Chapter 72
Pulmonary Hypertension (PH) and the Role of Transcatheter Atrial Flow Regulator (AFR) Device Implantation

Ata Firouzi and Zahra Hosseini

Abstract Pulmonary hypertension diagnosis is confirmed with RHC. Advanced cases of PH (mostly, group 1 and 4) who despite of optimized therapy (combination therapy), are being symptomatic (dyspnea functional class III/IV, recurrent syncope and clinical sign of RV failure), and eventually are candidate for lung transplant. In selected cases as bridge therapy, any strategy which can decrease the RV pressure and increase cardiac output, might improve RV function and patients symptoms. Atrial septostomy (which create a right to left shunt for increasing the cardiac output despite of desaturated the patients) has demonstrated to be effective in rare cases. But this procedure is very high risk and has the chance of reocclusion in short time. Recently in some pilot studies, the Occlutech AFR device which is a modified Figulla Flex II ASD Occluder that has been designed for using clinical conditions where creating the permanent inter-atrial communication which would lead to decompression of the right heart side. Further, it would increase cardiac output, blood pressure and organ perfusion and finally reduced the likelihood of syncope and acute right heart decompensation and death.

History

The patient was a 33 years old lady, a case of advanced Idiopathic PH who was referred with recurrent syncope attacks despite optimal medical therapy.

Pulmonary hypertension (PH) is defined as an increase in mean pulmonary arterial pressure (mPAP) of 20 mmHg or greater at rest, as assessed by right heart catheterization (RHC). Pre-capillary PH is defined as mPAP of 20 mmHg or more; a pulmonary capillary wedge pressure (PCWP) of 15 mm Hg or less; and a pulmonary vascular resistance (PVR) of more than 3 Wood units. Pre-capillary PH may

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being in group 1 pulmonary arterial hypertension (PAH), group 3 (PH due to lung disease), group 4 (CTEPH), or group 5 (PH due to unclear or multifactorial mechanisms) in origin. Post-capillary PH is present when the mPAP is 20 mmHg or more and the PCWP is 15 mm Hg or more [1]. Post-capillary PH is most common in group 2 patients or those with PH due to left heart disease. PH has multifactorial pathobiology in which an imbalance in vasoconstriction and vasodilation, thrombosis, and cell proliferation and remodeling of the walls of the pulmonary arteries contribute to increase PVR. Pulmonary vascular remodeling involves the intima, media, and adventitia of small pulmonary arteries (diameter <500 μm); all cell types (endothelial, smooth muscle, and fibroblastic), as well as inflammatory cells and platelets, may play a significant role in the condition. Pulmonary vasoconstriction has been regarded as an early component of the PH process, and excessive vasoconstriction has been related to abnormal function or expression of potassium channels and endothelial dysfunction. Endothelial dysfunction is characterized by impaired production of vasodilators such as a nitric oxide (NO) and prostacyclin, along with overexpression of vasoconstrictors such as endothelin-1. The most common initial symptoms of PH include exertional dyspnea or reduced exercise tolerance, chest pain, fatigue, and light-headedness. Manifestations of more advanced diseases include syncope, abdominal distention, lower extremity edema, and low cardiac output state attributable to right ventricular failure [2].

Diagnostic Work-Up

In physical exam, loud P2, RV heave, systolic murmur of TR, or diastolic murmur of PI and elevated JVP may be found. Central cyanosis, ascites, hepatomegaly, and peripheral edema is seen in advanced cases. In ECG, right axis deviation, RVH, and strain pattern are usually notable. TTE is the first noninvasive tool for evaluation the degree of LV, RV, RA, and LA size and dysfunctions, D-shaped septum, evaluation the causes of PH (left side valvular disease, HFPEF, congenital heart disease), TR and PI severity, estimation of PAP, stretched PFO and finally any pericardial effusion. Perfusion-ventilation lung scintigraphy is an essential tool for rule out of CTEPH. 6 MWT and CPET for evaluation of the functional capacity and finally, RHC is the gold standard modality for confirming the PH, hemodynamic assessment, treatment decision and predicting the prognosis of these patients (Fig. 72.1).

