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CASE REPORT

HEART CARE TEAM/MULTIDISCIPLINARY TEAM LIVE

Massive Right Ventricle Enlargement in Unrepaired Sinus Venosus Atrial Septal Defect

Repair or Not?

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ABSTRACT

Sinus venosus atrial septal defect (SVASD) with partial anomalous pulmonary venous return is a rare congenital heart defect that may present in adulthood when right heart enlargement has already occurred. We describe a case of unrepaired sinus venosus atrial septal defect and partial anomalous pulmonary venous return with massive right heart enlargement and recurrent atrial arrhythmias. (J Am Coll Cardiol Case Rep 2023;28:102091)

HISTORY OF PRESENTATION

A 37-year-old, previously healthy man presented with midsternal intermittent chest pain worsening on exertion and improving upon rest, shortness of breath, palpitations, and right lower quadrant abdominal pain. His vital signs were as follows: blood pressure of 137/97 mm Hg, heart rate of 102 beats/min, temperature of 36.7 °C; respiratory rate of 16 breaths/min, and oxygen saturation of 100% in room air. Physical examination findings were notable for a parasternal heave; displaced point of maximal impulse to the left midaxillary line; tachycardia with an irregularly irregular rhythm; a II/VI systolic ejection murmur at the left upper sternal border; jugular venous distention with prominent V waves; normal respiratory effort with clear lungs; and a distended, pulsatile liver.

LEARNING OBJECTIVES

- To demonstrate the importance of further evaluation when right-sided cardiac enlargement presents out of proportion to left-sided heart disease.
- To understand the role of multimodality imaging and cardiac catheterization for eligibility of surgical repair of SVASD and PAPVR.
- To appreciate the multiple arrhythmias that can occur in patients with late repair of SVASD and PAPVR.

INITIAL INVESTIGATIONS

Blood work showed normal complete blood count, creatinine of 0.99 mg/dL, troponin I of <0.01 ng/mL, and brain natriuretic peptide of 448.6 pg/mL. Liver evaluation revealed hepatomegaly without cirrhosis, normal hepatic enzymes, and normal synthetic liver function. Electrocardiogram showed atrial fibrillation

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ABBREVIATIONS AND ACRONYMS

ASD = atrial septal defect

LV = left ventricular

PAPVR = partial anomalous pulmonary venous return

RA = right atrial

RV = right ventricular

SVASD = sinus venosus atrial septal defect

TR = tricuspid regurgitation

at 100 beats/min with some aberrancy (Ashman's phenomenon) and ST-segment depression in V_4-V_6 (Figure 1A). Chest x-ray film revealed massive cardiomegaly. Transthoracic echocardiogram demonstrated severe tricuspid regurgitation (TR), severe right atrial (RA) and right ventricular (RV) dilation with normal systolic function, and normal size but moderately depressed function of the left ventricle (LV) (Video 1).

QUESTION 1: WHAT IS THE DIFFERENTIAL DIAGNOSIS, AND WHAT TEST WOULD YOU PERFORM NEXT?

The differential diagnosis for chest pain in a young adult is broad and includes pulmonary embolism; aortic dissection; pericarditis; pneumonia; acid reflux; costochondritis; and, rarely, myocardial ischemia. During the workup for this, the patient was incidentally noted to have right-sided cardiac enlargement with severe TR. This along with atrial fibrillation suggests long-standing pathology with a differential that includes intracardiac or extracardiac shunts, such as atrial septal defects (ASDs), partial anomalous pulmonary venous return (PAPVR), pulmonary-to-systemic arteriovenous connections or large coronary artery fistulas, and underlying valvular conditions such as Ebstein anomaly or congenitally abnormal pulmonary valve. Noncardiac causes of right heart enlargement include chronic pulmonary emboli or long-standing pulmonary hypertension. This patient needed evaluation for additional cardiac etiologies and for left atrial appendage thrombus before cardioversion, so a transesophageal echocardiogram was performed next.

Transesophageal echocardiogram demonstrated severe dilation of the RA and RV with preserved systolic function, severe TR, LV ejection fraction of 40%, and a sinus venosus atrial septal defect (SVASD) with predominately left-to-right shunting. Cardiac magnetic resonance was then performed to better asses the size and function of the RV. It confirmed a large SVASD measuring 3.8 cm, estimated a Q_p:Q_s of 3.6:1, and demonstrated a massively enlarged RV (RV end-diastolic volume index of 499 mL/m^2) and RA (81.5 cm^2) (Figure 2).

