind and

use optimum imaging modalities to determine the best management option on individual basis and avert adverse clinical sequels.

### CONFLICTS OF INTERESTS

There is no conflict of interest.

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