RHC

- So2: 92%
- Mixed venous saturation: 55%
- RVP: 100/20 mm Hg
- mRAP: 18 mm Hg

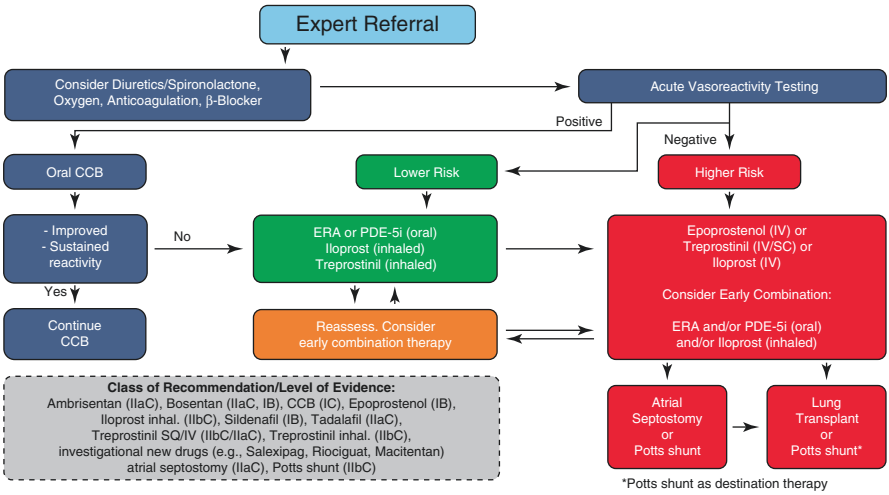

Fig. 72.1 Treatment approach

- mPAP: 70 mm Hg
- PVR: 22 WU
- SVR: 24 WU
- PCWP: 12 mm Hg
- CI: 1.8 L/min/m^2

Interventional Treatment

Balloon Atrial Septostomy (BAS) Atrial septostomy creates a right-to-left interatrial shunt, decreases the right-sided heart filling pressure, improves right ventricular function, and improves left-sided heart filling. Several case series have reported hemodynamic and clinical improvement following this procedure. Although the shunt created decreases systemic arterial oxygen saturation, the goal is an improvement in systemic oxygen delivery based on the improved CO. However, the procedural mortality rate is high, in the range of 9–22%, and it is driven by the severity of PAH and right-sided heart failure in patients undergoing this procedure. The recommended technique is graded balloon dilation of the fossa ovalis, which can be achieved in stages over a period of several weeks in unstable patients. It should not be performed in patients with impending death and severe right ventricular failure. Predictors of procedure-related failure or death include a mean right atrial pressure higher than 20 mm Hg, a PVR index higher than 55 units/m^2, or a predicted 1-year survival rate of less than 40%. Currently, atrial septostomy is recommended for patients with severe PAH and intractable right-sided heart failure despite maximal medical therapy. The goals of this procedure are palliation and restoration and main-

tenance of clinical stability until transplantation can be performed. Atrial septostomy should be performed only by experienced operators in centers with the resources to care for such critically ill patients. Expert-based consensus guidelines define the following as contraindications to atrial septostomy: mean right atrial pressure higher than 20 mm Hg, resting arterial oxygen saturation lower than 90% on room air, or LVEDP higher than 18 mm Hg [3]. The disadvantage of BAS is that the artificial shunt may be too large by inadvertent tear of the BAS, thus causing dangerous desaturation and potential death. Also, it may occlude within the short term due to the overgrowth of intimal cells.

Atrial Flow Regulator (AFR) Device Implantation An AFR device creates communication and allow blood to flow across the interatrial septum when the blood volume under right atrial side is high and the pressure increased. This would decompress the right heart side and reduce the clinically important right heart congestion. Further, it would give the left heart side additional blood volume, thus increasing the left-sided stroke volume, cardiac output, blood pressure, and organ perfusion and finally reduced the likelihood of syncope and acute right heart decompensation and death.

The Occlutech AFR device is a modified Figulla Flex II ASD Occluder that has been designed for using clinical conditions where creating permanent interatrial communication [4] (Fig. 72.2).

