Original Research Article

Sinus venosus atrial septal defect with partial anomalous pulmonary venous return: Case series

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Abstract

Background: Sinus venosus atrial septal defect (SVASD) is a rare adult congenital heart disease where there is shunting of blood from the systemic to the pulmonary circulation. It is commonly associated with partial anomalous pulmonary venous return (PAPVR). These patients may present with dyspnea, chest pain and palpitations and signs like tachycardia and murmur can be encountered. Cases of secondary pulmonary arterial hypertension have been reported. We here present a case series of five patients with SVASD with PAPVC. Diagnosis of SVASD was made on transthoracic echocardiography (TTE) and to look for PAPVC Cardiac CT (computed tomography) was done on multi-detector CT scanner. Treatment options include conservative management and surgical repair with ASD patching, intracardiac baffle, anomalous vein anastomosis, systemic vein translocation and Warden procedure inter alia.

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Received Date: 08/12/2022 Revised Date: 12/01/2023 Accepted Date: 02/02/2023

Access this article online

Quick Response Code:

Website: www.medpulse.in

DOI: https://doi.org/10.26611/10132611

INTRODUCTION

Atrial septal defect are the most common congenital heart defect after ventricular septal defects (VSDs) and the most common to become symptomatic in adulthood.[1] Atrial septal defects (ASD) account for 5% to 10% of all cases of congenital heart disease and as many as 30% of cases of congenital heart disease presenting in adulthood. Sinus venosus ASD account for 10% of ASDs.[2] PAPVR also known as partial anomalous pulmonary venous circulation (PAPVC) is the anomalous circulation where some of the pulmonary veins, but not all, drain into the systemic circulation rather than in the left atrium.[3]

CASE 1

A 37-year-old male known case of ASD and hypertension presented with complains of chest pain, heaviness in chest, facial puffiness since 1 year, breathlessness since 2 months. He had no history of TB or smoking.

His transthoracic echocardiography reveals atrial septal defect of 14mm with dilated RA-RV with mild pulmonary arterial hypertension with good LV function. His cardiac CT scan was done.

How to cite this article: Shireen Ramteke, Bhagyashree Bhoir, Avinash Gutte. Sinus venosus atrial septal defect with partial anomalous pulmonary venous return: Case series. *MedPulse International Journal of Radiology.* April 2023; 26(1): 01-05. http://www.medpulse.in/Radio%20Diagnosis/

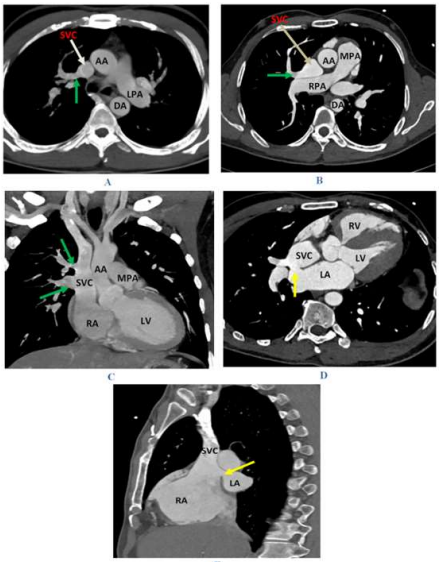

Figure 1: CT cardiac findings showing
A: Axial image shows right upper lobe pulmonary vein draining into SVC (green arrow).
B: Axial image shows right middle lobe pulmonary vein draining into SVC (green arrow).
C: Coronal reformatted image with maximum intensity projection (MIP) showing both right upper and middle lobe pulmonary veins draining into SVC separately (green arrows).
D: Axial reformatted image showing superior SVASD (yellow arrow).
E: Sagittal image showing SVASD (yellow arrow). Communication between the LA and SVC is seen due to SVASD.
AA: Ascending aorta. DA: Descending aorta. MPA: Main pulmonary artery. LPA: Left pulmonary artery. RPA: Right pulmonary artery. SVC: Superior vena cava. RA: Right atrium. RV: Right ventricle. LA: Left atrium. LV: Left ventricle.

CASE 2

A 42-year-old female presented with chest pain and breathlessness. Her transthoracic echocardiography was performed which revealed sinus venosus atrial septal defect of 26mm. Dilated right atrium and right ventricle with moderate tricuspid regurgitation and mild aortic regurgitation was seen. Patient also had moderate pulmonary arterial hypertension good LV function. Later her CT cardiac scan was performed.

