

The role of cardiac MRI in the diagnosis and management of sinus venosus atrial septal defect

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ABSTRACT

Sinus venosus atrial septal defects (SV-ASDs) are inter-atrial communications caused by a deficiency of the common wall between the superior or inferior vena cava and the right-sided pulmonary veins. The diagnosis can be challenging, especially in adults with delayed presentation. We present images that illustrate an example of the role of cardiac magnetic resonance imaging (CMRI) in the diagnosis and follow-up of a patient with SV-ASD.

Keywords: Atrial septal defect, adult congenital heart disease, cardiac MRI

INTRODUCTION

Sinus venosus atrial septal defects (SV-ASDs) are inter-atrial communications caused by a deficiency of the common wall between the superior or inferior vena cava and the right-sided pulmonary veins, and are typically associated with partial anomalous right pulmonary venous drainage (PAPVD). They account for 10-15% of all atrial septal defects. As with all ASDs, many patients with this type of defect can be free of symptoms even well into adulthood. This makes early diagnosis challenging and often difficult. Surgical repair is the only treatment for SV-ASD, and the most appropriate surgical strategy varies according to the configuration of the pulmonary veins. Late diagnosis and thus delayed surgical repair carries an increased risk of morbidity and mortality. We illustrate an example of the role of cardiac magnetic resonance imaging (CMRI) in the diagnosis and follow-up of a patient with SV-ASD.

CLINICAL SUMMARY

A 25-year-old asymptomatic male was incidentally detected to have a murmur. Clinical examination

revealed a prominent right ventricular (RV) impulse, a wide and fixed split of the second heart sound with a soft pulmonary component and an ejection systolic murmur at the left upper sternal border. Electrocardiogram (ECG) showed sinus rhythm with partial right bundle branch block pattern. Chest X-ray showed an enlarged right heart with increased pulmonary vascularity. A transthoracic echocardiogram (TTE) demonstrated a moderately dilated RV and right atrium with normal systolic ventricular function. There was trivial tricuspid valve regurgitation with a measured peak regurgitant gradient of 22 mmHg on Doppler evaluation. To further delineate a suspected ASD, a transesophageal echocardiogram (TEE) was performed. TEE demonstrated a SV-ASD with the right upper pulmonary vein connecting to the superior vena cava (SVC); however, the complete pulmonary venous anatomy was not adequately defined.

Hence, CMR examination (1.5 T) was performed. This included standard steady-state free precession (SSFP) cine imaging, breath-held fat suppressed three-dimensional SSFP pulse sequence, gadolinium-enhanced three-dimensional gradient-echo sequences for MR angiography (MRA), and velocity-encoded phase-contrast imaging for flow through the great vessels and shunt calculations (ratio of the net pulmonary flow/net aortic flow). Cine stacks using contiguous slices were obtained in the short axis of the heart and in a trans-axial plane through the entire cardiac mass.

CMRI confirmed a superior SV-ASD measuring 25 mm with anomalous drainage of the right upper pulmonary vein into a single right-sided SVC [Figure 1]. The other

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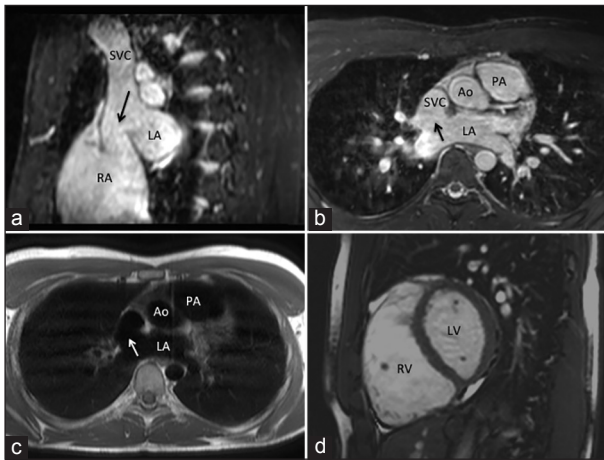


Figure 1: (a) Breath-held fat suppressed three-dimensional steady-state free precession (SSFP) pulse sequence in diastole in the sagittal view demonstrating sinus venosus atrial septal defect (SV-ASD) (arrow) between superior vena cava (SVC) and left atrium (LA). (b) Breath-held fat suppressed three-dimensional SSFP pulse sequence in diastole in the axial view demonstrating SV-ASD (arrow) between SVC and LA. (c) Turbo spin-echo black blood image in the same axial plane as ure 1b demonstrating SV-ASD (arrow) between SVC and LA. (d) SSFP image showing the dilated right ventricle (RV) and left ventricle (LV)

pulmonary veins were noted to be draining normally. The right-sided cardiac chambers were dilated with normal RV systolic function. The RV was not hypertrophied and the ventricular septal motion was normal. The indexed RV end-diastolic volume measured 198 ml/m² (normal range 55-105 ml/m²). The measured pulmonary to systemic blood flow (Q_p:Q_s) ratio was 2.1. The left ventricular size and function was normal.

The patient underwent surgery where a pericardial patch was used to redirect the anomalously draining right upper pulmonary vein to the left atrium and close the defect. Intraoperative TEE confirmed a good surgical result. The patient made a good recovery postoperatively.

At 1-year follow-up, he continued to be asymptomatic. The clinical findings of RV volume overload were no longer evident clinically. TTE showed persisting dilatation of the right heart chambers. CMRI showed no residual ASD with unobstructed and normally draining systemic and pulmonary venous flows. The indexed RV end diastolic volume measured 134 ml/m² (normal range 55-105 ml/m²), suggesting persisting moderate dilatation. The RV systolic function was preserved. There was normal size and function of the left ventricle.

DISCUSSION

This case highlights the increasingly important role played by CMRI in the diagnosis and management of SV-ASDs, especially in adults. This pathology can easily be ‘missed’ on TTE.^[1] TEE is a sensitive and cost-effective modality in diagnosing SV-ASDs. However, it

is an invasive procedure and may not always provide adequate anatomical and functional information.^[2,3] Importantly, both TTE and TEE can reliably estimate the right-heart pressure, a key pre-surgical criteria. Cardiac catheterization and contrast-enhanced computed tomography (CT) scan are other modalities that can be used; however, they involve exposure to ionizing radiation, and while the former is accompanied by the risks of an invasive procedure the latter does not always provide adequate functional information.^[4]

CMRI has inherent advantages over the traditionally used methods. CMRI can accurately quantify ventricular volumes and the magnitude of shunting.^[5] Muthurangu *et al.*, have noted that in certain situations, phase contrast MRI is more accurate than invasive cardiac catheterization in quantifying flow and shunt size.^[6] Gadolinium enhanced MRA is a sensitive method in detecting abnormalities of pulmonary venous return and can act as a surrogate for PA pressures along with detailed flow data.^[7] Additionally; CMRI is noninvasive, has high spatial resolution with large fields of view, and does not involve exposure to ionizing radiation. This allows for serial studies and documentation of normalization of the ventricular volumes, especially relevant in cases such as ours where, late diagnosis and repair is often associated with persisting cardiac dilatation and increased morbidity.^[8] These characteristics make CMRI an ideal imaging modality in the follow up of SV-ASDs.

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