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ATRIAL ARRHYTHMIAS

COMPLEX CASE STUDY

Pacemaker Considerations in Patients with Intracardiac Shunts

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ABSTRACT. A 74-year-old male with history of dyspnea on exertion, fatigue, and syncope presumed to be secondary to sick sinus syndrome and atrial flutter was referred for evaluation and management. Pre-procedural echocardiogram did not demonstrate any significant abnormalities. He underwent radiofrequency catheter ablation for atrial flutter. Fluoroscopy during the procedure demonstrated free movement of the ablation catheter between the right and left atria through the interatrial septum. Transesophageal echocardiogram using echo contrast as well as a contrast cardiac computed tomography scan confirmed the presence of a sinus venosus (SV)-atrial septal defect (ASD) with significant bidirectional flow as well as associated right ventricular dilatation. Because of the patient's significant symptoms of shortness of breath on exertion, palpitation, and sinus pauses associated with syncope in addition to signs of right heart failure and concerns for systemic embolization after pacemaker lead placement, the patient underwent surgical closure of the ASD followed by implantation of a dual-chamber pacemaker. This case demonstrates clinical features and challenges of managing atrial arrhythmias and device therapy in patients with unrepaired SV-ASD in the elderly population specifically those related to thromboembolic risks associated with pacemaker leads.

KEYWORDS. Atrial arrhythmias, cardiac pacing, congenital heart disease.

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Case

A 74-year-old male presented to an outside hospital with palpitations and syncope. He also complained of symptoms of dyspnea on exertion and frequent pre-syncope episodes that had progressed over the previous 6 months. At the time of presentation, he was diagnosed with new-onset atrial flutter and was referred to our center for further assessment and management.

His past medical history was significant for syncope 4 years earlier, which was not associated with any changes in position, prodrome, or post-ictal symptoms. A 24-h cardiac telemetry monitoring at that time did not reveal any arrhythmias. Prior transthoracic echocardiograms described right ventricular and bilateral atrial enlargement

with normal left ventricular size and function. An exercise nuclear perfusion stress test 1 year earlier was reported as normal. His other co-morbidities included hypertension, chronic right bundle branch block, dyslipidemia, obstructive sleep apnea, and diabetes.

The patient was admitted electively to the hospital to undergo atrial flutter ablation. Physical examination demonstrated a regular heart rate at 63 bpm and blood pressure of 130/70 mmHg. His SaO₂ was 95% at room air, and lung auscultation was unremarkable. Cardiac auscultation was only significant for a 2/6 holosystolic murmur at the apex with radiation to the axilla.

His medications included Coumadin, olmesartan, hydrochlorothiazide, Lasix, and insulin.

His electrocardiogram demonstrated typical atrial flutter with ventricular rate of 63 bpm, and right bundle branch block (**Figure 1**).

He underwent radiofrequency catheter ablation for typical atrial flutter after a limited transesophageal echocardiogram ruled out the presence of thrombus in the left atrial appendage. During the procedure, the ablation catheter

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Figure 1: An electrocardiogram demonstrated typical atrial flutter and right bundle branch block.

freely moved between the right and the left atria through the interatrial septum, as seen by fluoroscopy. After successful typical atrial flutter ablation, sinus rhythm was restored. However, there were episodes of symptomatic sinus bradycardia and sinus pauses suggestive of underlying sick sinus syndrome without being on any heart-rate slowing medications.

After atrial flutter ablation, a complete transthoracic echocardiogram was performed, which showed normal left ventricular size and function (left ventricular ejection fraction >55%), right ventricle dilatation without evidence of pulmonary hypertension, and moderate right atrial dilation. Agitated saline injection revealed the presence of right-to-left shunt across the interatrial septum without clear visualization of the location of shunting within the interatrial septum. There was no hemodynamically significant valvular abnormality.

A detailed transesophageal echocardiogram demonstrated a 3.0×1.9 cm sinus venosus (SV)-atrial septal defect (ASD) with significant bidirectional flow between the superior vena cava and the left atrium in addition to a patent foramen ovale. The right atrium and ventricle were dilated.

