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Case Report
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How to Deal with Multiple Shunt Lesions with Eisenmenger Syndrome: A Miracle Response

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Abstract

The intracardiac lesions are most common leading cause of Eisenmenger syndrome especially in developing countries. The lesions including pre-tricuspid or post-tricuspid shunts can result in the most advanced form of Pulmonary Arterial Hypertension (PAH) i.e., Eisenmenger syndrome. There is a considerable risk of death and also compromised the quality of life in patients with Eisenmenger syndrome. In this report, we present a case of 20 years old young girl diagnosed with Atrial Septal Defect (ASD) and Patent Ductus Arteriosus (PDA) with Eisenmenger syndrome. She was managed with transcatheter device closure of PDA and was placed on pulmonary vasodilators so addressing high pressure lesions first and keeping low pressure (safety valve) lesion later on as stepwise approach. Her quality of life improved enormously within 3 months of device closure of PDA.

Keywords: Atrial septal defect; Patent ductus arteriosus; Bidirectional shunt; Eisenmenger syndrome

Introduction

A single or multiple intracardiac shunt lesions or complete mixing, single ventricle or complex combinations can result in progressive increase in pulmonary arterial pressures and finally leading to a deadly complication Eisenmenger syndrome [1-3].

Pulmonary Arterial Hypertension (PAH) is common in patients with CHD and should always be excluded in all patients presenting with PAH. Careful, simple measurement of oxygen saturations in fingers and toes at rest and after exercise can aid in diagnosis of occult CHD [4]. Eisenmenger syndrome is highest in patients with complete atrioventricular septal defects or primum atrial septal defects, followed by Ventricular Septal Defect (VSD), Patent Ductus Arteriosus (PDA) and atrial septal defects and others [5].

Several case reports exist to manage such patients with single or solitary shunt lesion with Eisenmenger syndrome. Still, there is no consensus about the management strategy of patients with multiple shunts lesions with Eisenmenger syndrome [6,7].

The multiple shunt lesions like patients with ASD and VSD, VSD and PDA, ASD with PDA, ASD with Aortopulmonary Window Defect (APW) or multiple VSDs or any combinations thereof as our patient has ASD with PDA. She responded extremely extraordinary to eliminating the high-pressure shunt to lungs along with targeted PAH medications. We believe that in multiple shunt lesions, one can reduce the shear effect on pulmonary vasculature by abolition of high-pressure shunt lesion (step I) as Eisenmenger physiology is an ongoing process. The low-pressure lesion like Secundum ASD can closed later on (step II) by either way of fenestrated ASD device or patch.

Case Presentation

A 20 years old young girl presented with history of shortness of breath NYHA class III and become cyanosed after 50 m of walk. She was afebrile, normotensive, pulse rate of 95 bpm and respiratory rate was 30/m. Her fingers and toes were dusky in color; this discoloration was more prominent in toes with room air saturation 90% (UL) and 85% (LL).

2-D Echocardiography showed large secundum ASD with bidirectional more left to right shunt and large PDA (Figure 1 and Video 1) with bidirectional shunt more right to left shunt(diastolic left to right shunt). The right ventricle was dilated with RVSP 110 mmHg and pulmonary regurgitation jet velocity was 3.3 m/s with normal biventricular functions. She was oral sildenafil with inappropriate dosage. We had detailed discussion with the patient and family about the nature of disease and its

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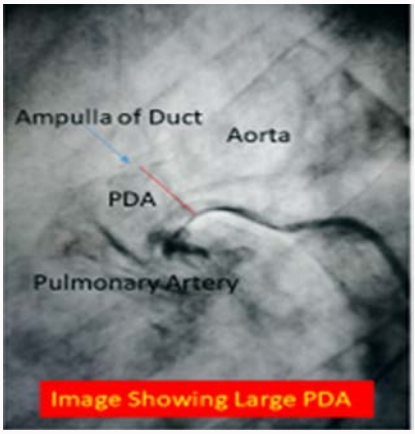
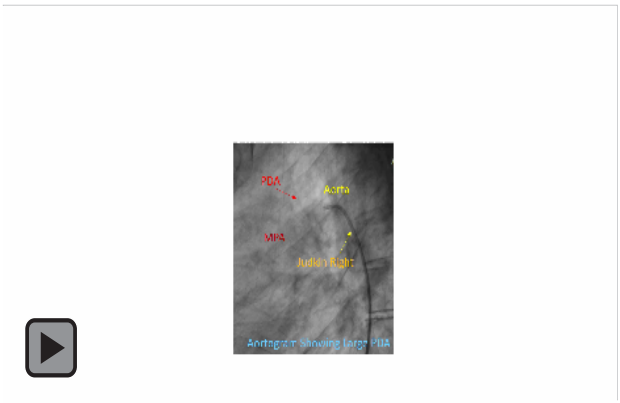
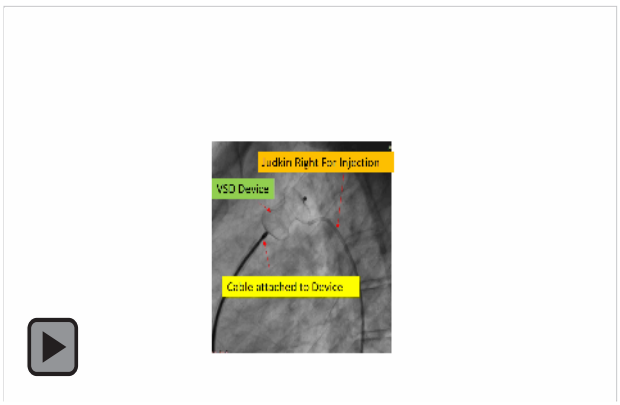


Figure 1: 2-D Echocardiography showed large secundum ASD with bidirectional more left to right shunt and large PDA.



