

Uncommon Facades of Atrial Septal Defect - A Case Series

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ABSTRACT

Atrial septal defect (ASD) is one of the most common congenital cardiac lesions in adults. While the most common symptoms include fatigue, exertional dyspnea, and palpitations, the presentation of ASD can be protean. In this article, we describe three uncommon presentations of ostium secundum ASD. Patient 1 presented with pericardial tamponade. Patient 2 presented with ventricular tachycardia. Patient 3 had moderately severe mitral regurgitation. These patients had chest radiographs and electrocardiograms typical of secundum ASD, but their presentations were atypical.

Keywords: ostium secundum, mitral regurgitation, pericardial tamponade, ventricular tachycardia

Learning objective

This article highlights the protean manifestations of a rather common congenital heart disease – atrial septal defect. Improved understanding of these uncommon presentations is essential in order to establish earlier diagnosis and concordantly plan treatment. The above cases presented a diagnostic challenge to us due to limited awareness, the lesion hiding behind more dramatic symptoms and not being evident at initial presentation. Early recognition of such manifestations helps in arriving at a timely diagnosis, thereby formulating an appropriate management plan for the patient.

INTRODUCTION

Atrial septal defect (ASD) is one of the most common congenital cardiac lesions in adults and is often asymptomatic until adulthood. Atrial septal defects (ASDs) are the most common form of congenital heart disease, and account for approximately 6–10% of congenital heart defects, with approximate population incidence of 1–3 per 1,000.⁽¹⁾ There are four types of ASDs, including ostium secundum defect, ostium primum defect, sinus venosus defect (inferior and superior), and coronary sinus defect. Although usually asymptomatic in childhood, ASD will be symptomatic in approximately 75% of adults.⁽²⁾ The most common symptoms include fatigue, exertional dyspnea, and palpitations. However, the presentation of ASD can be protean. While a detailed discussion is beyond the scope of this article, we will describe three variegated of patients with ostium secundum ASD. Patient 1 presented with pericardial tamponade. Patient 2 presented with ventricular tachycardia. Patient 3 had moderately severe mitral regurgitation. These patients had chest radiographs and electrocardiograms typical of secundum ASD, but their presentations were atypical.

CASE 1

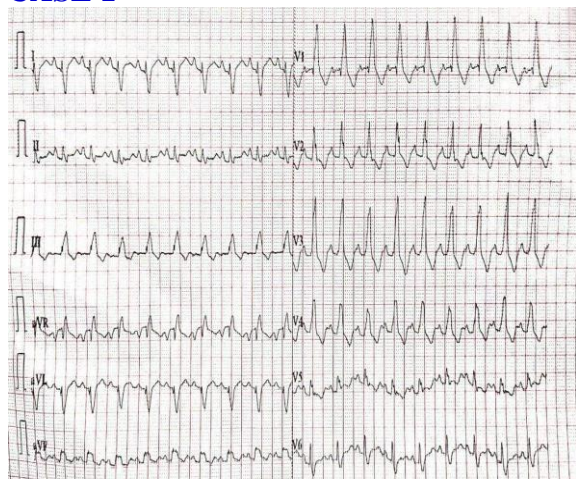


Figure 1a. ECG showed sinus tachycardia with right axis deviation. Lead V1 and aVR demonstrate a rsR prime pattern (pseudo RBBB) with slurring of the terminal R prime.

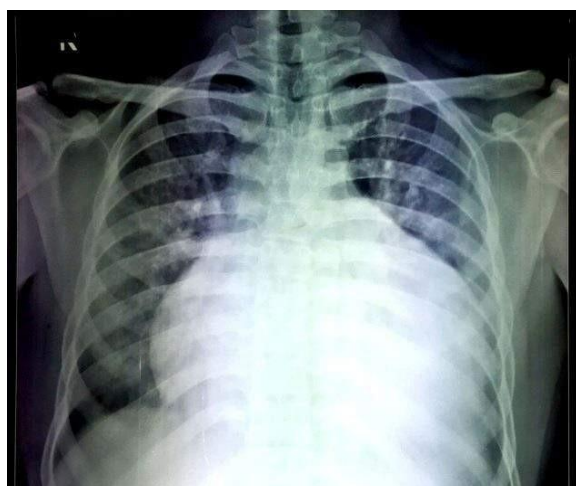


Figure 1b. Chest x-ray shows a grossly enlarged cardiac silhouette with a symmetrical and globular "water bottle" configuration suggestive of pericardial collection.