QUESTION 2: WHAT IS THE MOST COMMON DEFECT ASSOCIATED WITH SVASD?

PAPVR of 1 or more right-sided pulmonary veins is the classic congenital anomaly associated with SVASD.

To evaluate pulmonary venous anatomy, a computed tomography angiogram was performed and indeed revealed anomalous drainage of the right upper and middle pulmonary veins draining to the superior vena cava (Figure 3).

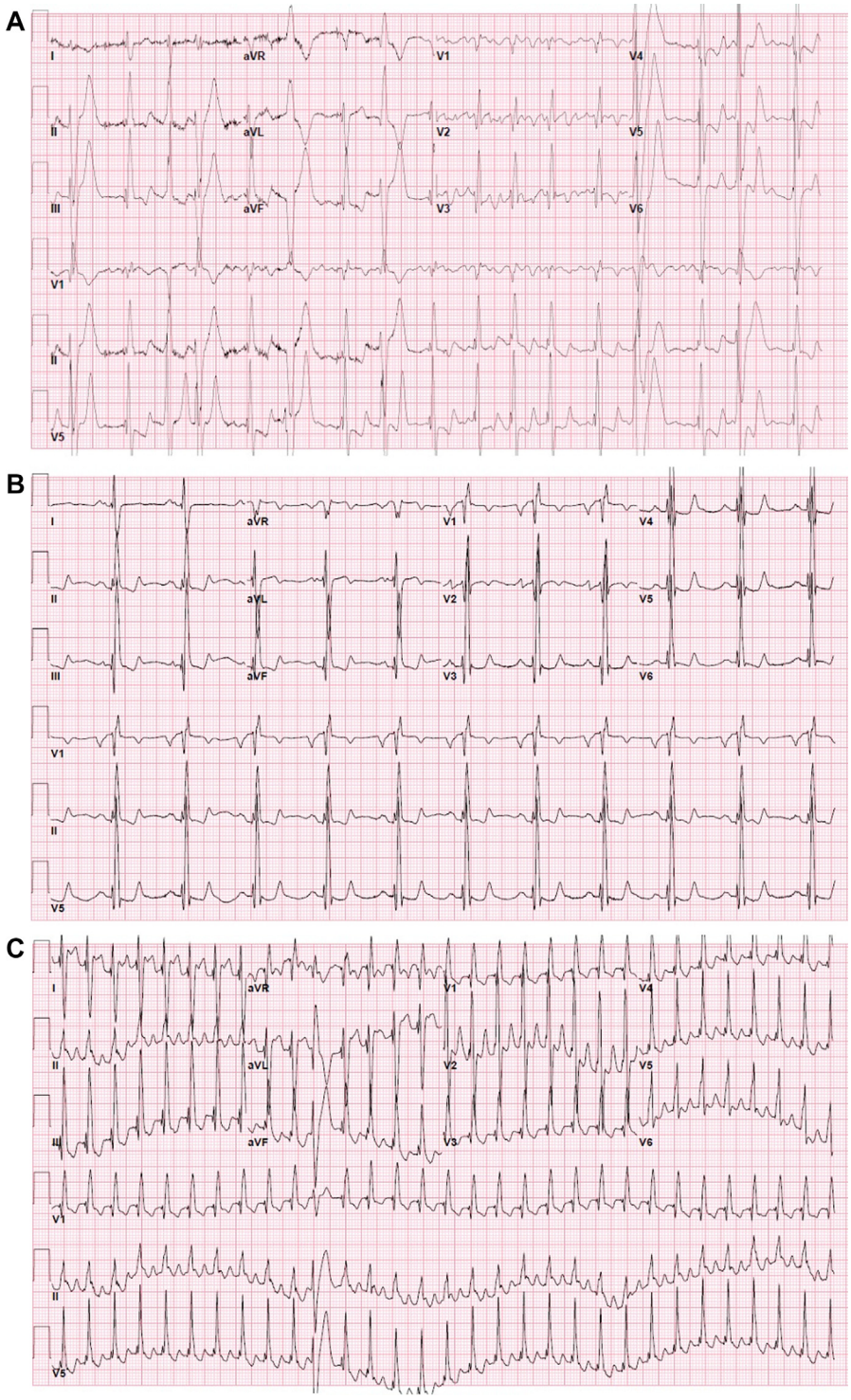
The patient was started on systemic anticoagulation for stroke prevention. He was given a diuretic agent and underwent a successful cardioversion with restoration of sinus rhythm (Figure 1B). Cardiac catheterization revealed a cardiac output of 3.3 L/min, Q_p:Q_s of >4:1, pulmonary vascular resistance of 0.5 WU, pulmonary artery pressure of 44/11 mm Hg with a mean of 20 mm Hg, and descending aorta pressure of 114/60 mm Hg.

