

Double Intervention in Single Sitting: Percutaneous Device Closure and Permanent Pacemaker Implantation in a Patient with Atrial Septal Defect

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Abstract

Context: Atrial septal defect may rarely be associated with other cardiac diseases such as arrhythmia, and may require additional intervention. **Case report:** A 16-year-old boy presented with effort dyspnea, tiredness, and fatigue. The electrocardiograph revealed right bundle branch block, atrioventricular block, and left axis deviation. Ostium secundum type of atrial septal defect was detected by transthoracic echocardiography and was confirmed by transesophageal echocardiography. The patient was advised to undergo percutaneous device closure. Permanent pacemaker implantation was also suggested considering the risk of fatal arrhythmias associated with atrioventricular block. Consequently, patient underwent percutaneous atrial septal defect closure and implantation of pacemaker in a single sitting. Both the procedures were successful, after which the patient showed remarkable symptomatic improvement. **Conclusion:** In atrial septal defect patients with unexplained atrioventricular block, closure of atrial septal defect and implantation of pacemaker in single sitting appear to be an attractive modality.

Keywords

Atrial Septal Defect, Ostium Secundum, Pacemaker, Percutaneous Device Closure, Double Intervention in Single Sitting

1. Introduction

Atrial septal defects (ASDs) are one of the most frequently diagnosed cardiovascular malformations in adults and adolescents. It affects over one in 1500 live births and accounts for nearly 10% of congenital heart defects. Ostium secundum is the most common type of atrial septal defect, accounting for about 85% of all ASDs [1]. Genetic analyses have shown that mutations in *NKX2.5* and *GATA4* genes are responsible for familial forms of secundum ASD [1]. Although the defect remains asymptomatic, it may contribute to significant morbidity and premature mortality, if not corrected [2]. It is reported that about 90% of patients with ASD die by the age 60. Hence, patients with atrial septal defect, even those who are asymptomatic, are recommended to undergo the correction of the anomaly [2]. The surgical closure of ASD is considered as the gold standard; however, the percutaneous closure using an occluding device is a safe and effective alternative to surgical closure. Percutaneous closure approach is not only reported to have comparable success rates to surgical closure approach, but also it has the advantages of reduced pain, lesser complications, and shorter hospital stay [3]. In several patients, ASD may be isolated or associated with other cardiac diseases such as valvular stenosis, conduction defects, or atrial arrhythmia, and may require additional intervention [1]. Here, we report a case of asymptomatic ASD in a 16-year-old boy who underwent percutaneous device closure and permanent pacemaker implantation in a single sitting.

2. Case Report

A 16-year-old male patient presented to our clinic with a complaint of shortness of breath, fatigue, and tiredness even after a mild exertion for the past one month. He had infrequent dry cough for the last several months. Palpitation and syncope were evident upon physical examination. The patient was a nonsmoker and had been apparently healthy without any known chronic illness or a history of surgical procedures before admission.

Laboratory investigations revealed that all hematological and biochemical tests were within normal limits. The electrocardiogram (ECG) revealed right bundle branch block, atrioventricular (AV) block, and left axis deviation (**Figure 1**). The transthoracic echocardiograph (TTE) demonstrated normal sized cardiac chambers and physiologically functioning cardiac valves except for a mild leak in the tricuspid valve. Further, an ASD of ostium secundum type was detected (**Figure 2**), which was subsequently confirmed by transesophageal echocardiography (TEE) and was measuring about 20 mm in diameter. The location of the defect with regard to mitral valve and aortic valve were adequate to perform the intervention. We did not observe absence of vital rims and there were no signs of deficient rims or aortic aneurysm. Mild pulmonary artery hypertension was observed and a Qp:Qs ratio of 1.7:1 was measured. Otherwise, the patient was hemodynamically stable. Additionally, we performed ECG in two of his siblings and found no evidences of ASD in both of them.