Figure 1c. 2D ECHO revealed a large circumferential pericardial effusion measuring 46 mm with diastolic collapse of right ventricle, suggestive of a cardiac tamponade.

A 25 year old male presented to the medicine OPD with dyspnea

associated with orthopnea and palpitations for the past 3 weeks. He also gave history of fever and productive cough for the past 2 days. He reported that his sister passed away 4 year prior due to some cardiac illness. On examination, his heart rate was 110/min and regular, blood pressure was 100/60 mm Hg, SpO₂ was 95% at room air, his heart sounds were muffled, respiratory examination revealed fine bibasal inspiratory crepitations. ECG showed sinus tachycardia with right axis deviation. The mean QRS duration was increased with an R/S ratio >1 in lead V1 signifying right ventricular hypertrophy. P waves have a peaking configuration in lead II suggestive of RA overload. Lead V1 and aVR demonstrate a rsR prime pattern (pseudo RBBB) with slurring of the terminal R prime (Figure 1a). Chest xray revealed a grossly enlarged cardiac silhouette with an almost symmetrical and globular configuration which suggests gross pericardial collection. Widening of the subcarinal angle is also noted (Figure 1b). An emergency bedside ECHO revealed a large circumferential pericardial effusion measuring 46 mm with diastolic collapse of right ventricle, suggestive of a cardiac tamponade (Figure 1c). The patient was shifted to the cathlab and after getting informed consent, pericardiocentesis was done via subxiphoid approach under echocardiographic guidance. Hemorrhagic pericardial fluid of approximately 450 mL was removed and the sample was sent for analysis. Patient symptoms improved dramatically following pericardiocentesis. Patient was then transferred to the IMCU. Post-pericardiocentesis, a detailed ECHO was done and it revealed a dilated right atrium and ventricle, with a large ostium secundum atrial septal defect of 42 mm with a left to right shunt (Figure 1d). A mild degree of pulmonary hypertension was also detected. The pericardial fluid analysis ruled out tuberculosis as evidenced by a negative CBNAAT and cytological studies ruled out malignancy as possible cause for the tamponade. The patient also denied any

history of trauma to the chest. CT pulmonary angiography showed no evidence of pulmonary thromboembolism. The patient was started on broad spectrum antibiotics and kept under observation. Patient condition improved significantly, with improvement of his symptoms, heart rate and blood pressure. On day 14 following admission, the patient was referred to the cardiothoracic surgery

department in view of need for closure of ASD and then discharged. On follow-up after 3 months, the patient had been admitted for a minimal invasive ASD closure via right thoracotomy. His ostium secundum ASD was repaired using a pericardial patch and he had tolerated the procedure well. Patient's post-op recovery was good and is currently on follow-up.