Contrast cardiac computed tomography confirmed a 3×2.7 cm diameter SV-ASD. The right superior and middle pulmonary veins emptied into the left atrium via separate ostia situated superiorly above the ASD and alongside the superior vena cava (Figures 2 and 3).

Progression of symptoms of dyspnea on exertion in previous months in the context of enlarged right atrium and ventricle suggested that the ASD and the associated left-to-right shunt were hemodynamically significant and due to right ventricular volume overload and pulmonary overcirculation. Furthermore, the implantation of a pacemaker in an un-repaired ASD would have potentially created the risk of paradoxical systemic embolization from the implanted leads. Therefore, the patient was assessed for surgical closure of the ASD before implantation of a dual-chamber pacemaker.

Coronary angiography did not show any evidence of significant epicardial coronary artery stenosis. Right heart catheterization showed normal pulmonary arterial pressures and hemodynamically significant left to right shunt at the level of the right atrium (Qp:Qs ratio of 1.55 and 7% oxygen saturation step up at the right atrial level).¹

Primary closure of the patent foramen ovale, pericardial patch repair of the SV-ASD, and patch augmentation of the superior vena cava were successfully performed. A dual-chamber permanent pacemaker was subsequently implanted (Figures 4 and 5).

The patient made a complete recovery and was discharged home without any complications. He was maintained on Coumadin. Holter monitoring before his 1-month follow-up did not show any evidence of atrial flutter or fibrillation and his symptoms of shortness of breath and fatigue had completely resolved.

Discussion

This case highlights some of the challenges related to the management of arrhythmias in elderly patients with symptomatic SV-ASD, including the risk of paradoxical

Figure 2: Transesophageal echocardiogram with color Doppler depicting left to right flow across sinus venosus atrial septal defect.

Figure 3: Transesophageal echocardiogram with color Doppler depicting left to right flow across sinus venosus atrial septal defect.

embolization after the implantation of an intravenous pacemaker.

SV-ASD should be suspected in patients with atrial arrhythmias who present with unexplained right atrial and ventricular enlargement. The diagnostic yield of transthoracic echocardiography for SV-ASD is between 12% and 44% and other imaging modalities such as transesophageal echocardiography, CT or cardiac magnetic resonance imaging are often necessary.^{2,3} Abnormal lead or catheter movement during pacemaker implantation or ablation across the interatrial septum should always raise the suspicion of an undiagnosed SV-ASD, even when the transthoracic echo is reported as “normal.”

The SV-ASD accounts for 5–10% of all ASDs and occur equally in both genders.⁴ The SV-ASD is located in the junction of the right atrium and superior vena cava and is usually associated with anomalous pulmonary venous return. (Figure 6) Atrial fibrillation or flutter in patients with ASD typically presents later in life (>40 years)

Figure 4: Transesophageal echocardiogram with post-surgical patch repair.

Figure 5: Transesophageal echocardiogram with post-surgical patch repair with color Doppler.