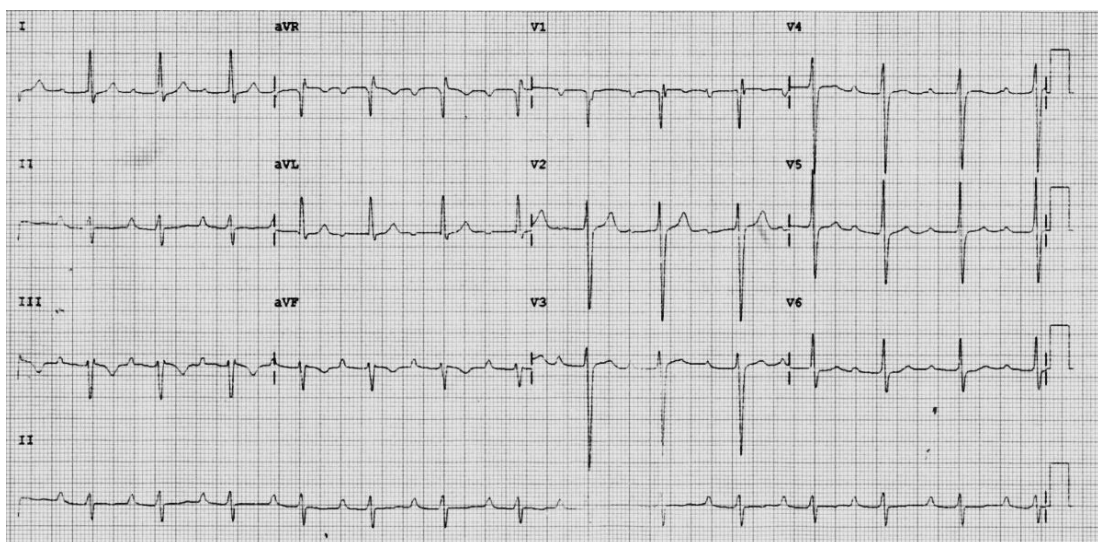


Figure 1. Electrocardiogram showing right bundle branch block, atrioventricular block, and left axis deviation at the time of presentation.

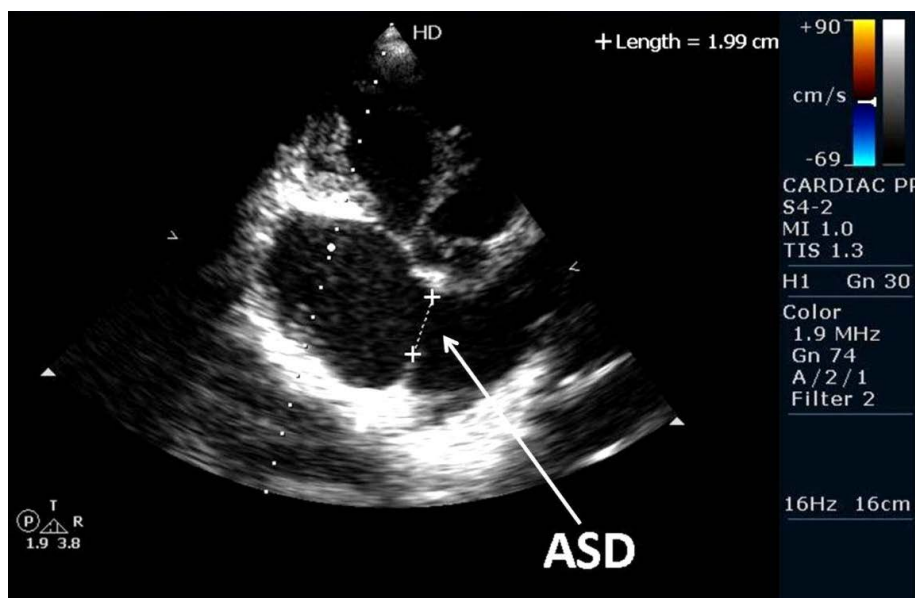


Figure 2. Transesophageal echocardiography confirmed the diagnosis of ostium secundum atrial septal defect.