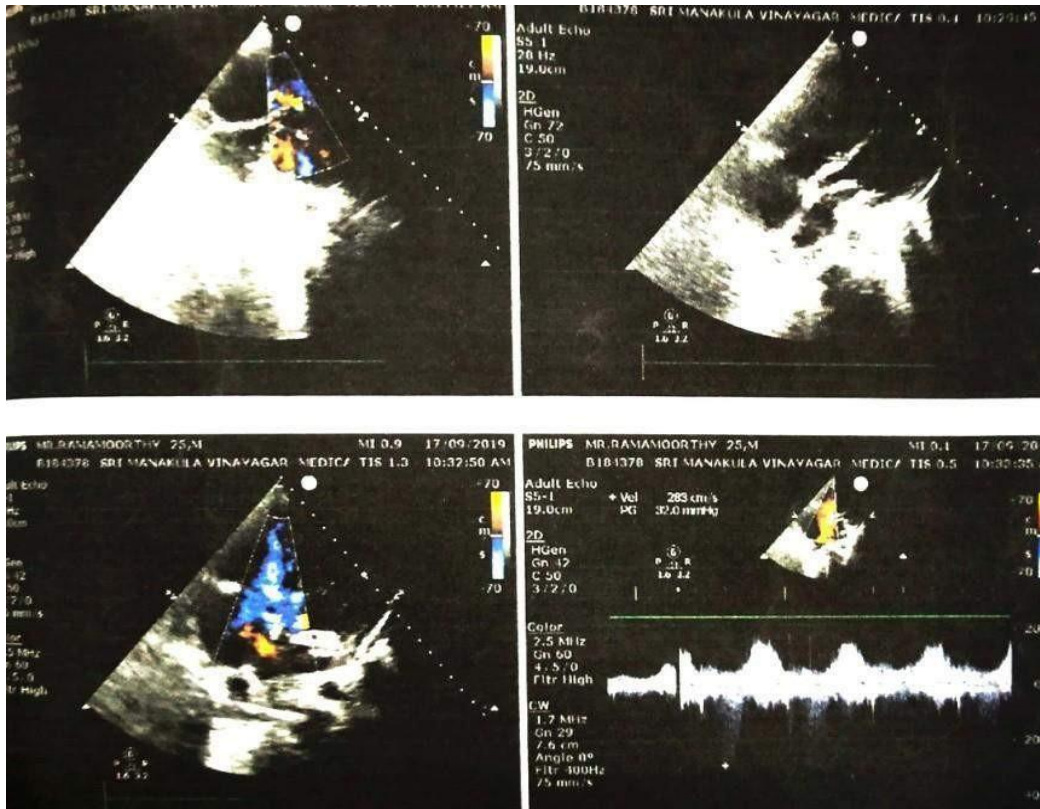


Figure 1d. Post-pericardiocentesis – 2D ECHO revealed a dilated right atrium and ventricle, with a large ostium secundum atrial septal defect of 42 mm with a left to right shunt.

CASE 2:

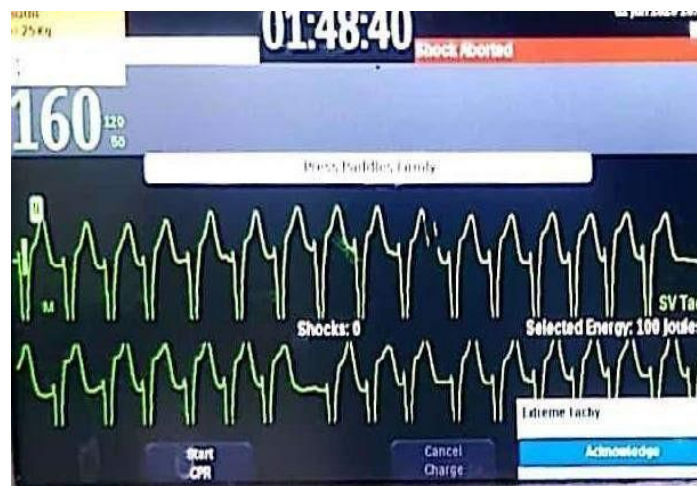


Figure 2a. Cardiac monitors showed broad complex tachycardia suggestive of ventricular tachycardia

