Diagnosis and management of residual atrial septal defect after surgical failure: A case series

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ABSTRACT

Residual defect after surgical closure of atrial septal defect is extremely uncommon. This communication reports four cases encountered in a tertiary care center during the last three decades. Clinical diagnosis was challenging, and the diverse presentations included acute ischemic stroke, cyanosis, and right ventricular volume overload. The morphology of the residual defects was complex, and multimodality imaging (transesophageal echocardiography, peripheral venous contrast studies, computed tomography, and balloon occlusion) enabled accurate recognition. Percutaneous device closure was feasible in one but required repeat surgery owing to unfavorable anatomy in the others. The communication focuses on difficulties in diagnosis and management.

Keywords: Device closure, failure of atrial septal defect surgery, multimodality imaging for diagnosis, residual atrial septal defect following surgery

INTRODUCTION

Atrial septal defect (ASD) is common and accounts for nearly 10% of congenital heart disease. Management of clinically significant ASD is achieved by closure with a surgical suture, patch, or percutaneous device. Surgical closure of ASD with the use of cardiopulmonary bypass has been the standard treatment for decades with excellent short- and long-term results.[1,2]

Failure of the surgical repair with resultant symptomatic presentation is rare. This communication describes four patients encountered during the last decades and focuses on the wide spectrum of clinical presentation, the complex anatomy of residual defects, and the role of multimodality imaging in management.

SUBJECTS AND METHODS

A retrospective data analysis of four patients of postsurgical residual ASD treated from October 1992 to September 2023 was performed. The diagnosis of surgical failure was made on clinical assessment. Relevant blood investigations, electrocardiogram (ECG), skiagram chest, tranthoracic Echocardiography (TTE), transesophageal echocardiography (TEE), peripheral venous contrast studies with TEE, three-dimensional (3D) cardiac computed tomography (CT), balloon sizing, and management (percutaneous or surgical) data were analyzed.

RESULTS

Tables 1 and 2 provide relevant clinical data. Table 1 summarizes the clinical data of the four patients.

Case 1

A 19-year-old female presented with a history of recurrent upper respiratory infection in childhood and

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effort dyspnea. She underwent direct suture closure of secundum ASD using a right lateral incision. A year later, she presented with features of right ventricular volume overload (RVVO) and needed a reoperation through a midline incision and using a pericardial patch. A single-chamber pacemaker was implanted in the postoperative period for prolonged sinus pauses, and a pulse generator replacement was needed after two decades. There has been persistent atrial fibrillation during the last 5 years, treated with appropriate pharmacotherapy and pacing support. Follow-up in May 2024 revealed no residual shunt across the patch.

Case 2

A 32-year-old female was diagnosed with a "hole in the heart" in early childhood but presented with effort dyspnea and palpitations. She underwent surgical closure for ASD using midline incision and pericardial patch closure. Twelve years after surgery, she presented with effort-induced New York Heart Association Class II dyspnea for the last 2 years. Clinical examination, ECG, and chest X-ray findings were consistent with a large left-to-right shunt at the atrial level. TTE and TEE revealed RVVO, a large left-to-shunt, and a 30-mm defect in the atrial septum. The anterior, posterior, superior, and inferior margins each were 4–5 mm. The defect was closed percutaneously using a 34-mm Block-Aid device (Shengai Shape Memory Alloy Co-operation limited, China) with the complete abolition of the shunt. The patient is asymptomatic and doing well at 20-year follow-up.

Case 3

A 4-year-old boy had undergone cardiac surgery using a midline incision and pericardial patch closure for secundum ASD along with pulmonary valvotomy. After uneventful 19 years of cardiac surgery, he was admitted with acute ischemic stroke (left-sided hemiplegia). Magnetic resonance imaging (MRI) of the brain revealed focal acute nonhemorrhagic infarction in the right corona radiata and normal intracranial vessels. Clinical examination, ECG, chest X-ray, relevant blood investigations, Holter monitoring, and lower limb venous Doppler were normal.

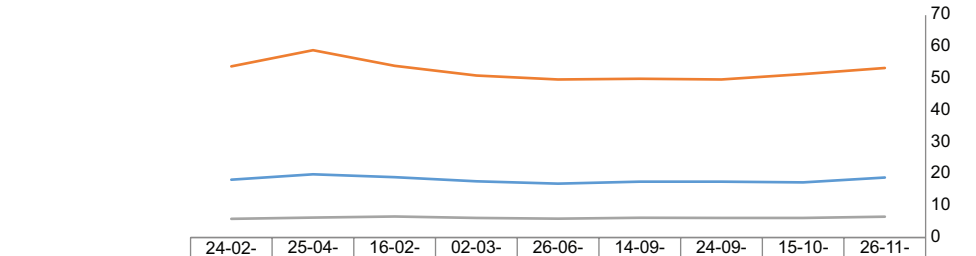
TTE revealed mildly dilated right heart chambers and prominent coronary sinus, raising the possibility of the left superior vena cava. TEE demonstrated a gap in the aortic and inferior vena cava (IVC) (inferior) margin of the patch with shunting from the right atrium (RA) to the left atrium (LA) [Figure 1a]. Cardiac CT angiography failed to demonstrate any residual defect or patch. Transcatheter device closure was planned under fluoroscopic, TEE, and angiographic guidance. Balloon interrogation revealed two defects of 10–12 mm in size [Figure 1b]. There was the persistence of bubble contrast from RA to LA on TEE despite the balloon occlusion of the defects. Device deployment was deferred as these observations suggested a complex anatomy. At surgery, there was degeneration and calcification in the lower portion of the patch, and an additional defect was visualized in the inferior margin of IVC [Figure 1c]. Both defects were closed using a

