Balloon pulmonary valvotomy and anesthesia implications

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Isolated pulmonary valve stenosis (PS) constitutes 7.5–9% of all congenital heart diseases. Percutaneous balloon pulmonary valvotomy (BPV) is the treatment of choice for the isolated PS. BPV is preferred in moderate-to-severe pulmonary stenosis and typical dome-shaped valvular PS. BPV is also the preferred treatment in neonates with critical pulmonary stenosis. BPV success rate is lesser with dysplastic valves. As a part of the team, an anesthesiologist not only helps in anesthesia management of the procedure but prompt management of the complications, especially during manipulation across RVOT, balloon inflation, and postoperative course.

Keywords:
balloon pulmonary valvotomy, Noonan syndrome, pulmonary edema, pulmonary valve stenosis, suicidal right ventricle

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Introduction

Isolated pulmonary valve stenosis (PS) constitutes 7.5–9% of all congenital heart diseases [1]. Percutaneous balloon pulmonary valvotomy (BPV) is the treatment of choice for the isolated PS. BPV has replaced surgery for this condition. It is a safe and effective procedure with very few complications [2].

We report two cases of different age groups who underwent BPV.

Case report

Case 1

A 16-year-old female married for 1 year was admitted with incomplete abortion. She gave a history of effort intolerance and bluish discoloration of skin since childhood. She also had complaints of dyspnea on exertion, which was exacerbated since 2 days. She was a known case of congenital heart disease, but details were not available with the patient. After evaluation, she got diagnosed with valvular PS.

On examination, pallor, cyanosis, and clubbing were present. Pulse rate was 70 bpm, blood pressure 92/60 mmHg. Ejection systolic murmur heard at pulmonary area, thrill present. SpO_2 was 70% on room air.

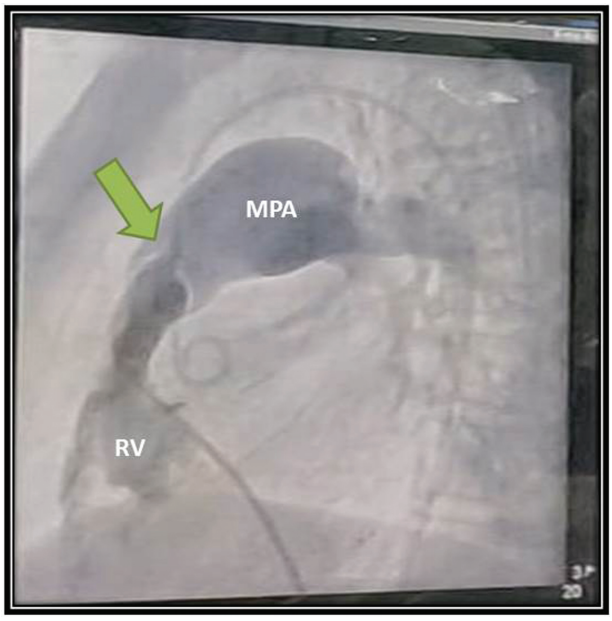
Two-dimensional echo findings were ostium secundum – atrial septal defect of 13 mm with predominant right-to-left shunt, pulmonary valve (PV) doming leaflet, severe valvular PS with gradient of 71 mmHg, PV annulus – 16 mm, main pulmonary artery – 21, TAPSE 14, and RV dysfunction present. Her blood investigations were within normal limits. The patient was then posted for BPV.

Monitored anesthesia care was decided as plan of anesthesia. Standard monitors were attached. Oxygen was started via nasal prongs at 4 l/min. Dexmedetomidine infusion was started at 0.5 μg/kg/h.

Right ventricular systolic pressure was 70 mmHg with an end-diastolic pressure of 12 mmHg. The left ventricular systolic pressure was 105 mmHg with an end-diastolic pressure of 10 mmHg. PV stenosis and post-stenotic dilatation of main pulmonary artery were noted on the right ventriculogram. After some struggle to pass guidewire across the stenosis, balloon dilatation was done (Fig. 1). An instant fall in saturation upto 60% and fall in BP upto 66/46 mmHg occurred. Both returned to the baseline on balloon deflation.

Postprocedure, the right ventriculogram revealed well-opened PV. Pressures measured were – RV systolic pressure decreased to 50 mmHg and PA systolic pressure of 33 mmHg. Saturation improved to 90%.

Figure 1



Right ventricle angioram, lateral view, showing pulmonary value stenosis (arrow), and main pulmonary artery (MRP) dilatation.

