Ibrahim Card Med J 2013; 3(1 & 2): 53-58

Severe Pulmonary Valvular Stenosis and Balloon Pulmonary Valvuloplasty (BPV): A Case Report

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ABSTRACT

Congenital pulmonary valve stenosis is a common congenital heart disease andIsolated pulmonary valve stenosis comprises 8-10% of all congenital heart disease. It is an acyanotic heart disease,but can present with severe cyanosis if it is associated with patent foramen ovale (PFO). Severe pulmonary stenosis with cyanosis can be misdiagnosed clinically. Proper evaluation and modern technique of treatment modality can save a life easily. We report a 5 years old boy with severe pulmonary valvular stenosis with PFO who was clinically misdiagnosed as a case of congenital cyanotic heart disease (Tetralogy of Fallot). After taking proper history, clinical examination and investigations we treated the baby by balloon pulmonary valvuloplasty (BPV) successfully without any complication. The short term (6 months) outcome of BPV showed excellent result.

Key Words: Severe pulmonary valvular stenosis, Balloon pulmonary valvuloplasty.

INTRODUCTION

Congenital pulmonary valve stenosis (PVS) is a common congenital heart disease. Isolated pulmonary valve stenosis comprises 8-10 % of all congenital heart disease.[1] Pulmonary stenosis with or without other associated lesions, occurs in 25-30% of all patients with congenital heart disease.[2] Recognizing the potential advantages of a less invasive approach, the first attempts at percutaneous catheter-based dilation of stenotic pulmonary valves were performed in 1950s.[3,4] But the traditional method of treatment for this obstructive lesion was surgical valvotomy up to 1982. That time Kan[5] first introduced the technique of percutaneous balloon pulmonary valvuloplasty (BPV) successfully. Stable positioning of a balloon within the pulmonary valve allows the radial forces of the balloon to separate the fused commissure during inflation.

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As the technique was refined and catheter and balloon technology have advanced, the results of balloon pulmonary valvuloplasty have improved and the approach has now become the standard of care for treating pulmonary valve stenosis.[6] This approach has reduced the need of using cardiopulmonary bypass, and post-surgical ICU admission with multiday hospitalization. The short[7-8] and mid term[8-11] results of BPV have been so good that now a days it has become the preferred method of therapy for moderate to severe PVS both in children as well as in adults. It is also safe and effective for relief of PVS in neonates.[5] However this safe procedure has mortality rate of < 0.25% and very few major complications.[12] The purpose of the article is to describe the techniques and current technology of balloon pulmonary valvuloplasty which can be successfully done without any significant post procedure complications.

CASE REPORT

A 5 years old boy weighing 15 kg came to Ibrahim Cardiac Hospital & Research Institute (ICHRI) with the complaints of bluish discoloration of whole body, exertional dyspnea and irritability for last 4 years. For last 5-6 months he had been suffering from chest pain. The boy was taken to a peripheral health complex where he was misdiagnosed as TOF and had been treated by beta blocker for last 5 months. A complete evaluation was done at our hospital and diagnosed as severe pulmonary valvular stenosis. Clinically the boy was severely cyanosed, irritable with heart rate (HR) 140/m, respiratory rate (RR) 30/m, SaO_2 85% at room air, blood pressure (BP) 110/70mmHg. His Jugular venous pulse wave was seen and palpable. Apex beat was at left 5th intercostal space (ICS) just lateral to mid clavicular line. S1 was normal followed by ejection click and S2 was soft, ejection systolic murmur was 4/6. Lungs were clear, liver enlarged 2 cm from right costal margin, level of haemoglobin 20.2 gm/dL, haematocrit (Hct) 69%, PT control, 14 second and PT patient 21 second. Other haemodynamic data were normal. Baby was given fresh frozen plasma with intravenous frusemide and partial exchange transfusion was done as a preparation of Balloon Pulmonary Valvuloplasty (BPV). Later on the patient's Hct was 50%, PT control 14 sec, PT patient 16 sec. X-ray showed cardiomegaly with right atrial enlargement, dilated convex right ventricle occupying the apex, prominent main pulmonary artery (MPA) segment with oligemic lung field. ECG revealed QRS axis 120, right axis deviation, R in V1, V2, V3 was 22 mm (normal 8-15mm) and R:S ratio in V6 is 0.5:1 (normal 2.5).