Table 1: Clinical data

Sex	Year	Age (years)		Surgery details	Presentation	Management
		At surgery	Symptoms recur			
Female	1994	19	20	Direct suture right lateral	RVVO	Resurgery (midline and pericardial patch)
Female	2001	32	44	Midline, pericardial patch	RVVO	Device closure (32-mm Block-Aid)
Male	2020	4	23	Midline, pericardial patch	AIS	Resurgery (midline and pericardial patch)
Male	2020	14	27	Right lateral, pericardial patch	Polycythemia	Resurgery (midline, pericardial patch, and rerouting of venous flow

RVVO: Right ventricular volume overload, AIS: Acute ischemic stroke, and Year: When the patient presented

Table 2: Serial hematocrit (%), hemoglobin (%), and red blood cell values in Case 4



	24-02-2015	25-04-2017	16-02-2020	02-03-2020	26-06-2020	14-09-2020	24-09-2020	15-10-2020	26-11-2020
Hemoglobin (g/dL)	18.2	19.9	19	17.7	17	17.6	17.6	17.4	18.9
Hematocrit (%)	53.9	59	54.1	51	49.8	50	49.8	51.5	53.4
RBC (10*6/uL)	5.89	6.3	6.61	6.16	5.93	6.25	6.21	6.18	6.56

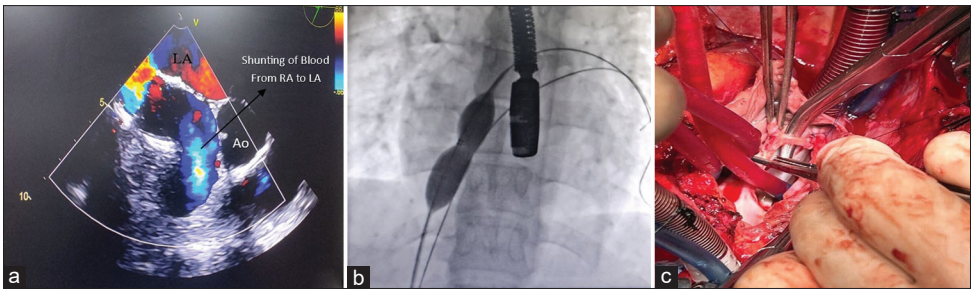


Figure 1: (a) Midesophageal 60° in transesophageal echocardiography showing deficient aortic margin (arrow) with a left-to-right shunt. (b) Balloon interrogation reveals two defects. (c) Surgery revealed two inferiorly located residual defects with degeneration of the patch. Ao: Aorta, LA: Left atrium, RA: Right atrium

fresh pericardial patch. The patient is asymptomatic at 6-month follow-up.

Case 4

A 14-year-old male had undergone cardiac surgery for a "hole in the heart" using right lateral thoracotomy with a pericardial patch. The preoperative echocardiography revealed a large (29 mm) ASD with a deficient inferior margin, RVVO, and normal drainage of pulmonary veins. In the postoperative period, he underwent multiple TTE, which was reported to be normal. After 13 years, he was referred for cardiac evaluation. During the previous 5 years, he underwent extensive investigations by a physician and hematologist for fatigue and polycythemia. Clinical examination was unremarkable, except for mild clubbing and uniform central cyanosis (88%–90% on pulse oximeter). Table 2 displays hematocrit, hemoglobin %, and red blood cells during the last 5 years.

TTE and TEE studies, along with an upper arm contrast study, revealed no shunt, pulmonary arteriovenous fistula, and normal pulmonary venous drainage. TEE with contrast studies from the lower limb revealed passage of contrast from IVC to LA and RA. CT angiography suggested biatrial communication of IVC with IVC draining into both RA and LA. The contrast flow from IVC was preferentially to LA and marginally to RA [Figure 2]. No shunt was observed across the pericardial patch. At reoperation, it was observed that the previous patch placement was faulty and prevented part of IVC blood from entering RA; instead, it was shunted to LA. A graft was sutured into the IVC to enlarge its diameter, close the residual defect, redirect flow to RA, and prevent flow to LA. The patient is asymptomatic at 6 months, following up with normal saturation.

DISCUSSION

Surgical repair of ASD is a well-established and highly durable treatment.[1,2] The defect is occluded through primary sutures or patches in the open-heart operation. The traditional approach for surgical ASD repair has been a median sternotomy. The right anterolateral submammary, the subpectoral approach, has been preferred in females to provide a cosmetic incision.[3] The failure of surgery in cases one and four could be attributed to right thoracotomy, which is known to restrict the operative field and precisely define the suture line. Suture dehiscence following direct closure might have been an additional contribution in Case 1.

