Summary

In Ebstein’s anomaly, percutaneous atrial septal defect (ASD) closure for the treatment of hypoxemia due to a right-to-left interatrial shunt remains controversial. We report the case of a 40-year-old woman with Ebstein’s anomaly who developed cyanosis and shortness of breath on exercise. Her symptoms improved after percutaneous ASD closure and her clinical course has been good during follow-up. The balloon ASD occlusion test, combined with dobutamine stimulation before the procedure, is useful to confirm treatment indication. A prior electrophysiological evaluation is also important because Ebstein’s anomaly is often complicated by atrioventricular recurrent tachycardia.

Key words: ASD occlusion test, Dobutamine stimulation, Electrophysiological study, Interatrial shunt, Cyanosis

CASE REPORT

Percutaneous Atrial Septal Defect Closure in Adult Ebstein’s Anomaly with Exertional Hypoxemia

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Einstein’s anomaly was first reported in 1866 by Wilhelm Ebstein. Clinically, it is a relatively rare congenital heart disease characterized by tricuspid valve dysplasia, usually tricuspid regurgitation, and systolic and diastolic dysfunction of the right ventricle. It results in right-to-left shunting through the interatrial septum, which may lead to cyanosis, syncope, and heart failure. The development of cyanosis usually occurs in adulthood and is associated with right-to-left shunt due to an atrial septal defect. In our patient, percutaneous ASD closure was performed with the balloon ASD occlusion test combined with dobutamine stimulation. The procedure was successful, and the patient’s symptoms improved. This case highlights the importance of patient selection and the use of diagnostic tests in managing Ebstein’s anomaly.

Key points:
- Percutaneous ASD closure is an option for treating hypoxemia due to right-to-left shunt in Ebstein’s anomaly.
- The balloon ASD occlusion test with dobutamine stimulation is useful for confirming the indication for ASD closure.
- Electrophysiological evaluation is important to rule out atrioventricular recurrent tachycardia.

Case Report

A 40-year-old woman was diagnosed with Ebstein’s anomaly at the age of 2 and had been followed-up without intervention from childhood. She gave birth to her first healthy baby at the age of 28 years, with no worsening of heart failure. She developed hypoxemia on exertion at the age of 32, during her second pregnancy; however, the cause could not be identified at this point. At 40 years of age, further examination by contrast-enhanced computed tomography revealed an ostium secundum ASD (Figure 1A), which had not been detected by transthoracic echocardiography. The patient suffered from shortness of breath during exertion, and fatigue, which was defined as New York Heart Association (NYHA) functional class II. A cardiopulmonary exercise test showed desaturation after exercise from 98% to 79% of percutaneous oxygen saturation (SpO2), suggesting that a right-to-left interatrial shunt was the cause.

At 40 years of age, the patient was admitted for evaluating the indication for percutaneous ASD closure. Cardiac examination revealed no fixed splitting of the second heart sound, a Levine II/IV systolic regurgitant heart murmur, no diastolic heart murmur, a blood pressure of 108/60 mmHg, and a heart rate of 80 beats/minute. Routine blood tests showed no anemia (hemoglobin 14.1 g/dL), normal kidney and liver function, brain natriuretic peptide of 56.5 pg/mL (reference range < 18.4 pg/mL), and human atrial natriuretic peptide of 106 pg/mL (reference range < 43 pg/mL). A chest X-ray revealed a cardiomegaly ratio of 0.54. Electrocardiography showed sinus rhythm with strikingly tall and peaked P waves and no delta wave (Figure 1B). Transesophageal echocardiography revealed a left ventricular ejection fraction of 80% and left ventricular end-diastolic diameter of 34 mm (78% of normal).
normal value). Tricuspid septal leaflet annular attachment showed a displacement of 22 mm toward the apex with mild tricuspid valve regurgitation (TR) (TR pressure gradient of 15 mmHg).

The Table shows a summary of cardiac catheterization findings. At rest, the patient showed an SaO$_2$ of 93%, a ratio of pulmonary to systemic blood flow (Qp/Qs) of 0.9, pulmonary artery pressure of 24/8 (14) mmHg, and central venous pressure (CVP) 7 mmHg. Her functional RV end-diastolic volume and RV ejection fraction calculated by biplane cineangiography were 80.8 mL (70% of normal value) and 55.4%, respectively. Transesophageal echocardiography (TEE) showed an ostium secundum ASD (13.3 mm in diameter), a circumferentially large enough atrial rim, bidirectional interatrial shunting via the ASD (Figure 1C), and mild TR (Figure 1D). During an ASD balloon occlusion test using a 24-mm diameter AMPLATZER$^\text{TM}$ Sizing Balloon (St. Jude Medical, St. Paul, MN, USA) (Figure 2A), SpO$_2$ increased from 93% to 97% with no hemodynamic changes, i.e., no elevation of CVP or decrease of systemic blood pressure. To assess the impact of exercise stress, catecholamine stimulation by dobutamine infusion (5-15 μg/kg/minute) was performed. We monitored systemic arterial pressure, CVP, SpO$_2$, and right ventricular cardiac output calculated by TEE during dobutamine stimulation over 40 minutes, revealing an increase of CVP from 7 to 8 mmHg and no exacerbation of TR according to TEE. RV cardiac output increased from 3.7 to 6.7 L/minute, suggesting that systemic cardiac output also increased to the same extent. Although we decided that the ASD could be closed, an electrophysiological study (EPS) revealed atrioventricular recurrent tachycardia (AVRT) with the right Kent bundle in the circuit. AVRT was easily induced by atrial extrastimuli (Figure 2B). Therefore, catheter ablation of the accessory pathway in the tricuspid annulus was performed two months later. The patient was followed-up for seven months without recurrence of tachycardia.