Color Doppler Echocardiography showed Situs solitus, d-loop ventricle, normally related great vessels (S,D,S), doming and mildly thickened pulmonary valve with annulus 12 mm (Fig-1), severe pulmonary valvular stenosis (effective opening of pulmonary valve 1mm) gradient 114 mmHg (Fig-2), no infundibular

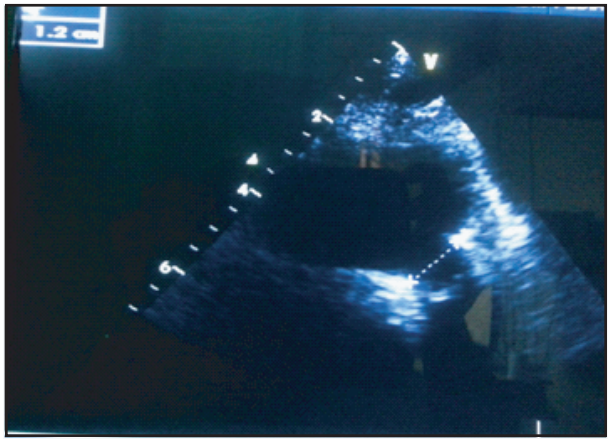

FIGURE 1 : *Doming pulmonary valve with pin head effective opening. Pulmonary valve annulus 12 mm*

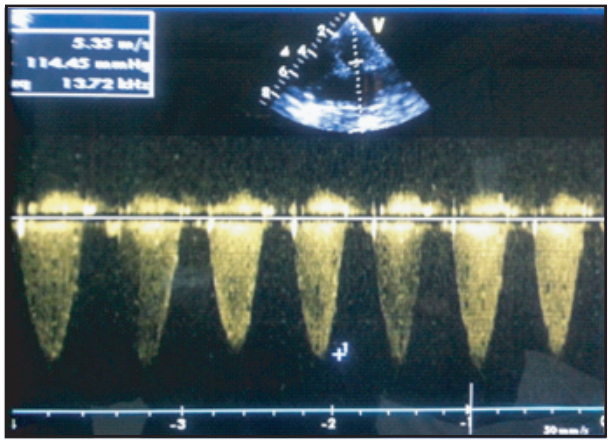

FIGURE 2 : *Severe Pulmonary valvular stenosis*

CASE REPORT

obstruction, post stenotic dilatation of main pulmonary artery with confluent branch pulmonary arteries. Right atrium was enlarged, RV hypertrophied with severe TR (though tricuspid valve anatomy was good), 3 mm pataent foramen ovale (PFO), right to left shunt (R-L), good biventricular function. Inferior vena cava (IVC) dilated and non-collapsing.

Right heart study before BPV

Premedication with a sedative and analgesic was given to the baby, followed by local anesthesia. Continuous ECG monitoring and blood pressure measurement was carried out and arterial oxygen saturation evaluated by means of pulse oxymeter. The baby was electively ventilated. Before starting catheterization all drugs and equipment for resuscitation were prepared. Right femoral vein and femoral artery were accessed percutaneously. After heparization and antibiotic prophylaxis the initial hemodynamic assessment was done. The right ventricular systolic pressure was 144 mmHg. The right ventriculogram was done with 5F Berman catheter which confirmed the presence of isolated pulmonary valvular stenosis with systolic doming of pulmonary valve.

A narrow jet of contrast was seen crossing the pulmonary valve with post stenotic dilatation of main pulmonary artery. Pulmonary valve annulus was 12 mm, right ventricle was hypertrophied and trabecullated and hypertensive. Infundibulum was hypertrophied without any obstruction. Biventricular function was good.

Balloon Pulmonary Valvuloplasty

It was difficult to enter into RV because of severe TR. We used 5F JR to enter into RV. Same catheter was used for the entrance intopulmonary artery with the help of straight tip (0.25 inches)terumo wire which was exchanged by 0.35 inches exchange guide wire. Zedmed Balloon (14mm) was used for dilation of pulmonary valve. Pressure was given into balloon upto full dilation of balloon (Fig-4). Proper waist formation confirmed the successful procedure (Fig-3). Post BPV pull through pressure from pulmonary artery to RV showed decreased pulmonary valve gradient from 114 to only 15 mmHg (Table 1).

