**Atrial Septal Aneurysm**  
**Classification, Clinical Implications and Management**  
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**Case Reports: Case No 1.** 35 year old male presented with history of palpitations usually on effort. In past about a decade back he had a check up for some chest problem. He was told to have some cardiac problem but he did not go for further evaluation. Clinical examination revealed average build. No anemia, cyanosis or jaundice. JVP was not raised. Pulse 80/min regular all peripheral pulses normal, blood pressure of 120/84. No signs of cardiac failure. Systemic examination was normal. Cardiac examination revealed tachycardia, normal $S_1$ wide fixed-split $S_2$, Grade 3/6 systolic murmur in pulmonary area. Hemogram, lipid profile, KFT and LFT were normal. ECG revealed normal sinus rhythm with RBBB. X-Ray chest has mild cardiomegaly, prominent central pulmonary arteries. Echocardiography was done which revealed dilated RA and RV, normal valves. IVS was intact. IAS had a large ($12\times14\text{mm}$) bulge predominantly towards right side although with significant oscillations of the bulge. No clot. CFM revealed Left to Right Shunt across a dropout in the bulge. There was a peak gradient of 20 mmHg across the Pulmonary valve (Figures 1 and 2).

![Fig. 1 Apical 4C showing dropout in ASA](image1)

![Fig. 2 Apical 4C CFM showing shunt across ASA](image2)

This patient has Atrial Septal Defect (ASD) with atrial septal aneurysm (ASA). There is a large left to right shunt. Possibly has partially closed ASD with aneurysm formation or else an association of ASA with ASD which is well known. This patient has ASD which can well explain palpitation. But Atrial Septal Aneurysm could also be contributory.

**Case No. 2:** 53 yrs old lady presented with transient left side weakness which improved slowly over 3 days. There was past history of palpitations on and off for few years prior to this episode. Clinical examination revealed irregular pulse 102/min with pulse deficit of 6/min, BP 130/80mmHg. Peripheral pulses were felt normal. CVS examination revealed irregular heart rate, normal heart sounds, systolic click and grade iii/iv systolic murmur in pulmonary area. CNS examination revealed mild weakness in left lower limb but patient was able to walk without support. Hemogram, serum lipids, kidney function tests and liver function tests were within normal range. ECG revealed atrial fibrillation with ventricular rate of 90/min normal QRS and T waves. Chest X-Ray showed normal cardiac size and clear...
lungs. CT brain was normal with no evidence of hemorrhage. Transthoracic Echocardiogram revealed normal size chambers, normal valves, and prominent moderator band in right ventricle. IVS was intact. IAS showed a large bulge in fossa ovalis area towards right atrium with limited excursions of this part of atrial septum. No evidence of thrombus in any chamber or atrial appendage. No evidence of any shunt (Figures 3-5). Carotid Doppler and ultrasound studies were normal.

Patient has Ischemic Cerebrovascular embolic stroke with left side hemiparesis with Atrial fibrillation and Atrial Septal Aneurysm Fossa ovalis type 1A. This patient obviously has atrial fibrillation with ischemic cerebrovascular accident most likely cardioembolic stroke. Atrial Septal Aneurysm (ASA) though an accidental finding could be contributing to cardioembolic stroke though we could not see any thrombus in aneurysm or left atrium in transthoracic echo. Patient was started on anticoagulants and rate control for atrial fibrillation. Both of these patients have Atrial Septal Aneurysm. First one presented with palpitations and second one with cerebral embolism. First case has associated ASD and second case has atrial fibrillation and cerebral embolism.

**ATRIAL SEPTAL ANEURYSM (ASA)** Atrial septal aneurysm is a localized “saccular” deformity, generally at the level of the fossa ovalis, which protrudes to the right or the left
atrium or both sides. Atrial septal aneurysm though rare is well recognized cardiac abnormality. It was previously diagnosed on autopsy but with advent of echocardiography it is being picked up frequently on routine echocardiography or evaluation of ischemic stroke. Clinical significance is uncertain though many studies link it with peripheral embolism and cardioembolic stroke (1), pulmonary embolism, atrial arrhythmias.

Atrial septal aneurysm can be secondary to interatrial pressure difference or can be primary malformation involving fossa ovalis region or entire septum (1). Since atrial septal aneurysms are rare in patients with chronically elevated atrial pressures as in mitral stenosis so acquired origin seems unlikely and most likely there is congenital malformation of the atrial septum contributing to development of ASA as was suggested by Hanley PC and colleagues (2). 