After consulting patient's family about his medical condition, the patient was scheduled for elective percutaneous device closure of ASD along with a permanent pacemaker implantation in a single sitting. Prior to the procedure, the patient received antibiotic prophylaxis and a bolus of heparin (100 IU/kg) followed by maintenance dose to keep an activated clotting time > 200 seconds throughout the procedure. The interventions were performed under general anesthesia with TTE and fluoroscopy guidance.

For the percutaneous closure of ASD, femoral vein was punctured and the defect was crossed with a Judkins right 3.5 - 5 French (F) catheter using a 0.035 × 260 cm guide wire. After pressure measurement, the catheter was advanced in the left upper pulmonary vein. Successively, the 3.5 - 5 F catheter over the guide wire was replaced with a 14 F long delivery sheath (Cook Medical). A Cocoon ASD occluder device (Vascular Innovations Co. Ltd., Thailand) of 28-mm size was then loaded over the delivery catheter and was introduced in the left atrium. Subsequently, the device was deployed over the ASD (**Figure 3(A)**). The correct device position was verified by both TTE and fluoroscopy, which displayed that the discs were parallel to each other and were separated by the atrial septum (**Figure 3(B)**). Post-procedural TTE confirmed the non-interference of the device with major adjacent structures and valves, and revealed the normal functioning of mitral and tricuspid valve and the absence of residual shunting.

After concluding the ASD closure, percutaneous single-chamber pacemaker implantation was performed via extra thoracic left subclavian vein using the Seldinger technique [4]. Screw-in lead was inserted in the right ventricular apex wall (**Figure 4(A)**) with satisfactory parameters. The lead was affixed and a subcutaneous pocket was formed for the implantation of pacemaker generator. Once the lead was confirmed to be in proper position, the pulse generator was connected to the lead. Finally, the pacemaker pocket was closed in two layers using absorbable subcuticular sutures (**Figure 4(B)**).

Both interventions, percutaneous device closure and pacemaker implantation, were successful. The total fluoroscopy time was 12 min, while the procedural time was 40 min for percutaneous device closure and 35 min for pacemaker implantation. The patient displayed an uneventful anesthesia recovery. Procedure-or device-related thromboembolic and vascular complications, cardiac tamponade, or sustained arrhythmia were not observed. On the next day, a follow-up TTE revealed stable positioning of the device without residual shunt flow. The patient was discharged after satisfactory observations following an administration of aspirin 300 mg/day during hospital stay. After discharge, the patient was advised to take aspirin 150 mg/day for 6 months.

At 1-month follow-up, the patient reported a remarkable decrease in symptoms of fatigue, dyspnea, and palpitations. A considerable decrease in the right ventricle size was also observed without the presence of residual shunt flow.

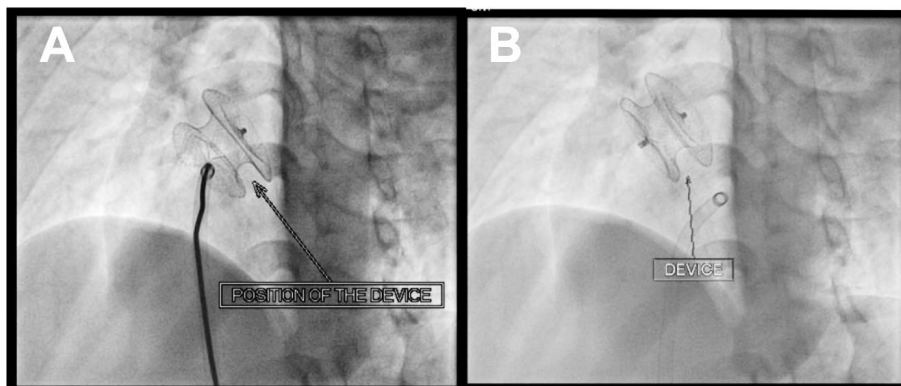


Figure 3. (A) Percutaneous closure of atrial septal defect by cocoon occluder device; (B) Post-procedure verification of the correct device position.