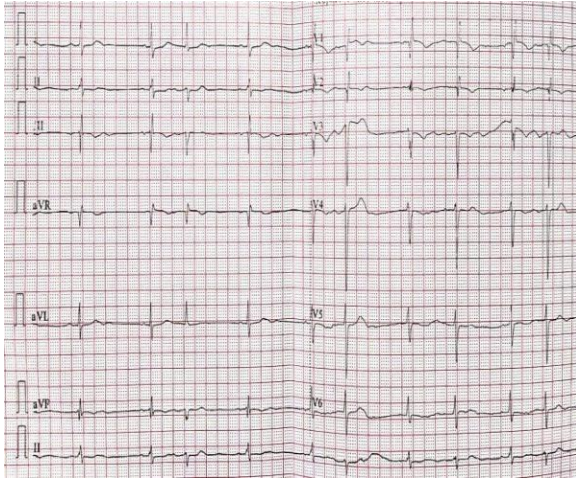


Figure 2b. Q waves present in right precordial leads and absent in left precordial leads signify that shunt traverses through right ventricle than the left ventricle.



Figure 2c. Chest x-ray shows cardiomegaly with prominence of pulmonary trunk. The ascending aorta is inconspicuous indicating that the intra-cardiac shunt does not traverse the aortic root.

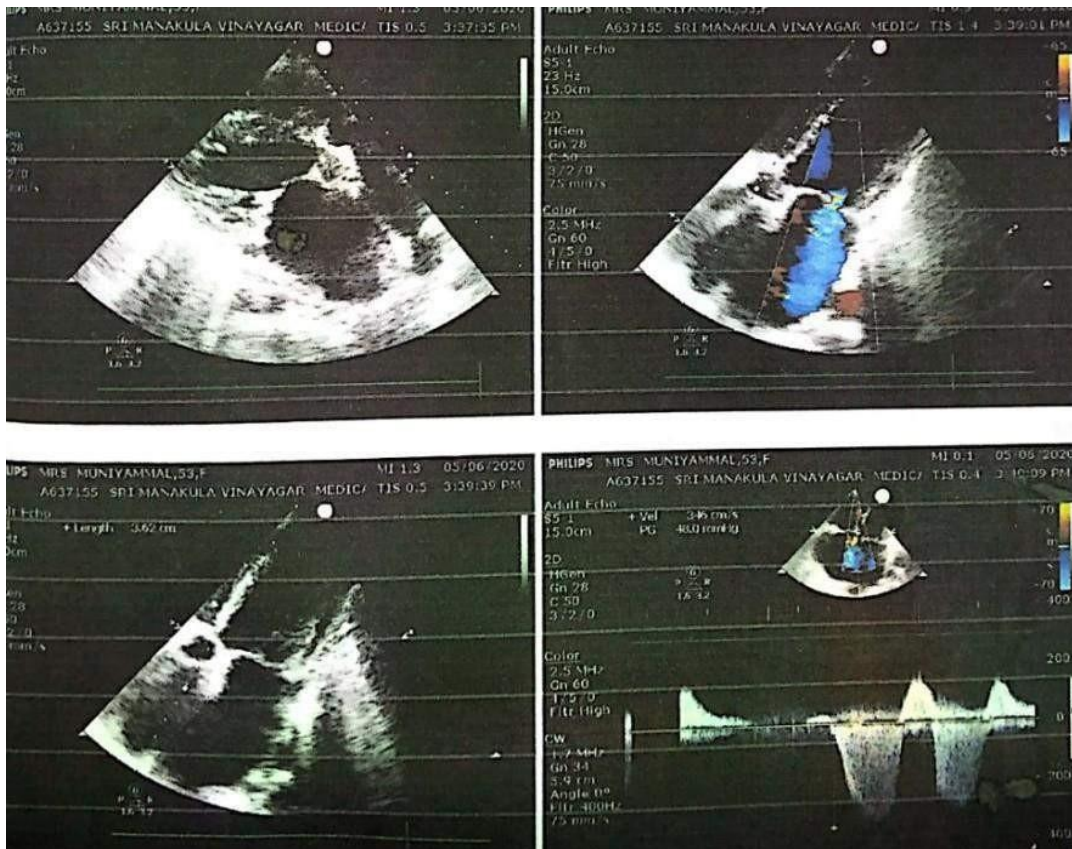


Figure 2d. 2D echo showed a dilated right atrium, right ventricle and an atrial septal defect of ostium secundum type measuring 35 mm.