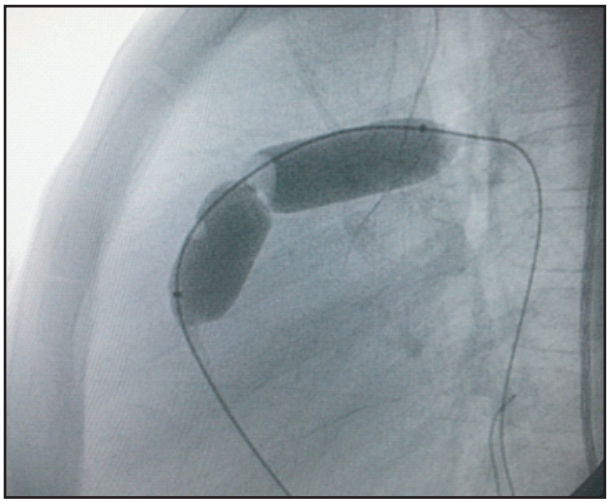

FIGURE 3 : *Waist formation during balloon dilation*

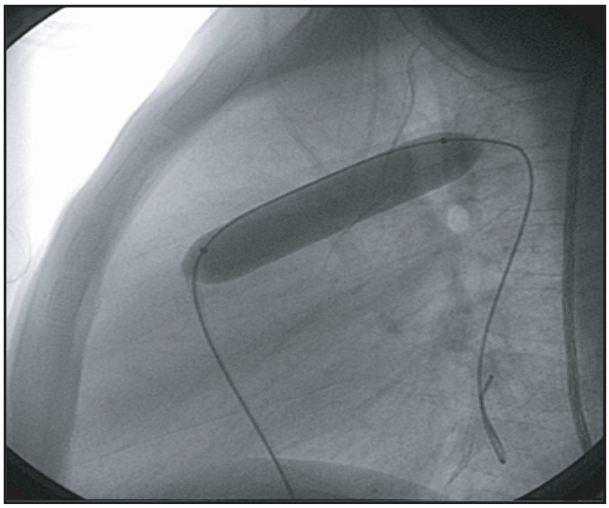

FIGURE 4 : *Full dilation of balloon indicating a successful BPV*

TABLE I. Pre & post procedure pressure gradient

Site	Pre BPV Pressure	Post BPV Pressure
Right ventricle	144 mmHg	40 mmHg
Pulmonary Arteries	30 mmHg	25mmHg
Pressure gradient	114 mmHg	15 mmHg

Post Balloon Pulmonary valvuloplasty echocardiography showed free flow of blood through the pulmonary valve with minimal pulmonary valve gradient and trivial pulmonary regurgitation. RA and RV were mildly dilated & mildly hypertensive with good biventricular functions. Clinically the baby was acyanotic with 99.9% saturation in room air.

Post Catheterization care:

Patient was transferred to Intensive care unit where stabilization was carried out with recording of vital signs and pedal pulses. Patient received intravenous fluid and broad spectrum antibiotic for 2 consecutive days. He was discharged in a stable clinical andhaemodynamic condition day after the procedure.

Follow up:

Follow up after 3 months included clinical evaluation and echocardiographic studies. Clinical evaluation focused on symptoms while echocardiography studies assessed the maximum peak instantaneous systolic pressure gradient across the pulmonary valve and the presence of pulmonary regurgitation. On follow up after 3 months the baby was clinically well with pulmonary valve gradient 10 mmHg.

DISCUSSION

In patients with relatively normal cardiac output, classification of severity of pulmonary stenosis is routinely based on measurement of right ventricular pressure and valve gradient which determine the necessity of percutaneous balloon pulmonary vavuloplasty. Mild stenosis is charachterized by pulmonary valve gradient <35 to 40 mm Hg. In moderate stenosis valve gradient is 40-60 mmHg and insevere stenosis it is >60-70 mm Hg.