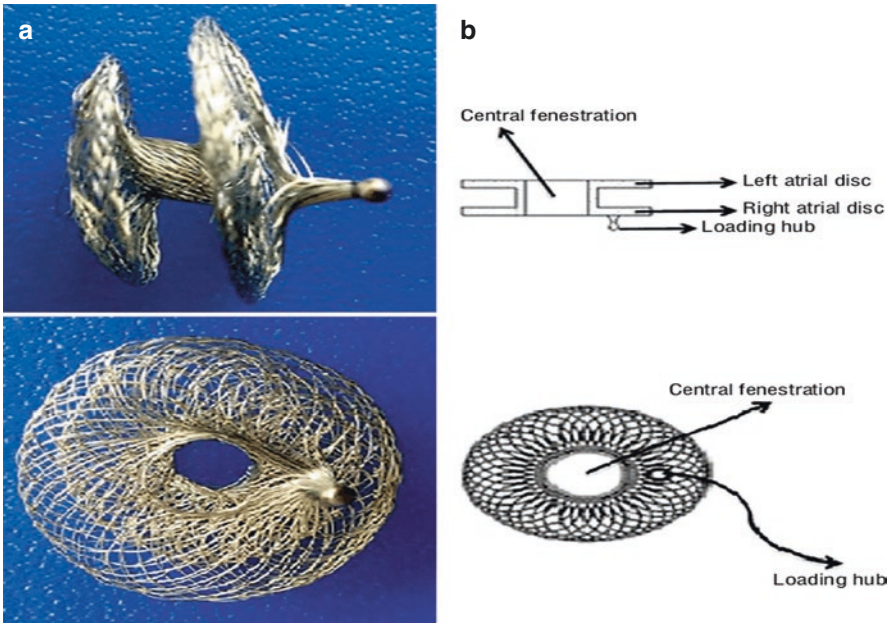

Fig. 72.2 AFR design

Procedural Technique

Under general anesthesia, and closed hemodynamic monitoring by heart failure team (Milrinone and Norepinephrine infusion), after insertion of femoral artery and vein sheaths (6F), under TEE guidance, septostomy was done by Brockenbrough needle with contrast injected to confirm of being in LA (Fig. 72.3) and spiral wire inserted in LA then IV Heparin completed to achieve ACT > 250 s. After septal dilatation by peripheral balloon 10*30 (OTW) (Fig. 72.4), the wire exchanged with Amplatzer Superstiff wire with its tip in LUPV, then the delivery sheath (Occlutech 10F) was crossed through the wire to mid LA and the dilator was removed to allow back bleed. After loading the device (AFR 8 mm) and connecting to the delivery cable, the device was passed over the sheath to reach the tip of the sheath. By retracting the sheath, the distal disk (DD) was deployed in LA then the whole system was pulled backed to the septum, after checking the orientation of the device by TEE, the proximal disk deployed (Fig. 72.5). Finally, after complete assessment of the device position and stability and the degree of right to left shunt, the device was released (Fig. 72.6). Final RHC showed the RAP decreased to 12 mm Hg and SO_2 was 90%. The patient referred to ICU for closed observation and hemodynamic monitoring.

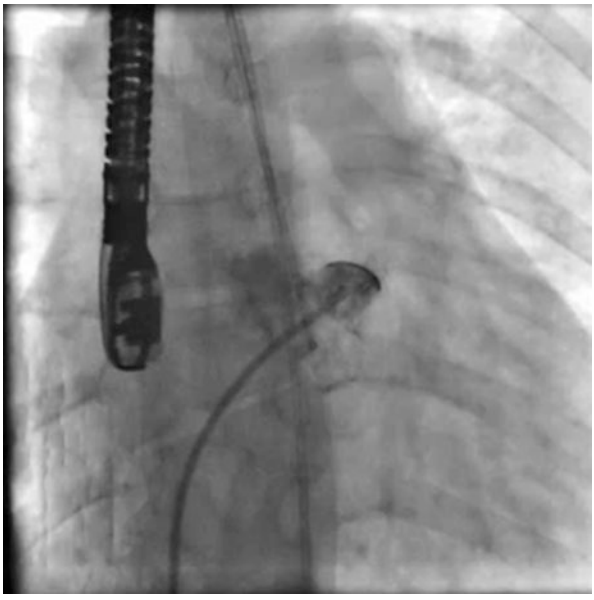

Fig. 72.3 After septostomy, diluted contrast showed the correct procedure

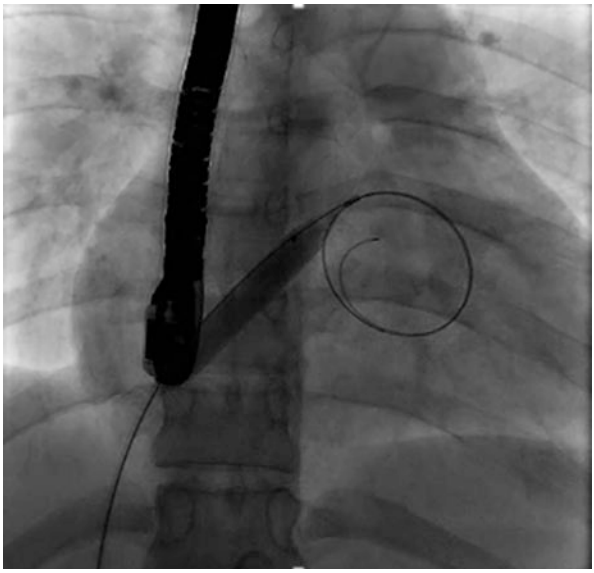

Fig. 72.4 Through the spiral wire, septal dilatation was performed with Powerflex (OTW) 10*30