QUESTION 3: WHAT ARE THE CRITERIA FOR SURGICALLY REPAIRING SVASD AND PAPVR?

Patients with SVASD and PAPVR should undergo surgical repair if the defect causes impaired functional capacity, right-sided cardiac enlargement, or a hemodynamically significant shunt (Q_p:Q_s ≥1.5:1) so long as the patient is a good surgical candidate without cyanosis and with systolic pulmonary arterial pressure of <50% systemic arterial pressure and R_p:R_s of <1/3 (Class of Recommendation: 1, Leve of Evidence: B-NR).[1] However, there is a paucity of literature regarding the upper limit of right heart dilation at which surgical correction has been shown to be beneficial in terms of decreased RV size and preserving RV function. Recently, an RV end-systolic volume index of 75 mL/m^2 has been suggested as a possible threshold before the RV size normalizes after the closure of an ASD.[1] Our patient presented with an RV end-systolic volume index (395 mL/m^2) that was more than 5 times this threshold, raising the question if surgical repair of the SVASD and PAPVR would lead to normalization of the right heart parameters. At the same time, leaving the patient with a very large unrepaired shunt would be deleterious.

The patient met criteria for surgical repair, and following medical optimization with diuresis and milrinone for RV support, he underwent double-patch SVASD and PAPVR repair, tricuspid valve annuloplasty, cryomaze pulmonary vein isolation, cavotricuspid isthmus ablation, and RA reduction. A patch fenestration was created as a "pop-off" in case of significant RV dysfunction. The immediate postsurgical period was complicated by ventricular tachycardia at the termination of cardiopulmonary bypass, treated with amiodarone, lidocaine, and defibrillation. For RV support, he was started on inhaled nitric oxide and inotropic support that was gradually weaned off. He developed sinus node dysfunction postoperatively with a return to an ectopic atrial rhythm after the antiarrhythmic

FIGURE 1 **Electrocardiograms**



(A) Initial electrocardiogram revealing atrial fibrillation with rapid ventricular response, aberrant conduction, ST-segment depressions in the lateral precordial leads, and evidence of right ventricular hypertrophy. (B) Following cardioversion, electrocardiogram showed normal sinus rhythm with right atrial abnormality and biventricular hypertrophy. (C) Electrocardiogram showing intra-atrial re-entrant tachycardia with 2:1 conduction and a premature ventricular contraction. (D) Following ablation of the intra-atrial re-entrant tachycardia, electrocardiogram revealed junctional rhythm. (E) Electrocardiogram 2 days post- revealed recovery of a low atrial rhythm.