Three patients had ASD repair using a pericardial patch, resulting in secure, low profile, and effective closure with an interatrial septum aligned with native tissue. The surgical belief is that patch closure precludes any residual shunt, trivial or otherwise. However, residual shunting after surgical closure varies from 2% to 7.9% in the long-term follow-up.[1] The residual defect may be associated with deficient rims, inadequacies in the surgical technique, or degeneration (retraction, shrinkage, and calcification) of the patch. Glutaraldehyde preservation at the time of surgery can aggravate the degeneration. The patch failure in this study became clinically apparent 12 months–17 years after surgery. However, residual defects can manifest within days, months, or later.[4-7]

Diagnosing residual defects can be challenging as clinical manifestations can be diverse, and RVVO can be absent [Table 1]. Comprehensive evaluation enabled the conclusion that paradoxical embolization from degenerated and calcified pericardial patches caused a stroke in Case 3. Recurrent neurological events have been described following open surgical ASD patch repair in a patient with persistent tunneled defects.[8]

Case 4 had an extremely unusual presentation with cyanosis, which was misdiagnosed as primary polycythemias [Table 2]. These patients are also at a high risk of paradoxical embolization. Iatrogenic right-to-left shunting at the atrial level was more frequent before the routine use of cardiopulmonary bypass and is rare in the current surgical practice. This complication results from mistaking the Eustachian valve for the lower margin of the defect, incomplete closure, or inadvertent diversion of IVC flow. The latter phenomenon has been reported following surgery for low-lying large ASD with

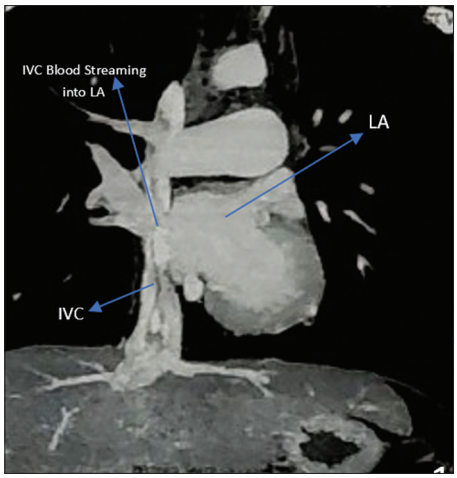


Figure 2: Arrow showing the preferential flow of inferior vena cava blood into the left atrium. IVC: Inferior vena cava, LA: Left atrium

no inferior margin, sinus venosus defect of inferior vena caval type, or inferior ASD complicated by partial anomalous pulmonary venous drainage.[9,10] Cyanosis usually appears early in the postoperative period but can be delayed for four decades. Cyanosis is attributed to the partial distribution of the suture line many years after the operation. Diagnosing this complication was challenging as conventional TTE, TEE, and upper arm peripheral vein contrast studies were normal. A high index of suspicion and multimodality imaging established the right-to-left shunting. TEE-combined contrast echocardiogram performed from the venous access in the lower limb and 3D echocardiography and CT angiography substantiated the biatrial commitment of the IVC with the predominant flow of contrast from IVC to LA. Cardiac MRI and cardiac catheterization with IVC angiogram have also been used for diagnosis.

It is widely agreed that residual defects should be treated on the principles of native ASD and should be closed if there is a significant shunt causing RVVO. Case 1 had repeat surgery using a pericardial patch. Case 2 had percutaneous closure of the defect with sustained benefits in the long term. Device closure of residual defects using different devices has emerged as an alternate option to surgery.[4-8] In our experience, great caution is needed while evaluating residual defects for transcatheter closure. These defects can be at unusual locations, malaligned, with deficient posterior–inferior rim, and difficult to visualize on routine imaging. In Case 3, the precise delineation of the residual defect was impossible despite TEE, CT angiography, contrast studies, and balloon interrogation. Live 3D echocardiography can help guide transcatheter closure.[11] Surgical findings revealed two defects and a degenerated, calcified patch, which was the likely source of the stroke and the residual shunt. In retrospect, the decision to defer the transcatheter procedure was justified. Large residual defects with deficient inferior–posterior rim may not be suitable for device closure.[4]

Similarly, Case 4 had a complex anatomy that could be defined only after lower limb venous access contrast echocardiography and CT angiography. The inferior margin of the defect was reported as deficient in surgical records. The percutaneous way of closing this defect was to place a covered stent from the IVC into LA and then into RA through the patch. This was discussed with the family, but the combined decision was against the same due to the young age, uncertainty regarding placing a stent on the systemic side of the heart with a need for anticoagulation, and excellent safety of repeat surgery. Resurgical repair successfully allowed the flow of blood into RA. Transcatheter treatment can be considered in patients who cannot tolerate open-heart surgery.[12]

It is rare to see the surgical failure of ASD (patch closure/direct suture) with varied clinical presentations, including stroke and cyanosis. The anatomy of residual defects can be complex and should be adequately defined with all available modalities before planning the closure approach.

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Conflicts of interest

There are no conflicts of interest.

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