Case Report

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TANAFFOS

Curable Syncope in Primary Pulmonary Hypertension with Novel Atrial Flow Regulator

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Pulmonary artery hypertension (PAH) occurs when mean pulmonary artery pressure (PAP) is higher than 25 mmHg in rest or 30 mmHg during activity. Idiopathic pulmonary artery hypertension (IPAH) is defined as PAH without a definite reason. The IPAH is a rare disease with a high mortality rate if left untreated. To date, there has been no definite cure for this entity, and most treatments are for symptom relief or improvement in the quality of life. For many years, decompressing the right heart through a hole in the interatrial septum has been advised to alleviate some of their symptoms, specifically syncope. Atrial flow regulator (AFR) is a device to make an iatrogenic interatrial hole and finally a unidirectional shunt. It has been used for some PAH patients for symptom relief. Herein, we report a 36-year-old female case with diagnosed IPAH for 6 years. In the last 3 years, the case had frequent syncope with shortening intervals. The AFR was implanted for her without any complications. Then, within 6 months of follow-up, she had only one syncope episode. A significant change was observed in her 6-minute walk and PAP.

Key words: Pulmonary artery hypertension; Idiopathic pulmonary artery hypertension; Syncope; Interatrial shunt; Atrial flow regulator

INTRODUCTION

Pulmonary artery hypertension (PAH) occurs when pulmonary artery pressure (PAP) is higher than 25 mmHg at rest or 30 mmHg during exercise. Pulmonary hypertension is classified into five subtypes by the Word Health Organization. Idiopathic pulmonary artery hypertension (IPAH) is in class one (1–3). The PAH does not have a specific sign or symptom (4). To date, there has been no specific treatment for IPAH; therefore, any treatment is for a better quality of life or symptom relief (5–7.)

Patients with atrial septostomy (AS) have shown a better survival rate. In addition, AS can be considered a palliative pretransplant therapy (8, 9). Dizziness and syncope are two devastating clinical symptoms of a pulmonary hypertensive crisis, and their incidence decreases by AS (10). This is a case report of novel atrial flow regulator (AFR) implantation in Iran. It is a device to create an iatrogenic unidirectional right to left interatrial shunt. The AFR is suitable for PAH patients with frequent syncope, and its usage makes them have a better quality of life.

CASE SUMMARIES

A 36-year-old female case with diagnosed IPAH was admitted to implant an Occlutech AFR (Occlutech, Sweden

Company). The patient was initially diagnosed 6 years before administration. Dyspnea during exercise and coughing were her first symptoms of the disease. She had no history of another disease except IPAH. Previously, she had two pregnancies without any complications. She was a non-smoker without any significant family history of pulmonary or rheumatologic diseases. Her drug history included tadalafil, spironolactone, digoxin, furosemide, bosentan, and ilomedin.

Based on the New York Heart Association (NYHA) classification, she was categorized in function class 3. During the last 3 years, she had 16 episodes of syncope crisis, with shortening intervals before admitting to a hospital. Meanwhile, last month before device implantation, she had 8 episodes of traumatic syncope. Before device implantation, her syncope was investigated, and other neurocardiogenic reasons were ruled out. After obtaining the approval of the ethics committee of the National Research Institute of Tuberculosis and Lung Diseases, Shahid Beheshti University of Medical Sciences, Tehran, Iran (IR.SBMU.NRITLD.REC.1397.055) and patient consent, she was scheduled for device implantation.

Technique and Procedure

Device implantation was performed under general anesthesia. Due to severe pulmonary hypertension, routine general anesthesia was high risk in this special population. We recommended a cardiac anesthesiologist who was familiar with pulmonary hypertension hemodynamics to guide during the anesthetic time. Transesophageal echocardiography (TEE) was also used during the procedure to help device positioning and transatrial septostomy. Before the procedure, the patient underwent right-heart catheterization, and chamber pressures and oxygen saturations were analyzed.

Moreover, pulmonary angiography was performed with 50 cc of a contrast agent in a nonselective manner to evaluate pulmonary artery and venous return. Right atrium (RA) pressure, pulmonary capillary wedge pressure (PCWP), right ventricular (RV) pressure, and PAP were reported as 18, 8, 110, and 110/80 mmHg, respectively. Additionally, the mean PAP was 90 mmHg. Oxygen saturation was 42% in the RA, and systemic oxygen saturation was 98%. Cardiac output with the Fick method was 4.8 L/m, and pulmonary vascular resistance was 4 wood units.

Based on two-dimensional TEE guidance, a transseptal puncture was performed with a Brockenbrough needle. A 0.035-inch stiff guidewire was advanced to the pulmonary vein, and a 12-F sheath as the device delivery system was inserted. The AFR device was loaded on the delivery system and advanced into the place. The proper position was checked with fluoroscopy in the left anterior oblique 30° position and double-checked with TEE. The left atrial disc was first deployed, and then the entire system withdrew up to reaching the septum. Finally, the RA disc was released. After confirming the proper position by TEE, the delivery cable was released and withdrawn.

After the procedure, oxygen saturation decreased to 83%. Furthermore, postprocedural PCWP was 10 mmHg, and RV pressure was 90 mmHg. Moreover, RA pressure was 6 mmHg after deployment. The TEE showed a unidirectional right to left shunt. The patient was transferred to the intensive care unit; however, she was hemodynamically stable. Three hours after the procedure, she was awake without any evidence of pulmonary congestion. She was discharged without any complications.