A 53 year old female presented to the casualty with complaints of acute onset breathing difficulty, palpitations and giddiness for the past 1 hour, not associated with chest pain and syncope. She was diagnosed to have atrial septal defect 3 years back and managed conservatively. Her pulse was rapid and thready, BP was 80/60

mm Hg and saturation at room air was around 86%. Respiratory examination revealed bibasal crepitations. She was connected to the cardiac monitors which showed broad complex tachycardia with occasional capture beats suggestive of ventricular tachycardia (Figure 2a). She was started on nasal O₂ therapy and after

obtaining consent, delivered with 100 J of DC shock following which the VT was reverted back to sinus rhythm. She was then shifted to the IMCU where she was started on dobutamine and amiodarone, and planned for stabilization. Post-DC shock ECG showed HR of 50/ min, irregular rhythm with the presence of P-wave (Figure 2b). Q waves present in right precordial leads and absent in left precordial leads signify that shunt traverses through right ventricle than the left ventricle. Inferior leads demonstrate rSR prime pattern with lead II demonstrating a notch near the apex of R wave (cochretage), an electrocardiographic sign highly probable of a diagnosis of ASD. Chest X-ray PA view demonstrated cardiomegaly with gross enlargement of the right atrium, right

ventricle and pulmonary trunk as evidenced by the laterally shifted right heart border, left heart border and RV apex (Figure 2c). The pulmonary vascularity is increased with prominence of pulmonary trunk. The ascending aorta is inconspicuous indicating that the intra- cardiac shunt does not traverse the aortic root. 2D echo showed a dilated right atrium, right ventricle and an atrial septal defect of ostium secundum type measuring 35 mm with a left to right shunt, associated with moderately severe pulmonary hypertension (Figure 2d). Patient was stabilized in the IMCU, and later shifted to the ward. She was then referred to a higher centre for electrophysiological studies and radiofrequency ablation.

CASE 3

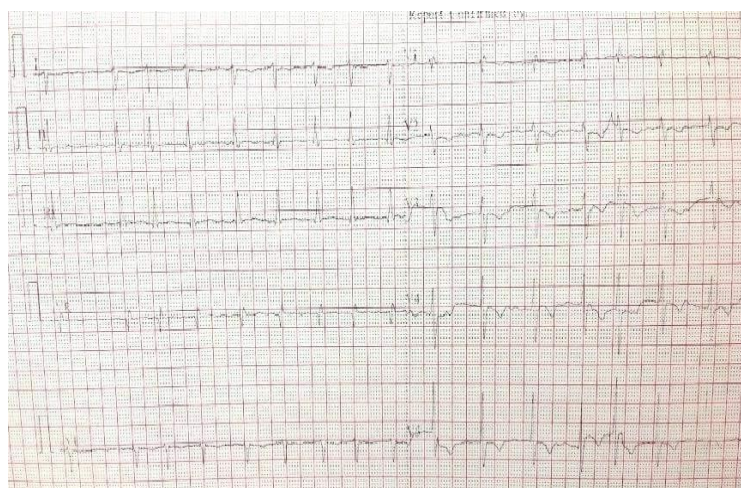


Figure 3. ECG showed a heart rate of 108, with varying RR intervals and absent p waves suggestive of atrial fibrillation.

A 40 year old female presented with chief complaints of breathing difficulty for the past 6 months, which was aggravated over the past 1 week and was associated with palpitations. No history of hypertension/ previously detected cardiac illness. No history of previous hospitalizations or similar complaints in the family members. Her heart rate was 98/min, irregularly irregular, variable volume with all peripheral pulses felt. Bp was 100/60 mm Hg in the right upper arm in supine position. A pansystolic murmur of grade 3 was heard in the left third intercostal space.