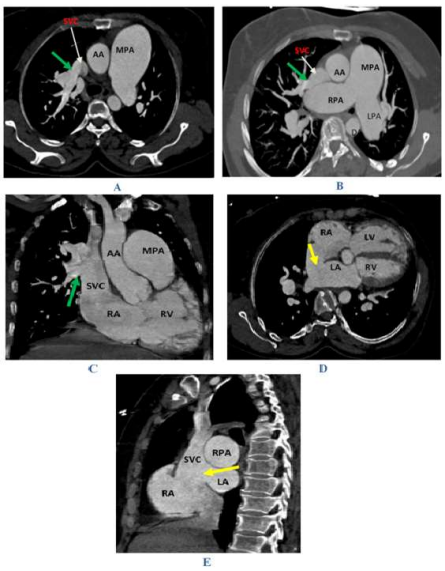

Figure 2: CT cardiac findings
A: Axial image showing right upper lobe pulmonary vein draining into SVC (green arrow).
B and C: Axial and coronal images with maximum intensity projection showing right middle lobe pulmonary vein draining into SVC (green arrow). MPA, RPA and LPA appear dilated.
D and E: Axial and sagittal images showing atrial septal defect (superior sinus venosus type) - yellow arrow. The superior and eccentric position of the defect (yellow arrow) along the interatrial septum allows communication between the LA and SVC. Cardiomegaly is appreciated with dilated right atrium and right ventricle.

CASE 3

A 3-year-old male child was presented with complains of breathlessness. He had no history of fever or cough.
His transthoracic echocardiography revealed 10mm sinus venosus ASD with left to right shunt with dilated right atrium and pulmonary arterial hypertension. His Cardiac CT scan was done under sedation.

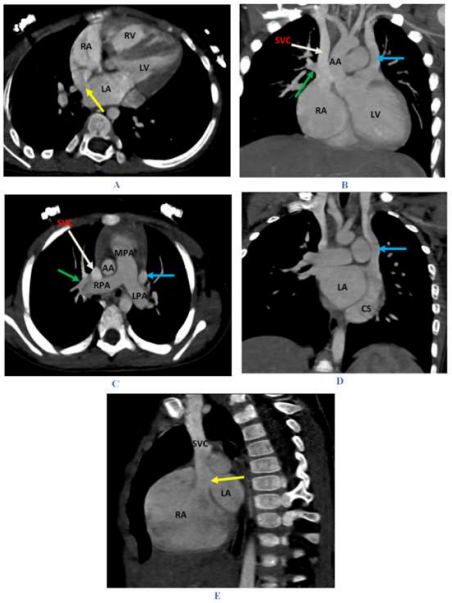

Figure 3: CT cardiac findings

A: Axial image shows superior sinus venosus ASD (yellow arrow).

B and C: Coronal and axial image with maximum intensity projection showing right upper lobe pulmonary vein draining into superior vena cava (green arrow). Dilated main pulmonary and left and right pulmonary arteries are seen. Persistent left side superior vena cava is seen (Blue arrow).

D: Coronal and reformatted image shows persistent left sided superior vena cava (blue arrow) which is draining into coronary sinus into right atrium.

E: Sagittal image showing SVASD superior type (yellow arrow). Communication between the LA and SVC is seen secondary to SVASD.

Abbreviation: CS: Coronary sinus. Rest of the abbreviations as in Figure 1.

CASE 4

A 40 year old female patient presented with complains of chest pain on and off associated with dyspnea on exertion since one year. She did not have fever, cough, cold, palpitation or generalized weakness.

She was a known case of hypothyroididsm on medication since one year.

No history of hypertension or diabetes mellitus.

Her transthoracic echocardiography revealed paradoxical septal motion of inter-ventricular septum with 22 mm ASD, severe PAH, dilated right atrium and right ventricle with good LV function. Her CT cardiac scan was performed.