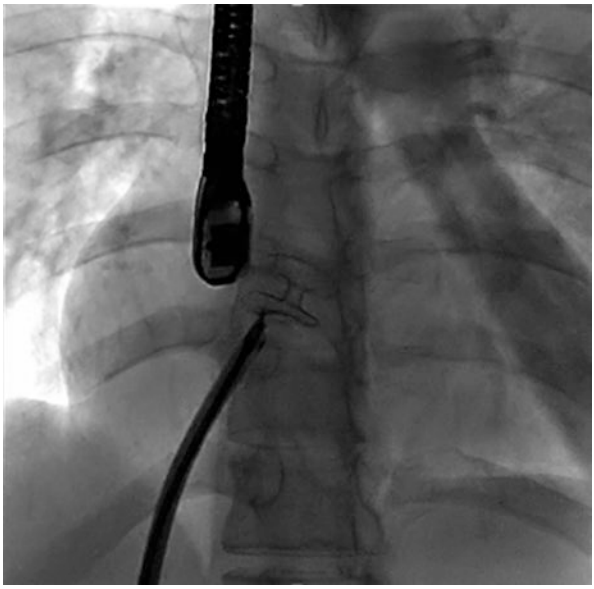

Fig. 72.5 By retracting the sheath, the distal disk (DD) was deployed in LA then the whole system was pulled back toward the septum, after checking the orientation of the device by TEE, the proximal disk deployed

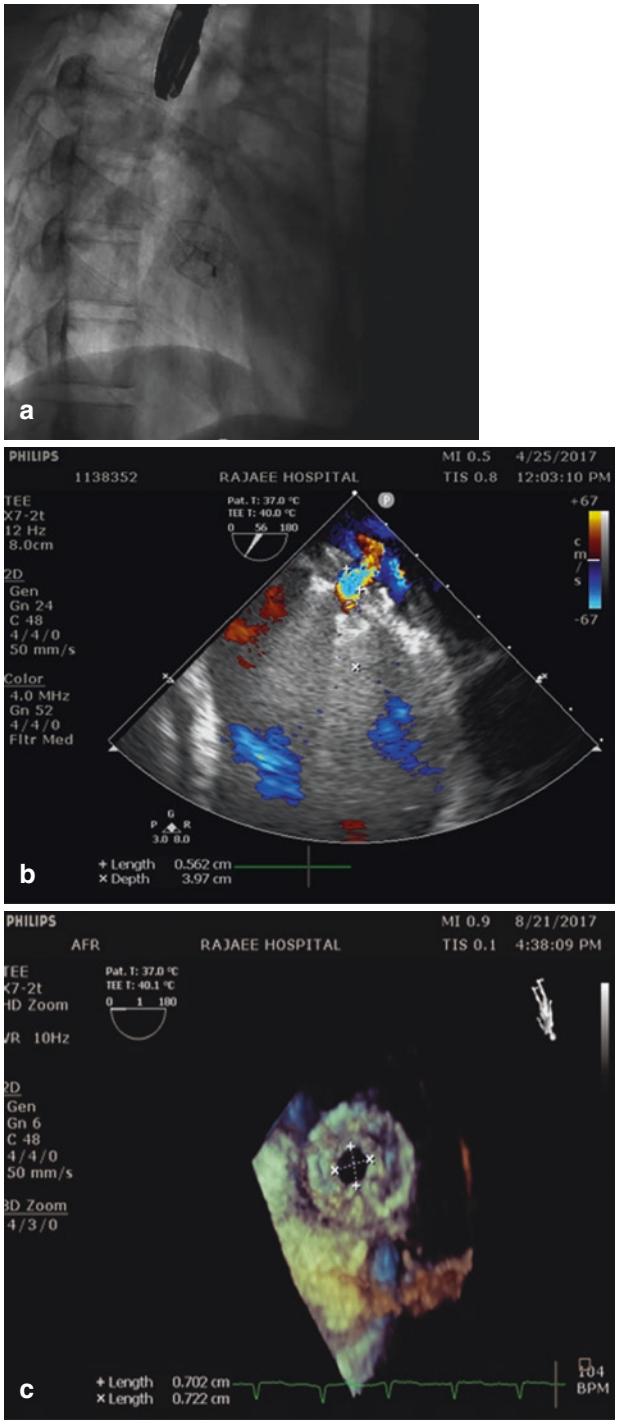

Fig. 72.6 (**a**–**c**) After complete assessment of the device position and stability and the degree of right-to-left shunt, the device was released

Conclusion

Although patients with advanced PH have a very poor prognosis, but any palliative interventions (bridge therapy) to buy the time for them to prepare these patients for lung transplant, is valuable. AFR implantation seems to be a safe procedure that needs more studies for determining the optimal time of intervention and long-term outcomes of these patients [5].

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