Case Report

Home-made fenestrated amplatz occluder for atrial septal defect and pulmonary arterial hypertension

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Abstract
We report the management of a patient with secundum atrial septal defect (ASD) and severe pulmonary hypertension. A 65-year-old male with recently diagnosed atrial septal defect was referred to our centre for decompensated right heart failure with rest and exercise induced dyspnea and severe pulmonary hypertension. Right heart catheterization confirmed a mean pulmonary pressure of about 55 mmHg and a Qp/Qs of 2.7. An occlusion test with a compliant large balloon demonstrated partial fall of pulmonary arterial pressure. The implantation of a home-made fenestrated Amplatzer ASD Occluder (ASO) was planned in order to decrease left-to-right shunt and promote further decrease of pulmonary arterial pressure in the long-term. Thus, by means of mechanical intracardiac echocardiography study with a 9F 9 MHz UltraIce catheter (Boston Scientific Corp.), we selected a 34 mm ASO for implantation. Four millimeter fenestration was made inflating a 4 mm non-compliant coronary balloon throughout the waist of the ASO, which was successfully implanted under intracardiac echocardiography. After six months, a decrease of pulmonary arterial pressure to 24 mmHg and full compensated right heart failure was observed on transthoracic echocardiography and clinical examination. This case suggests that transcatheter closure with home-made fenestrated ASD in elderly patients with severe pulmonary hypertension is feasible.

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Keywords: atrial septal defect; pulmonary arterial hypertension; interventional therapy

1 Introduction

The repair of secundum atrial septal defect (ASD) with surgical techniques or transcatheter closure is a safe and effective treatment. Previous studies have suggested excellent long-term results with reduction in arrhythmias, pulmonary hypertension, and paradoxical cerebral embolism after ASD closure at young age.[1-2] Over the past years, an increasing number of elderly patients (age > 60 years) have been admitted for transcatheter closure to prevent ongoing congestive heart failure from volume overload. However, transcatheter repair of secundum atrial septal defect is generally contraindicated in patients with associated severe pulmonary hypertension. Recent data highlight the risk of serious acute left ventricular dysfunction leading to pulmonary edema immediately after surgical or transcatheter ASD closure in some patients.[3]

We here present a case of intracardiac echocardiography-aided transcatheter ASD closure in patients with associated severe pulmonary hypertension using a home-made fenestrated Amplatzer ASD Occluder (ASO).

2 Case report

A 65-year-old male with recently diagnosed atrial septal defect, was referred to our center for decompensated right heart failure with rest and exercise induced dyspnea and severe pulmonary hypertension. On transesophageal echocardiography, a mean pulmonary pressure of 55 mmHg was observed as well as dilated right chambers and a moderate left-to-right shunt within the ASD (Qp/Qs 2.5), which the estimated diameter was about 32 mm. The patient was initially submitted to complete right heart catheterization which confirmed a mean pulmonary pressure of about 55 mmHg (systolic 70 mmHg, diastolic 47 mmHg, mean 55 mmHg) and a Qp/Qs of 2.7.

An occlusion test with a compliant AGA medical Balloon demonstrated a decrease of mean pulmonary pressure to 40 mmHg. The implantation of a home-made...
fenestrated ASO was planned in order to decrease left-to-right shunt and promote further decrease of pulmonary arterial pressure in the long run. Thus, a mechanical intracardiac echocardiography study with a 9F 9 MHz UltraIce catheter (Boston Scientific Corp.) was performed as previously described[4-5] and a 34-mm ASO was selected for implantation (Figure1). After having inflated a 4-mm non-compliant coronary balloon throughout the waist of the ASO in order to make a 4-mm fenestration, the ASO was successfully implanted on intracardiac echocardiography. Pre-discharge 24-h trans-thoracic echocardiography revealed decrease of mean pulmonary pressure to 40 mmHg. After six months, further decrease of pulmonary arterial pressure to 24 mmHg and full compensated right heart failure was observed on transthoracic echocardiography and clinical examination.

3 Discussion

In patients < 40 years old with secundum ASD, surgical closure has been demonstrated to be superior to medical treatment in improving both the composite of major cardiovascular events and overall mortality.[6] This superiority seems related to the mean pulmonary artery pressure, age at diagnosis and cardiac index. Survival rates especially favor those patients closed before 25 years (85% vs. 74% at 10 years) but improved quality of life can also be expected for patients closed over the age of 40 years.[7]

Transcatheter repair of secundum atrial septal defect is generally contraindicated in patients with associated severe pulmonary hypertension. In older patients, decompensated right heart failure may develop and is often associated with pulmonary hypertension, as shown in our present patient, which is caused by excessive pulmonary flow over a long period of time.[8] Although Khan et al.[9] demonstrated that transcatheter closure of secundum ASD at advanced age resulted in favorable cardiac remodeling and improvement in functional class, there is controversy over the management of patients with associated severe pulmonary hypertension.

Home-made fenestrated device has been recently suggested in the management of such patient,[10-13] all reporting favorable results in the short term with significant decrease of pulmonary artery pressure. Differently from previous reports in which the fenestration was fixed by means of surgical ligature of the nitinol meshes forming the border of the fenestration, we choose to leave the newly-formed hole free for eventual spontaneous closure by the fibrotic process. Further decrease of pulmonary pressure is expected when the device and the fenestration is closed and completely covered by the fibrotic process. Although this patient was at relatively old age, the positive occlusion test suggested favorable results regarding improvement of congestive heart failure in the near future. In more advanced age and with less positive occlusion test, fixing the fenestration with surgical legation in order to maintain patent the fenestration would probably avoid congestive heart failure over the time.

In conclusion, the case reported here suggests that transcatheter closure using home-made fenestrated ASD in patients with associated severe pulmonary hypertension is clinically feasible even in younger elderly population. However, in patients at more advanced age and with partially positive occlusion test, fixing the fenestration with surgical legation in order to maintain patency of the hole and preventing congestive heart failure may still remain the choice of treatment.

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