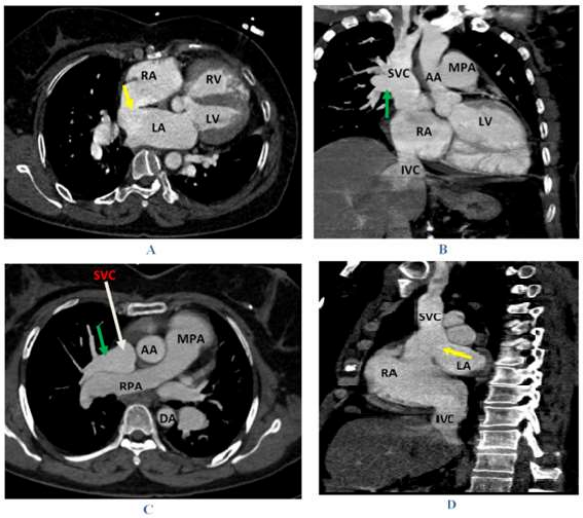

Figure 4: CT cardiac findings

A: Axial image shows superior sinus venosus ASD (yellow arrow).

B and C: Coronal and axial image with maximum intensity projection showing right upper lobe pulmonary vein draining into SVC (green arrow). Dilated main pulmonary and right pulmonary artery is seen.

D: Sagittal image showing SVASD superior type (yellow arrow). Communication between the LA and SVC is clearly seen due to eccentric position of ASD.

Abbreviation: IVC: Inferior vena cava. Rest of the abbreviations as in Figure 1.

CASE 5

A 37 year old female patient was presented with chest pain on exertion with breathlessness since 2 years. She had no history of palpitation, fever, cough or weakness. No history of hypertension or diabetes mellitus.

Her transthoracic echocardiography revealed sinus venosus ASD and dilated right atrium and right ventricle with pulmonary hypertension. Her Cardiac CT scan was done.

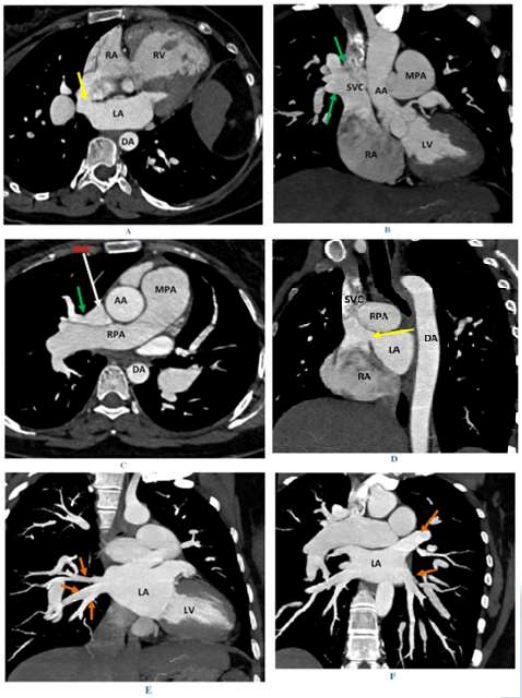

Figure 5: CT Cardiac findings

A: Axial image shows superior sinus venosus ASD (yellow arrow).

B and C: Coronal and axial image with maximum intensity projection showing Right superior pulmonary vein is draining into superior vena cava (green arrow). Dilated main pulmonary and right pulmonary artery is seen.

D: Sagittal image showing SVASD superior type (yellow arrow). Communication between the LA and SVC is seen due to SVASD.

E and F: Five pulmonary veins are seen draining into morphological left atrium (three on right and two on left shown in orange arrows). Abbreviations as in Figure 1.

DISCUSSION

Atrial septal defect (ASD) is one of the most common types of congenital heart defects, accounting for 5% to 10% of all cases of congenital heart disease.[2] An atrial septal defect occurs when there is a failure to close the communication between the right and left atria. It encompasses defects involving both the true septal membrane and other defects that allow for communication between both atria.[6]