Percutaneous ASD closure was then performed. The ASD occlusion test was repeated, with the same findings. Balloon sizing of the ASD was 15.9 mm, and the patient underwent successful percutaneous ASD closure with a 17-mm diameter AMPLATZER$^\text{TM}$ Septal Occluder (St. Jude Medical, St. Paul, MN, USA). She was discharged without complications. After the procedure, her subjective symptoms improved from NYHA functional class II to I. Her clinical course has been uneventful, with no exacerbation of heart failure during three years’ follow-up.
Table. Summary of Hemodynamics of Cardiac Catheterization

| Balloon occlusion | Dobutamine (μg/kg/minute) | HR (minute) | AoP (mmHg) | CVP (mmHg) | RVCO (L/minute) | SpO2 (%) |
|------------------|--------------------------|------------|--------------|-------------|-----------------|----------|
| -                | -                        | 64         | 83/51        | 7           | 3.7             | 93       |
| +                | -                        | 65         | 105/53       | 7           | 4.2             | 97       |
| +                | 5                        | 63         | 98/51        | 7           | 3.9             | 97       |
| +                | 10                       | 96         | 137/56       | 8           | 6.2             | 97       |
| +                | 15                       | 124        | 128/54       | 8           | 6.7             | 97       |
| -                | 15                       | 127        | 138/58       | 8           | -               | 93       |

ASD balloon occlusion and the dobutamine stimulation test were performed to confirm the treatment indication for ASD closure. After ASD balloon occlusion, SpO2 increased from 93% to 97%, with no elevation of CVP or fall in systemic blood pressure. Dobutamine infusion revealed a slight increase of CVP, from 7 to 8 mmHg. ASD indicates atrial septal defect; SpO2, percutaneous oxygen saturation; CVP, central venous pressure; HR, heart rate; AoP, aortic blood pressure; RVEDP, right ventricular end-diastolic pressure; and RVCO, right ventricular cardiac output.

Discussion

To date, there is no definitive guideline for the treatment of adult patients with Ebstein’s anomaly who have progressive heart failure and hypoxia due to a right-to-left interatrial shunt. Surgical valvuloplasty or replacement of the tricuspid valve with ASD closure are considered to be the standard treatment for these patients. Percutaneous ASD closure for treatment of desaturation in Ebstein’s anomaly is controversial. Extensive examination of hemodynamics by ASD test occlusion, combined with dobutamine stimulation and electrophysiological evaluation, are important to confirm the indication for ASD closure. Accurate assessment of the severity of TR is required to assess whether surgical intervention should be performed to improve right heart function. We evaluated this patient in accordance with this strategy, and treatment was successful with a good subsequent clinical course.

It has been reported that NYHA functional class ≥ III or a cardiothoracic ratio of > 0.6 on chest X-ray increase perioperative mortality, suggesting that early surgical repair may improve long-term outcome. If right ventricular dysfunction is too severe to perform biventricular repair, 1.5 ventricular repair or a Fontan procedure may be performed even in adult patients. However, as in the present case, careful consideration is required to decide on a treatment strategy in patients with hypoxemia due to a right-to-left interatrial shunt and mild to moderate TR because clinical symptoms may improve without surgical intervention for the tricuspid valve. In addition to the degree of TR, functional RV volume is also important. In the present case, although we evaluated functional RV volume and ejection fraction by the measurement of biplane cineangiography, more detailed examination, such as cardiac magnetic resonance imaging, might be useful for assessing RV function.
Conclusions

Percutaneous ASD closure for Ebstein’s anomaly patients with exertional hypoxemia may improve exercise tolerance by eliminating the right-to-left shunt. However, to avoid exacerbation of right heart failure and low cardiac output, careful preoperative assessment of treatment indication should include the degree of TR and right ventricular function. To predict hemodynamic changes after ASD closure, ASD occlusion with dobutamine stimulation are useful. To reduce or eliminate the likelihood of arrhythmias, especially in WPW syndrome, it is important to perform EPS and catheter ablation before ASD closure. Since long-term prognosis after percutaneous ASD closure remains unclear, further follow-up and investigation in a larger series of patients is required.

Disclosure

Conflicts of interest: None.

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