For the best management, pulmonary valvuloplasty is currently the first line of treatment for pulmonary valve stenosis at any age and most would agree, for any valve morphology. Valvuloplasty should be performed in any symptomatic patient as soon as the diagnosis is made. Even asymptomatic patients with severe obstruction should be treated semi-electively with valvuloplasty shortly after diagnosis. Patients with moderate obstruction should undergo elective valvuloplasty. No intervention is necessary for patients with mild obstruction.[2]

Our study confirmed earlier reports that balloon pulmonary valvuloplasty may provide effective relief of pulmonary valve stenosis. The patient aged 5 years had typical doming pulmonary valve with severe pulmonary valvular stenosis. This stenosis is due to thickened leaflet with fused commissures with suprasystemic right ventricular pressure. But the present case had no infundibular stenosis.

In a previous study Sullivan et al[13] with 23 infants and children aged 7 days to 12 years estimated pulmonary valve diameter by cross sectional echocardiography to assist in the choice of balloon size. The external balloon diameter that was equal to or slightly less than the measured pulmonary valve ring diameter was selected for the first 14 procedures. Balloon diameters about 20% greater than the pulmonary valve ring diameter were used subsequently except in one patient, in whom the largest balloon diameter available was 20 mm (5 mm less than the measured pulmonary valve ring diameter). Similarly we estimated our patient's pulmonary valve annulus diameter by cross sectional echocardiography which is considered gold standard. During procedure we used 20% larger balloon diameter than the pulmonary valve annulus diameter which did not damage the right ventricular outflow tract or did not cause any pulmonary incompetence. The study of Sullivan et al[13] showed significant reductions in the ratio of right ventricular to systemic systolic pressure and pulmonary systolic pressure gradient immediately after balloon dilatation. Before dilatation 10 patients had suprasystemic right ventricular pressure. Twelve patients underwent

recatheterization (11 at six months and one at one week after balloon dilatation) which showed further improvement with significant reduction in right ventricular pressure or pulmonary valve gradient or both, particularly in the patient with the least satisfactory initial results. This improvement was attributed to resolution of the obstruction at infundibular level.[13] Our patients had no infundibular stenosis and post procedure RV graphy showed good flow to the pulmonary field.

Although BPV is a safe procedure Mohanty et al[14] described a case of pulmonary valvular stenosis which developed acute severe refractory pulmonary edema immediately after balloon pulmonary valvotomy. Similarly Walker et al[15] reported pulmonary edema after BPV in an elderly patient, which resolved with diuretics, ventilation and minimal ionotropic support. This was presumed to be due to a sudden increase in blood flow after BPV in severe pulmonary stenosis. However, two other cases reported by Shrivastava et al[16] were in young children without any associated left heart pathology. Their patients had significant tricuspid regurgitation. They reasoned that the presence of long standing significant tricuspid regurgitation led to a state of chronic hypoperfusion of lungs and sudden opening of the pulmonary valve precipitated the pulmonary edema. Similarly in our study the patient was untreated for long time and had hypertensive RV with severe pulmonary valvular stenosis with severe tricuspid regurgitation. Post procedure the baby was in ICU under close observation. Just after procedure his saturation was 100% with 2 liter oxygen. He was on intravenous Frusemide only for 6 hours and after that diuretic and afterload reducing agent Enalapril was given orally. Post procedure color Doppler echocardiography revealed good flow through pulmonary valve with minimal pulmonary incompetence and mild gradient between RV and pulmonary blood flow. The baby did not develop any pulmonary edema or any other complications.

A low immediate post-BPV gradient was found to be the most important predictor of successful pulmonary valvuloplasty in infants with severe valvular pulmonary stenosis. This is in agreement with McCrindle et al[17] who reported that a higher gradient immediately after the procedure predicts suboptimal outcome.

Similar observations were made by Gupta et al[18] who reported high post BPV gradient (>25 mmHg) as a good predictor of a suboptimal long term result. Similarly, Castillo et al[19] identified both post BPV higher right ventricular Systolic Pressure (RSVP) and dysplastic pulmonary valve as significant risk factors for procedure failure. Our observation is also same that low immediate post-BPV gradient is the most important predictor of successful pulmonary valvuloplasty.

Thus, the above findings highlight the fact that proper history taking, clinical examination and investigation can pick up the simple cases of PVS earlier, a prerequisite for safe and successful procedure which would ensure a good outcome with less complication.

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