Case 2

A 2.5-month-old male child who was a known case of Noonan syndrome, weighing 3.8 kg, born preterm at 34 weeks by emergency LSCS i/v/o fetal distress, presented with respiratory distress since birth. The mother was a case of pregnancy-induced hypertension. On day 3, he developed neonatal hyperbilirubinemia, and late-onset sepsis on day 4. Later, he was diagnosed to have neonatal encephalopathy with seizure, osteopathy of prematurity, anemia, and coagulopathy. He was extensively managed medically for these conditions. On preanesthesia examination, the patient was having swelling over the whole body. He was hemodynamically stable with HR 122/min, RR 60/min, and afebrile and on oxygen support via nasal prong at 4 Lpm, with SpO_2 65%. Blood investigations were within normal limits. ABG – pH – 7.24, pCO_2–50, PO_2 31, Lac 2.6, and HCO_3 21. On echocardiography, severe LVH (HCM, PW 5 mm), severe PS – valvular, dysplastic leaflets, PSG 98 mmHg, PV annulus 8 mm, ASD 4 mm with L–R flow, VSD 4 mm muscular, PDA 4 mm with L-to-R flow, and TAPSE 16 were noted. He was posted for palliative BPV considering the symptoms and high risk for cardiac surgery. It was anticipated for difficult mask ventilation and intubation. The patient was shifted to cath lab on PGE1 infusion. General anesthesia was given with Inj fentanyl plus sevoflurane induction followed by Inj succinylcholine. The patient got intubated by a senior cardiac anesthesiologist and maintained on oxygen, air, and sevoflurane.

Post-BPV, the pressure gradient was not significantly decreased. The patient was shifted intubated to NICU.

Discussion

BPV is a percutaneous procedure for pulmonary stenosis. It has emerged as the treatment of choice for the isolated PS. It is associated with complications ranging from minor and related to vascular puncture to severe and fatal such as suicidal right ventricle and postprocedure pulmonary edema [3].

Presentation of pulmonary stenosis can be in pediatric as well as in adult age group. In pediatric population, presentation can be as preterm and as early as day 1, which imposes different challenges. They may be associated with congenital cardiac disease such as TOF and Noonan syndrome. As in our case 2, the patient was preterm-born known case of Noonan syndrome who had presented for BPV after stabilization and addressing other medical conditions. So, intraoperative management not only directs PS but also associated cardiac and noncardiac anamolies.

In adults, PS may present as progressive symptoms such as exertional dyspnea, light-headedness over the years, or during pregnancy or underlying conditions such as carcinoid syndrome, rheumatic heart disease or cardiac tumor, and depending on the severity of PS [4]. Long-standing and severe obstruction may present as RV failure as in our case 2.

PS can be valvular, supravalvular, and subvalvular according to location of obstruction. In typical valvular PS that is the most common one, the commissures are fused, and leaflets are thin and pliable.

BPV is preferred in moderate- (gradient 40–60 mmHg) to-severe (>60 mmHg) pulmonary stenosis and typical dome-shaped valvular pulmonary stenosis in severe stenosis (gradient >60 mmHg). BPV is also the preferred treatment in neonates with critical pulmonary stenosis [5]. Successful BPV is when with the balloon dilatation of pulmonary stenosis leads to decrease of gradient across PV.

PS with dysplastic leaflet accounts for 10–20%. BPV success rate is lesser with dysplastic valves. Still, some

case series reported up to 65% success. In our case 2, although the patient had dysplastic PV, BPV was done as a palliative procedure considering high-risk status [6].

Complications associated with BPV procedure are transient fall in BP and saturation at the time of balloon inflation as we noticed in case 1, which requires vigilance and prompt correction if hemodynamics are not returned to baseline after balloon deflation.

Another complication is post dilatation pulmonary edema. It is due to increase in blood flow in a pulmonary vascular bed unprepared and associated left-side diastolic dysfuction or mitral stenosis or large left-to-right shunt [7]. Its treatment includes use of diuretics, ventilation, and ionotropic support if required [1]. In our case 2, we anticipated similar complication as the patient was having HCM too, but fortunately we did not face it.

A dreaded complication of BPV is severe RV outflow spasm or obstruction. Efforts to manipulate the catheter in the RVOT during cath study or when positioning the catheter for an outflow angiogram can precipitate severe dynamic sudden RVOT obstruction, leading to cessation of forward flow and sudden hypotension [8].

Management is stopping any further manipulation around RVOT, volume expansion, and intravenous betaâ blockers or calcium channel blockers [9].

Similar situation can occur in severe PS, after relief with balloon valvuloplasty or surgery, RV infundibular spasm creates suprasystemic RV pressure and this can lead to low cardiac output. This life-threatening clinical condition is known as suicidal right ventricle. It can occur in children and adults with preprocedure suprasystemic RV pressure. Its treatment is as mentioned in RV outflow spasm [9].

Plan of anesthesia in these cases depends on age of patients, hemodynamics, and associated congenital anamolies. General anesthesia in pediatric patients and MAC is preferred for adults in our institute. Cardiac grid for the management of severe pulmonary stenosis is avoiding tachycardia, maintaining preload, afterload, and contractility.

As a part of the team, an anesthesiologist knowledge of the disease as well as presentation and pathophysiology of pulmonary stenosis not only helps in management of the procedure but helps in avoiding and prompt treatment of the complications, especially during manipulation across RVOT, balloon inflation, and postoperative course.

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Conflicts of interest
There are no conflicts of interest.

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