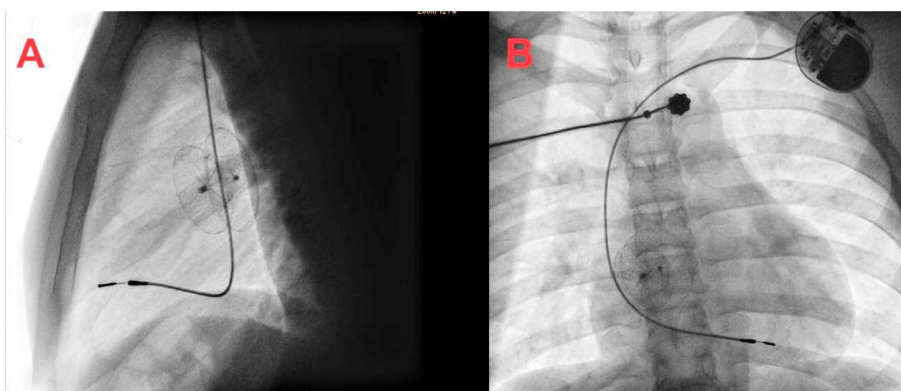


Figure 4. (A) Successful implantation of single-chamber pacemaker in the right ventricular apex wall; (B) Closure of pacemaker pocket in two layers with absorbable subcuticular sutures.

3. Discussion

The atrial septum is one of the most genetically sensitive cardiac structures [1]. In ECG, the significant characteristics of a patient with ostium secundum ASD is right bundle branch block and right axis deviation. Contrary to this, the baseline ECG in our patient exhibited right bundle branch block, AV block, and left axis deviation. While the right bundle branch block suspected the presence of ostium secundum ASD, which was confirmed by TTE, the presence of AV block and left axis deviation were atypical.

It is suggested that all available relatives should also be examined when unexplained left axis deviation or prolonged AV conduction is observed in a patient with a secundum ASD [5]. Earlier, Emanuel *et al.* had proposed a genetic connection to rationalize left axis deviation and prolonged AV conduction in patients with secundum ASD [5]. Mutations in the transcription factors of *NKX2.5* and *GATA4* genes are found to be associated with nonsyndromic ASDs. It is cited that the heterozygous mutations in the *GATA4* genes are responsible for autosomal dominant ASDs and normal conduction while the mutations in *NKX2.5* homeobox genes are responsible for autosomal dominant ASDs with progressive AV block [1]. The genetic link in our patient was not verified as the genetic testing was not done due to financial constraint. However, we investigated the siblings of the patients and found no evidences of familial ASD. Accordingly, we opted for ASD device closure with a percutaneous approach to manage ASD.

Friedman *et al.* had previously reported a case of pediatric patient with ASD and long QT syndrome who underwent percutaneous device closure followed by the implantation of transvenous lead, with an interval of 8 weeks between the procedures [6]. Recently, Bitar *et al.* reported a case in which the ASD closure and transvenous lead implantation were performed during the same hospitalization but not during the same procedure [7]. To the best of our knowledge, this is the first case of elective double cardiac intervention, *i.e.* percutaneous ASD closure and permanent pacemaker implantation, performed simultaneously in a single sitting. The procedures

were successful and the patient showed remarkable symptomatic improvement.

4. Conclusion

The presence of AV block in a patient with ASD should alert the cardiologist against the future risk of fatal arrhythmias, which can be prevented by pacemaker implantation. We conclude that the simultaneous closure of ASD and implantation of pacemaker in single sitting appears to be an attractive modality as compared to surgical or percutaneous treatment in two different sittings.

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