Echocardiographic definition of ASA is somewhat arbitrary. Hanley PC and colleagues after evaluating 80 consecutive cases of ASA devised echocardiographic criteria and classification of ASA (2). They defined an aneurysm if atrial septum or part of it exhibited aneurysmal dilatation protruding at least 15 mm beyond the plane of the atrial septum or if the atrial septum exhibited phasic excursion during the cardiorespiratory cycle exceeding 15 mm and if the base of aneurysmal protrusion was at least 15 mm in diameter. They also classified aneurysms into 

- Those involving entire septum
- Those involving fossa ovalis.

Fossa ovalis type aneurysms are further classified according to direction of maximal excursions.

- **Type 1** with maximal excursions towards right atrium and
- **Type 2** with maximal excursions towards left atrium.

Type 1 has further two types i.e. **type 1A** with oscillations less than 5mm and **type 1B** with oscillations more than 5mm. 

Type 2 aneurysms were characterized by markedly redundant fossa ovalis membrane protruding maximally into left atrium. All type 2 aneurysms were mobile with movement synchronized to cardiac cycle. There was increased excursion toward the left atrium during inspiration and more excursions toward the right atrium during expiration. Atrial septal defect (ASD) was more common in Type 1B implying that presence of ASD could induce a phasic oscillation of atrial septal aneurysm.

Silver and Dorsy (3) defined atrial septal aneurysm as protrusion of 10mm or more beyond septal plane relatively less restrictive as compared to definition given by Hanley and colleagues.

Pearson AC and coworkers also defined ASA as septal protrusion of 10mm either side or total excursion of 10mm. (7). Mugge and coworkers also used same criteria i.e. 10mm (1).

To add to the confusion Echocardiographic Classifications of Atrial Septal Aneurysm Motion has been proposed at different stages as follows.

**Alexander Oliva-Reyes et al (17) studied 205 patients and suggested the following classification:**

- **Type 1R:** The ASA protrudes from the midline of the atria to the right atrium throughout the cardiorespiratory cycle.
**Type 2L:** The ASA protrudes from the midline of the atrial septum to the left atrium throughout the cardiorespiratory cycle.

**Type 3RL:** The maximal excursion of the ASA is toward the right atrium with a lesser excursion toward the left atrium.

**Type 4LR:** The maximal excursion of the ASA is toward the left atrium with a lesser excursion toward the right atrium.

**Type 5:** The ASA movement is bidirectional and equidistant to the right as well as to the left atrium during the cardiorespiratory cycle.

Carlo Longhini and coworkers (18) studied 23 cases and suggested following classification:

**Type 1:** The ASA projects into the right atrium during diastole, with early systolic bulging into the left atrium, followed by a rightward crossing-over motion in mid-systole and during inspiration or expiration.

**Type 2:** Sustained rightward deviation during expiration and a leftward motion only during inspiration in early ventricular systole.

**Type 3:** The ASA remains in the right atrium, with an undulating motion during all phases of the cardiorespiratory cycle.

J Hanley PC, Tajik AJ and others (2) studied 80 cases and gave a detailed classification as already mentioned above. According to motion of aneurysm they classified ASA as follows:

**Type 1A:** The bulging in the right atrium is motionless.

**Type 1B:** The bulging is confined to the right atrium but with rapid phasic oscillation during inspiration.

**Type 2:** The ASA protrudes maximally into the left atrium and is accompanied by excursion into the right atrium.

**Associated cardiac abnormalities**

Atrial septal aneurysms may be an isolated anomaly or else may be associated with other abnormality. Commonest association is patent foramen ovale (PFO). Silver and Dorsy found patent foramen ovale in 8 out of 16 patients (3). Other common associations are Atrial septal defect (2), mitral valve prolapse (4,5) tricuspid valve prolapse, marfans syndrome, sinus of Valsalva aneurysm, aortic dissection (1). Shunt across ASA is picked up more frequently in transesophageal echocardiography as compared with transthoracic echocardiography (1). Association with mitral and tricuspid valve prolapse and other abnormalities like Marfans syndrome, sinus of valsalva aneurysm may point to common inherent connective tissue deficiency (6). Familial clustering of ASA has also been reported (13).