secondary to atrial dilation and results in sudden worsening of symptoms of shortness of breath on exertion and fatigue, commonly seen in patients with hemodynamically significant ASD.⁵ Classical physical examination findings of the wide and fixed split S2, loud P2, right ventricular lift, and diastolic rumble at the lower left sternal border are not always present. Electrocardiographic findings in SV-ASD include inverted P waves in inferior leads due to the absence, dysfunction of, or surgical trauma to the sinus node (6%), atrial fibrillation or flutter (14%) and presence of various degrees of right bundle branch block, right ventricular hypertrophy, or right axis deviation as a result of right ventricular volume overload (71%).⁶ ASD closure for right atrium or right ventricle enlargement and no evidence of pulmonary arterial hypertension with or without symptoms has been given a class I indication by the most recent ACC/AHA guidelines.⁹ Closure is indicated to prevent long-term complications such as atrial arrhythmias, reduced exercise tolerance, hemodynamically significant tricuspid regurgitation, paradoxical embolism, overt congestive cardiac failure, and pulmonary vascular disease.⁹ Older patients remain at high risk of stroke even after surgical closure of ASD, and as such some authors have advocated continued thromboprophylaxis for older patients with an ASD for 6–12 months after closure.⁴ Pacemaker implantation is often required for patients with SV-ASD and symptomatic bradycardia. In a retrospective cohort study of 202 patients with intracardiac shunts, Khairy et al.⁷ demonstrated that the presence of transvenous leads was independently associated with an increased risk of systemic thromboembolism (HR=2.6) in patients with intracardiac shunts, and this risk increased with older age and the presence of atrial fibrillation and flutter. Interestingly, in the same study, prescription of aspirin or warfarin were not protective against thromboembolism.⁷ Therefore, transvenous pacing should be avoided in patients with unrepaired ASDs.⁸ For older patients with SV-ASD, atrial fibrillation or flutter and symptomatic sinus bradycardia who undergo surgical repair, placement of epicardial pacemaker leads may be advantageous despite their higher capture thresholds and shorter generator longevity.⁷

Figure 6: Schematic drawing showing the location of different types of ASD, the view is into an opened right atrium. HV: right ventricle; VCS: superior vena cava; VCI: inferior vena cava; 1: upper sinus venosus defect; 2: lower sinus venosus defect; 3: secundum defect; 4: defect involving coronary sinus; 5: Primum ASD.

After taking into consideration the extent of the patient's symptoms including shortness of breath on exertion, the need for pacemaker implantation, the risk of paradoxical thromboembolization secondary to pacemaker leads as well as potential surgical risks and the patient's age, a decision was made to proceed with surgical correction of SV-ASD before pacemaker implantation. Management of patients with SV-ASD that needs cardiac pacing requires a comprehensive approach that involves cardiac electrophysiologists, cardiovascular surgeons, and cardiac imaging specialists.

References

1. Therrien J, Warnes C, D'Alto L, et al. Canadian Cardiovascular Society Consensus Conference 2001 update: recommendations for the management of adults with congenital heart disease part III. *Can J Cardiol.* 2001;17(11):1135–1158.
2. Shub C, Dimopoulos IN, Seward JB, et al. Sensitivity of two-dimensional echocardiography in the direct visualization of atrial septal defect utilizing the subcostal approach: Experience with 154 patients. *J Am Coll Cardiol.* 1983;2(1):127–135.
3. Pascoe RD, Oh JK, Warnes CA, Danielson GK, Tajik AJ, Seward JB. Diagnosis of sinus venosus atrial septal defect with transesophageal echocardiography. *Circulation.* 1996;94(5):1049–1055.
4. Webb G and MA Gatzoulis, atrial septal defects in the adult: Recent progress and overview. *Circulation.* 2006;114(15): 1645–1653.
5. Gatzoulis MA, Freeman MA, Siu SC, Webb GD, Harris L. Atrial arrhythmia after surgical closure of atrial septal defects in adults. *N Engl J Med.* 1999;340(11):839–846.
6. Attenhofer Jost CH, Connolly HM, Danielson GK, et al. Sinus venosus atrial septal defect: long-term postoperative outcome for 115 patients. *Circulation.* 2005;112(13):1953–1958.
7. Khairy P, Landzberg MJ, Gatzoulis MA, et al. Transvenous pacing leads and systemic thromboemboli in patients with intracardiac shunts: A multicenter study. *Circulation.* 2006; 113(20):2391–2397.
8. Silversides CK, Marelli A, Beauchesne L, et al. Canadian Cardiovascular Society 2009 Consensus Conference on the management of adults with congenital heart disease: executive summary. *Can J Cardiol.* 2010;26(3):143–150.
9. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 Guidelines for the management of adults with congenital heart disease: Executive Summary. *Circulation.* 2008;118(23): 2395–2451.