Atrial septal defects occur as singular defects. However, ASDs can be seen in association with Mendelian inheritance, aneuploidy, transcription errors, mutations, and maternal exposures. Atrial septal defects are noted in patients with Down syndrome, Treacher-Collins syndrome, Thrombocytopenia-absent radii syndrome, Turner syndrome, and Noonan syndrome. These syndromes occur as a result of Mendelian inheritance. Maternal exposure to rubella and drugs, such as cocaine and alcohol can also predispose the unborn fetus to develop an ASD. Additionally, ASDs have been associated with familial genetic disorders and conduction defects.[6] The sinus venosus ASD is when the location of the defect is eccentric either superiorly or inferiorly in the interatrial septum at the sites of inflow of the superior vena cava (SVC) and inferior vena cava (IVC), respectively.[2] Sinus venosus defects (SVDs), represent about 2%–10% of all atrial septal defects. The remainder is composed of ostium secundum (70%), ostium primum (20%) and unroofed coronary sinus (<1%) defects. Approximately 90% of SVDs are associated with partial anomalous pulmonary venous return (PAPVR). The superior SVD is associated with PAPVR from the right upper lung lobe into the systemic venous circulation. Inferior SVDs are less common and result from anomalous communication of the RA and inferior vena cava (IVC) junction with the left atrium. They are less commonly associated with PAPVR of the right lower lobe pulmonary vein into the IVC.[5] The sinus venosus defect typically results in a left-to-right shunt at the level of the atria. The presence of PAPVR compounds the problem with additional left-to-right shunt pathways.[4] Four types of PAPVR have been described; supra-cardiac, cardiac, infracardiac and mixed. In our case series we present supra-cardiac PAPVR with sinus venosus ASD.[8] Most patients are asymptomatic but as cardiac failure develops they may present with shortness of breath, palpitations, and weakness. Chest auscultation classically reveals an ejection systolic murmur heard at the left upper sternal border, attributed to increased flow across the pulmonary valve rather than blood shunting across the defect itself [1]. Complications associated with ASDs are atrial dysrhythmias, pulmonary arterial hypertension, right-sided congestive heart failure, transient ischemic attack/stroke and eisenmenger syndrome.[6] Diagnostic imaging is important in determining the size of the defect and is crucial in determining treatment options. A transthoracic echocardiogram is the gold standard imaging modality. A transthoracic echocardiogram allows one to detect the size of the defect, understand the direction of blood flow, find associated abnormalities (involvement of the endocardial cushions and atrial-ventricular valves), examine the heart for structure and function, estimate pulmonary artery pressure, and estimate the pulmonary/systemic flow ratio. A transesophageal echocardiogram is a better tool for diagnosing the rarer cardiac defects.[6] Though echocardiography is the gold standard for evaluation of ASDs, other diagnostic modalities include cardiac CT and MRI. Both CT and MRI examine structures surrounding the heart and in the thoracic cavity. CT and MRI also have refined assessment of pulmonary venous anatomy with variants and anomalies being increasingly recognized. Anomalous pulmonary venous circulation can be asymptomatic or non-specific in presentation and the presence of right heart dilatation

should prompt particular scrutiny of the pulmonary venous anatomy. Pulmonary veins are a recognized source of ectopic foci triggering episodes of atrial fibrillation. Therefore mapping of the relevant anatomy is crucial, to facilitate clinical decisions and planning of surgical and catheter based interventions such as RFA. For these reasons, knowledge of anomalous pulmonary venous drainage and variant anatomy is important and the cardiac imaging specialist is important, not only in these cases but also in the diagnostically challenging patient.[7] Chest x-ray findings are not as helpful diagnostically, though the chest x-ray assists providers to monitor clinical status by identifying cardiomegaly and pulmonary artery enlargement. Cardiac catheterization is contraindicated in patients who are young and present with small, uncomplicated ASDs.[6]

Therapeutic options include surgical repair with ASD patching, intracardiac baffle, anomalous vein anastomosis, systemic vein translocation and Warden procedure inter alia.[8] Indications for treatment include stroke, a hemodynamically significant shunt greater than 1.5:1 and evidence of systemic oxygen desaturation. If an ASD requires closure, options include percutaneous and surgical intervention. Percutaneous transcatheter closure poses less risk for the patient, but it is only useful for the closure of ostium secundum defects. Percutaneous transcatheter ASD closure has a post-procedural complication risk of 7.2% compared to a post-surgical complication risk of 24%. Complications associated with percutaneous closure risk include arrhythmias, AV blocks, cardiac erosion, and thromboembolism. Surgical closure of atrial septal defects requires the placement of a patch over the lesion through an incision in the right atrium.[6]

CONCLUSION

Atrial septal defects can present with pulmonary arterial hypertension and can develop cardiac failure in adults. With the increased use of CT, careful assessment for these defects, and their associated pulmonary venous anomalies in patients with enlarged central pulmonary vessels, may be beneficial as surgical repair of these defects can improve outcomes.

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Source of Support: None Declared
Conflict of Interest: None Declared