The first month after the surgery, she was syncope-free, with a severe cough. Her coughing was treated with an antireflux drug. Meanwhile, her condition improved one month after the procedure, and ilomedin infusion was stopped. During 6 months of follow-up, the case had only one episode of syncope. Additionally, there was a significant improvement in her 6-minute walk test and PAP after 6 months of follow-up. Her dyspnea decreased, and NYHA classification improved to class 2 function.

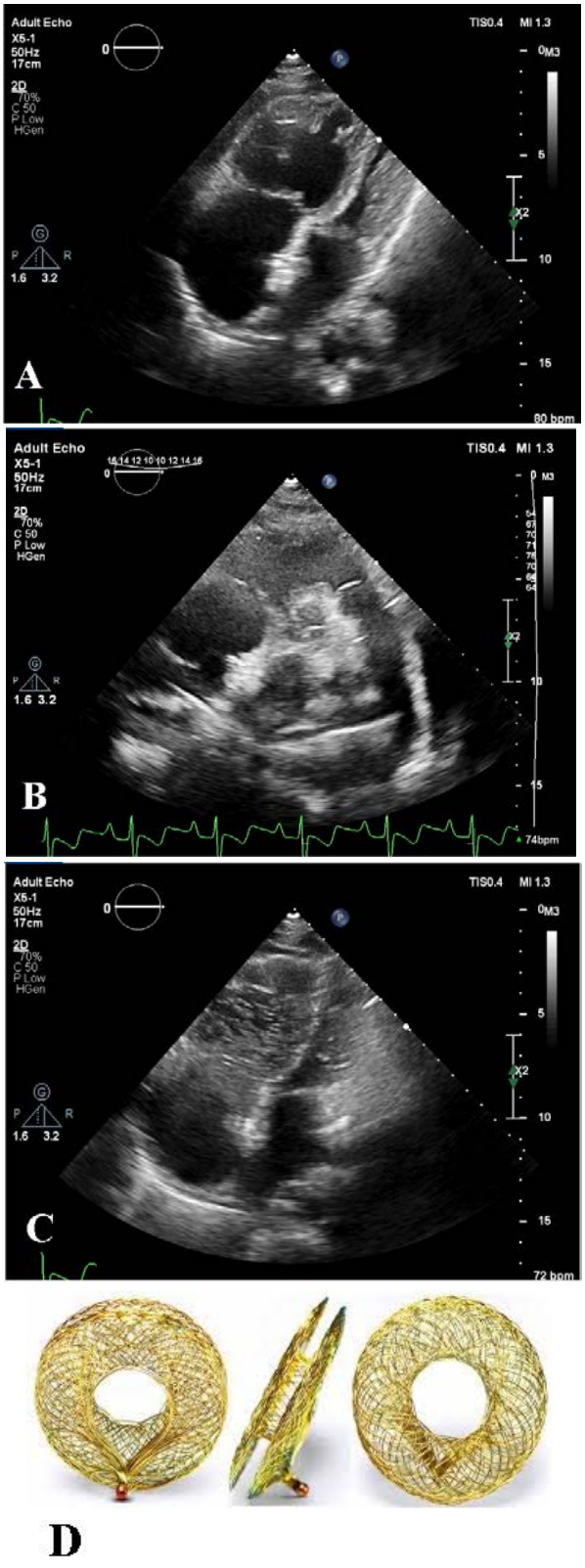

Figure 1. Echocardiography after 6 months follow up with atrial flow regulator (AFR) in inter atrial septum. A) four chamber view, B) short axis view, C) contrast view (arrow shows AFR), D) Occluthech AFR

DISCUSSION

The IPAH is a progressive disease due to pulmonary precapillary vasculature damage. It has an unknown etiology; some genetic or environmental factors might influence the incidence (11). The IPAH is a rare disease with a prevalence of 4 to 6 per million incidences worldwide and nearly 140 deaths per year in the United States of America. Females are more susceptible to the disease (12). It has a nonspecific presentation, and exertional dyspnea is the most common symptom. Lower extremity edema, presyncope, and syncope suggest the advanced stage of the disease (13). The increase in the RV afterload proposes a different pathology for PAH and makes the patient susceptible to syncope (14). There is no definite treatment, and all managements are performed for a better quality of life (5–7). It has a poor prognosis with a mean survival of 2 to 3 years if left untreated.

Besides drug treatment, one of the treatments for an end-stage patient is lung transplantation (15). There is no definite treatment for syncope in PAH patients. Palliative therapy for syncope includes avoiding strenuous activity, physiotherapy, and cardiopulmonary rehabilitation. Furthermore, some prescriptions, such as antipsychotics, should be stopped or titrated (14). Unfortunately, lung transplantation is the only available definite treatment for one-third of patients in a developed country and is not possible in many countries (16).

The AS is the recommended procedure for patients who have limited access to lung transplantation or wait for lung transplantation. By this procedure, they will have fewer symptoms of PAH, especially syncope (17). The AS has its complications, such as early closure or severe hypoxemia, due to a large defect (18). The AFR device makes a predictable and durable unilateral interatrial shunt with an implantable device (19). The AFR can be used for different categories of PAH, especially patients with frequent syncope. It is a double-disc, circular, and self-expanded device, with nitinol wire mesh and unique shape, memory, and elasticity.

A similar study conducted on 12 patients with syncope and PAH showed dramatic improvement. All patients were syncope-free during their follow-up (17). Our case underwent AFR due to frequent syncope. As previously

mentioned, her functional capacity, distance in the six-minute walk test, and PAP improved significantly after device implantation. Novel atrial flow regulator can be curable for syncope in primary pulmonary hypertension patients.

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