Video A:



Video AP:

progression. She was planned for right heart catheterization and if test occlusion of duct was successful then device closure of duct. Her lab Markers were Hb 16.8 g/dl, TLC 9.5 × 10^3/ul, Platelets 179 × 10^3/ul, urea 39 mg/dl creatinine 0.98 mmol/L, LFTs-NAD, CXR-NAD, COVID-19-Negative.

An informed written consent was taken and the procedure was done under general anesthesia. Right heart cardiac catheterization was performed PAP was suprasystemic 124/74/97 mmHg and AoP 116/73/90 mmHg, Qp:Qs. 9:1 with PVR 24 wood u.m^2. A 20 mm double disc VSD device was loaded and deployed across duct (Video 2 and Figure 2), there was immediate drop of PAP to 86/59/69 mmHg and AoP 116/84/91 and mPAP from 97 mmHg to 69 mmHg. The device was not released and she was monitored for 30 m and her pressures remained stable, then the device was unscrewed (Video 3). She was shifted to ward and was discharged home next day on PAH-targeted medications along with home oxygen therapy.

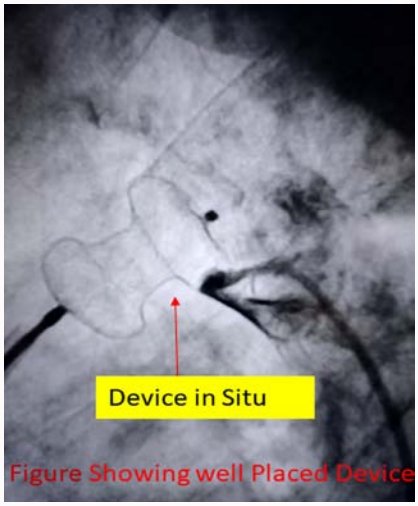


Figure 2: A 20 mm double disc VSD device was loaded and deployed across duct.

Video APC:

Discussion

Patients with multiple shunt lesions with Eisenmenger syndrome are challenging cases to manage, given our patient had ASD with PDA. After 12 weeks of Bosentan therapy, patients with single shunt lesion with Eisenmenger syndrome had successful repair [7,8]. There are publications in the literature on successful surgical treatment of patients with Eisenmenger syndrome [9-13].

Recent researches show the possibility of remodeling lung vessels when taking PAH-specific therapy and there is no single tactic for managing such patients. The evolution of pulmonary vascular disease is more aggressive in patients with high flow and pressure lesions like VSD and PDA as compared to low pressure lesion like ASD. This is because the high blood flow and pressure lesions may induce more pulmonary vascular endothelial damage and induce early smooth muscle cell hypertrophy, proliferation and adherence and activation of platelets and leukocytes.

Despite having high pulmonary vascular resistance, our patient's mean PAP dropped 30% immediately and responded well to PAH-targeted medications. Moreover, it has been suggested that endothelin receptor antagonists may have anti-proliferative effects causing reverse remodeling in pulmonary circulation [14]. It has been suggested that without correction of underlying high-pressure defects like large VSDs, PDA and APW, the PAH therapies leads to an increase in flow and shear stress in pulmonary circulation [15]. In addition, there is no consensus on whether intracardiac communication should be completely eliminated or whether fenestration should be left on the patch or double flap technique, acting as a shunt.

Maintaining fenestration on patch, which subsequently acts as a valve, was proposed by Charles P. Bailey [16]. With continued improvements in the diagnosis, preoperative management, refinement of surgical techniques and intra and postoperative management strategies, patients with Eisenmenger syndrome using a diagnostic treat and repair strategy are operable with safety and efficacy in the current era with advanced pulmonary arterial hypertension therapy.

Zhenlei Hu et al. [17] operated 41 adult patients with excellent results following treat- and repair strategy with overall mortality 4.9%. All patients had bidirectional shunt or even right-left shunt compared to our patient who was having two shunt lesions with ASD mainly left-right and at PDA mainly right-left shunt. Now after the device closure of PDA, our patient is maintaining oxygen saturations 95% on room air. There will be a possibility of closing the ASD with fenestrated device or patch after completion of one-year PAH-targeted therapy after establishing the criteria for closure at that time.

Result

The mean Pulmonary Artery Pressure (mPAP) was significantly decreased (30%) immediately after test occlusion of duct. She improved WHO Functional Class (WHO-FC) from class III to I, Six-Minute Walking Distance (6MWD) from 50 m to 550 m, RVSP reduced from 110 mmHg to 55 mmHg and room air saturation in fingers and toes improved from 89% to 95% after 3 months of PDA closure and PAH-targeted drugs.

Conclusion

Eisenmenger Syndrome with congenital heart defects is difficult to manage, however in multiple shunt lesions complete elimination of large high-pressure shunt (step I) and keeping low pressure lesion (step II) as stepwise approach with PAH-targeted drugs play a crucial role in clinical outcome.

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