**Clinical Manifestations**

ASA would be taken as incidental finding of no clinical significance. But number of studies has underscored its clinical implications. Two important manifestations attributed to ASA are atrial arrhythmias and arterial embolism.

**Arrhythmias**
IAS aneurysm can act as an arrhythmic focus, generating focal atrial tachycardias. Hanley et al (2) noted atrial arrhythmias in 20 out of 80 patients (25%). Mugge and coworkers (1) in a multicenter study of 195 patients found atrial tachyarrhythmia in 47 patients (24%). In this study 28 patients (>14%) had atrial fibrillation. Schneider B et al (8) reported prevalence of atrial tachyarrhythmia in 26% cases of ASA. Mechanism of increased prevalence of atrial tachyarrhythmia in ASA is not clear though redundancy of atrial septum could be responsible for pathogenesis of arrhythmia.

**Embolism**

Arterial embolism is another complication associated with ASA. IAS aneurysms tend to aggravate stasis of LA blood flow and predispose to minute LA clots and systemic thromboembolism. Statistical association has been found between ASA and arterial embolism in various studies. In fact cardiac embolism has been reported in 20-52% cases of ASA. Gallet B et al reported embolic stroke in 2 of 10 patients (9). Belkin RN and coworkers (10) reported cerebrovascular event in 10 of their 36 consecutive patients (28%) with ASA. Pearson AC et al (7) studied 410 patients of which group 1 (133) were referred for assessment of source of emboli and Group 2 (277) were referred for other reasons. ASA was diagnosed in 32 patients (8%). Transthoracic echo picked up only 12 cases. Transesophageal echo was more sensitive in picking up aneurysm. ASA was significantly more common in patients with stroke than in patients without stroke (15% vs. 4%, p<0.05). Most of patients had left to right shunt (70%). Agmon Y et al (11) reported prevalence of ASA in 7.9%cases of cerebrovascular accident compared to 2.2% in general population in their study. In a retrospective study by Mugge and coworkers found that patients with ASA especially those with shunts showed increased frequency of clinical events in past compatible with cardiogenic embolism. In substantial number of patients ASA was only source of embolism as judged by transesophageal echocardiography (1). Salmasi AM reported higher prevalence of ASA and patent foramen ovale in Afro-Caribbean population as compared to Indo-Asians. This could be a possible cause of increased incidence of strokes in Afro-Caribbeans. (12). Mechanism of cardioembolic stroke could be right to left shunting as is detected in most of cases with ASA or thrombogenic properties of aneurysm itself. A non ejection click may be occasionally heard possibly as the IAS aneurysm bulges and tenses within LA/RA cavity thus ASA is one of the causes of systolic click (20).

**Diagnosis**

By far the most important investigation to diagnose ASA is echocardiography. Usually ASA is picked up on routine echocardiography or more commonly in patients referred for evaluation for cardioembolic cerebrovascular stroke and peripheral embolism. Transesophageal echo is more sensitive in picking up ASA compared to transthoracic echo. (1, 7) Cardiac Computed Tomography and MRI are also useful for diagnosis of ASA (14, 15)

**Treatment**

Uncomplicated and isolated ASA does not need any specific treatment. It needs follow up. Patient should be evaluated for presence of thrombus in aneurysm. Therapeutic options for prevention of recurrent stroke in patients with an atrial septal abnormality, including patent foramen ovale (PFO), ostium secundum atrial septal defect (ASD), and atrial septal aneurysm (ASA), are medical therapy with antiplatelet agents or anticoagulants, and surgical or percutaneous closure of the defect.
In presence of shunt to prevent recurrent paradoxical embolism it is preferable to close the shunt and transcatheter procedure is now commonly used for closure of shunt and is safe and effective (16). Although in a landmark CLOSURE 1 trial published recently did not prove any superiority of closure of PFO over adequate medical treatment. (21).

In case of atrial arrhythmia specific treatment is given. In case of embolic episode patient needs antiplatelet drugs and preferably oral anticoagulation for secondary prevention of cardioembolic episode.

The efficacy of aspirin therapy is suggested by the French PFO-atrial septal aneurysm (ASA) study, which found that among the 216 patients with a cryptogenic stroke who had a PFO alone, the incidence of a recurrent stroke on aspirin was only 2.3 percent after four years, a value that was comparable to the 4.2 percent risk in patients with neither a PFO nor an ASA (19).

References:


Conflict of Interest: None.
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