The patient also had a wide fixed split, appreciated best in the left second intercostal space. A grade 2 non-radiating pansystolic murmur was heard in the left fifth intercostals space. ECG showed a heart rate of 108/min with absent p waves and varying RR intervals suggestive of atrial fibrillation (Figure 3). ECG also demonstrated an incomplete RBBB pattern with an axis shifted to the right. 2D Echo revealed Dilated Left atrium/ Right atrium/ Right ventricle/ Moderate mitral regurgitation/ Mild RV dysfunction with no pulmonary hypertension. It also showed an

Ostium secundum ASD - 12mm (L→R shunt).The patients started on beta blockers and spironolactone. She was also initiated on anticoagulant therapy to prevent embolic stroke secondary to atrial fibrillation. The patient general condition improved significantly and tolerated therapy well, and is currently on follow-up.

DISCUSSION

Atrial septal defect (ASD) is a common congenital acyanotic heart disease encountered in clinical practice. It is the first congenital heart disease recognized by Leonardo DaVinci in 1531.

The first clinical diagnosis of ASD was by Belford and his colleagues in 1941. There are four types of ASD are ostium secundum defect (74%), ostium primum defect (20%), sinus venosus defect (5%) and coronary sinus defect (1%). In this article, we want to highlight some of the uncommon presentations of ostium secundum type of ASD. While literature is riddled with anecdotes of pericardial effusion and tamponade after surgical repair of atrial septal, the rare instance of a cardiac tamponade in the natural course of an atrial septal defect is seldom come across.³⁻⁵ The chronic left-to-right shunt associated with ASDs leads to increased hemodynamic load and geometric remodeling, both at a cellular and macroscopic level. This chronic volume stress leads, in turn, to electrical remodeling that may precipitate development of arrhythmias. Atrial myocyte electrophysiological properties are altered, with increased intra-atrial conduction time being a common finding, likely from combination of interstitial fibrosis and chamber enlargement. Arrhythmias and conduction disorders are well-described in patients with ASDs. Atrial tachyarrhythmias such as atrial flutter and atrial fibrillation are rare in childhood, but become increasingly common with older age at time of repair, larger shunt size and comorbidities. In unoperated adults, the estimated incidence of atrial arrhythmias is approximately 10 % under the age of 40

years, rising to at least 20 % with increased age.⁶ To our knowledge there is neither any reports of ASD presenting with ventricular tachyarrhythmia nor any conclusive evidence of an increased risk of ventricular arrhythmias in patients with ASDs, despite the evidence of geometric and electrical remodeling of the RV myocardium.⁷ Medical treatment of arrhythmias in patients with ASDs is similar to treatment strategies in the general population. Procedural treatments such as ablation should be undertaken in consideration of timing and type of ASD closure. Incidence of arrhythmias decreases after closure of ASD, but remains elevated in comparison to general population, particularly with late age of repair.

Finally, we discuss a rather peculiar association of atrial septal defect with mitral regurgitation. The incidence of mitral regurgitation is high in secundum ASD.⁸ While the reported 5% incidence of severe MR in the secundum ASD population provides some clinical perspective to this association, one study has emphasized the increased frequency of severe MR specifically in the older patient, which was 15% in patients over 50 years of age.⁹ The pathology of the mitral valve in older ASD patients with severe MR consists of thickening and shortening of the chordae. These valves are densely fibrotic with scattered microscopic foci of myxomatous degeneration. However, the clinical significance and overall perspective of the association between secundum atrial septal defect and mitral regurgitation has not been fully characterized. Awareness of the frequent coincidence of severe MR in older ASD patients has clinical importance. In patients with ASD, the magnitude of MR is often underestimated because the ASD allows decompression of the regurgitant left ventricular volume; MR may even be in apparent preoperatively. However, following ASD closure, left atrial decompression across the ASD is no longer possible, thus patients with severe MR develop symptoms of pulmonary venous

hypertension. Awareness of the frequent occurrence of severe MR in older patients with ASD is thus important to the surgeon for intraoperative mitral valve evaluation and to the physician managing these patients following ASD repair.

Conflict of interest: The authors declare no conflict of interest.

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