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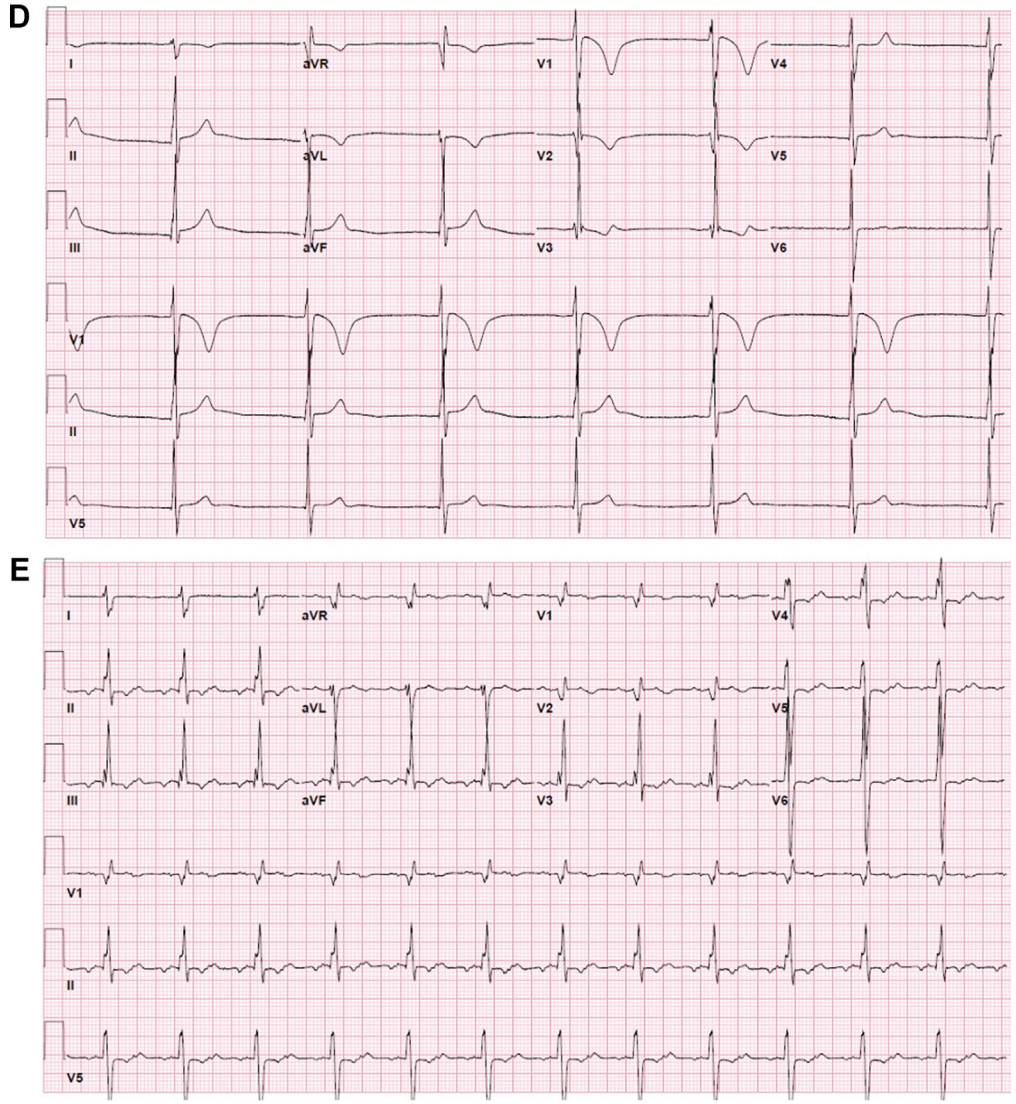
FIGURE 1 Continued



medications were discontinued. His echocardiogram at 1-week postrepair showed improved RV size, though it still remained moderately dilated and with moderately depressed function. The LV was mildly dilated with improved LV function (Video 2). He remained stable and was discharged on postoperative day 11.

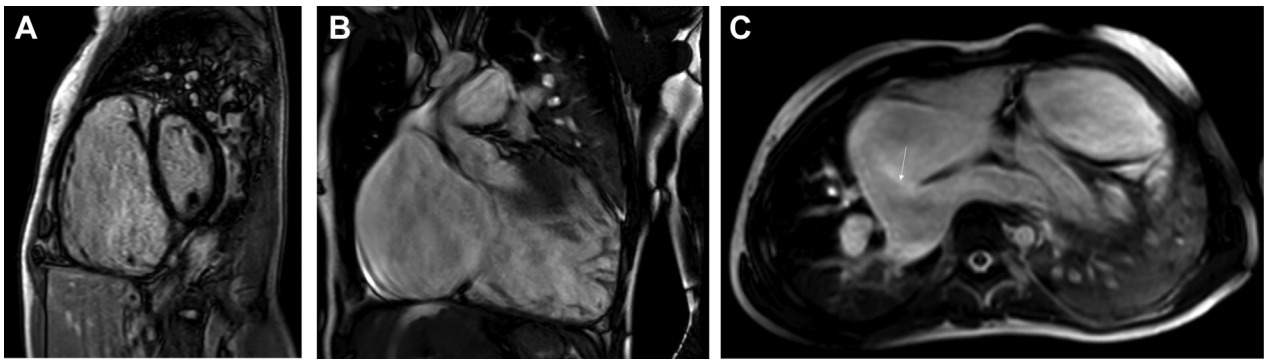
QUESTION 4: WHAT ARE THE LONG-TERM COMPLICATIONS OF LEFT-TO-RIGHT SHUNTS?

Atrial septal defects affect up to one-third of adults with congenital heart disease.[2] SVASD is a rare type of ASD often associated with PAPVR and, when left unrepaired, can lead to right-sided volume overload with right heart enlargement; pulmonary overcirculation and possible pulmonary arterial hypertension; sinus node dysfunction; atrial tachyarrhythmias; and, rarely, end-organ dysfunction secondary to right heart failure. A high index of suspicion for a left-to-right shunt should always be kept when patients have right-sided heart dilation that is out of proportion to their left-sided heart disease or any underlying pulmonary disease.

QUESTION 5: FOR WHICH ARRHYTHMIAS ARE PATIENTS WITH SVASD AND PAPVR AT RISK?

Atrial arrhythmias are among the leading causes of short- and long-term comorbidities in patients with unrepaired or late repair of any left-to-right shunt.[3]

FIGURE 2 Cardiac MRI Findings



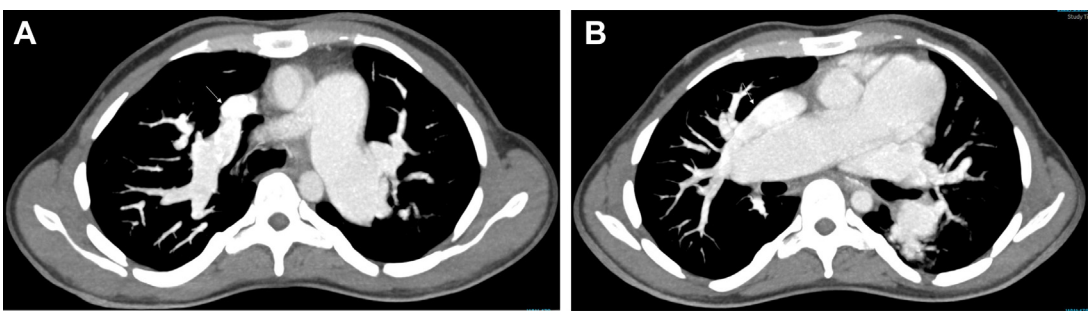
Cardiac magnetic resonance showed massive right atrial and right ventricular enlargement in the (A) sagittal and (B) coronal views. (C) In axial imaging, the sinus venosus atrial septal defect is shown.

Sinus node dysfunction is also relatively frequent in patients with repaired SVASD.[4] The onset of sinus node dysfunction has been associated with anatomic abnormalities, fibrosis, or trauma during repair to the internodal tracts.[5] Our patient developed sinus node dysfunction shortly after the surgical repair (**Figure 1B**); however, a low atrial rhythm predominated, and he was discharged in a stable rhythm with atrioventricular synchrony. The prevalence of atrial fibrillation ranges between 20% and 50% in patients with ASD.[6] The volume load and stretch of the left atrium are the most like mechanisms for atrial fibrillation. Another proposed mechanism for the onset of atrial fibrillation is RA volume overload, leading to conduction delay or block in the intercaval region and Bachmann's bundle.[7]

FOLLOW-UP

Transthoracic echocardiogram at 2 months post-surgery showed an LV ejection fraction of 52%, moderate RV dilatation with mildly decreased function, severe RA dilation (area: 33.8 cm^2), a small left-to-right shunt across the surgical atrial septal fenestration, trivial TR, and no evidence of pulmonary hypertension. He presented with interatrial re-entry tachycardia (**Figure 1C**) 10 weeks post-operatively with associated biventricular dysfunction that was successfully ablated. He again experienced junctional rhythm (**Figure 1D**) in the immediate post-ablation period secondary to sinus node dysfunction, highlighting the multiple types of arrhythmias patients with late repair of SVASD and PAPVR are at risk

FIGURE 3 Cardiac CT Findings



(A) Computed tomography axial image showed the right upper pulmonary vein draining anomalously into the superior vena cava (arrow). (B) Computed tomography axial image showed the right middle pulmonary vein draining anomalously into the superior vena cava (arrow). Pulmonary arteries were severely dilated.

for and the complexity of treating them. He had recovery of a low atrial rhythm (Figure 1E). The severe biventricular dysfunction improved with directed treatment of the arrhythmia. Currently, his RV remains moderately enlarged with moderate systolic dysfunction and trivial TR. The patient has resumed normal activity with no limitations.

CONCLUSIONS

Longstanding left-to-right shunting secondary to SVASD and PAPVR can result in massive right heart dilation, for which the potential of reverse remodeling is unknown. We present a patient with late repair of SVASD and PAPVR complicated by atrial arrhythmias who, in the short-term, has demonstrated reduction in the size of the RV with moderate RV dysfunction and normalization of LV systolic function.

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KEY WORDS atrial arrhythmias, atrial septal defect, multimodality imaging, right ventricle

APPENDIX For supplemental videos, please see the online version of this paper.