CASE REPORT

Successful High-Risk Pulseless-Balloon Aortic Valvuloplasty Procedure in Uncorrected Pulmonary Atresia with Severe Congenital Aortic Stenosis and Low Left Ventricular Function

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ABSTRACT

Background: Pulmonary atresia with ventricular septal defect (PA-VSD) and severe bicuspid aortic stenosis is an uncommon condition that presents significant surgical challenges. The dual obstruction leads to chronic pressure overload, resulting in ventricular hypertrophy and decreased systolic function. A low left ventricular ejection fraction (LVEF) increases the risk of mortality during surgical interventions. Percutaneous balloon aortic valvuloplasty (PBAV) poses additional risks due to the intentional induction of pulselessness during balloon inflation. This case report aims to detail the perioperative management strategies employed during PBAV in a patient with PA-VSD, severe aortic stenosis, and low LVEF.

Case: We present a 19-year-old female patient weighing 45 kg with shortness of breath and fatigue. Her oxygen saturation was measured at 90% across all extremities. Electrocardiogram (ECG) findings indicated sinus rhythm with biventricular hypertrophy and incomplete left bundle branch block (LBBB). Chest X-ray revealed cardiomegaly and pulmonary artery dilation, while echocardiography confirmed PA-VSD, severe aortic stenosis with a mean pressure gradient (mPG) of 55 mmHg, and an LVEF of 41.3%. A percutaneous pulseless-BAV was performed using a Tyshak balloon via an antegrade transvenous femoral approach under fluoroscopy and transesophageal echocardiography guidance.

Discussion: Anesthesia management focused on gradual medication titration, minimizing patient movement during the procedure, and ensuring comfort due to transesophageal echocardiogram (TEE) probe insertion. A temporary pacemaker was placed in the right ventricular apex for pacing at 220 beats per minute until cardiac arrest occurred post-balloon inflation, and defibrillation successfully restored spontaneous circulation.

Conclusion: This case illustrates that PBAV in uncorrected PA-VSD with severe aortic stenosis and low LVEF is feasible despite its inherent risks when conducted by a skilled team utilizing careful judgment throughout the procedure.

Keywords: aortic stenosis; balloon aortic valvuloplasty; cardiovascular anesthesia; congenital heart disease; pulmonary atresia

INTRODUCTION

Pulmonary atresia with ventricular septal defect (PA-VSD) is a rare congenital heart defect, occurring in approximately 7 out of every 100,000 live births.[1] This condition represents a severe form of Tetralogy of Fallot (ToF), necessitating distinct management strategies, particularly in late-presenting patients, where clinical manifestations can vary significantly.[2] Accurate screening and diagnosis are essential and can be effectively achieved through segmental sequential analysis of echocardiography.[3]

The presence of bicuspid aortic valves in patients with PA-VSD is uncommon.[4] However, the coexistence of severe bicuspid aortic stenosis alongside PA-VSD has not been previously reported in the literature. This highlights the uniqueness and complexity of our case. The combination of these conditions poses significant challenges due to their impact on left ventricular function, which may be compromised as evidenced by decreased left ventricular ejection fraction (LVEF).

This case report aims to elucidate the management approach for a patient diagnosed with pulmonary atresia, ventricular septal defect (VSD), and severe bicuspid aortic stenosis accompanied by diminished left ventricular function. Specifically, we will detail the high-risk procedure involved in performing pulseless balloon aortic valvuloplasty (BAV). By sharing this experience, we hope to contribute valuable insights into managing similar cases that present unique challenges within pediatric cardiology.

CASE

A 19-year-old woman presented to the outpatient clinic with complaints of easy fatigability for the past year. Her symptoms were relieved by rest and worsened with physical activity. Her parents reported that she often turned blue while crying since the age of 3. There was no history of recurrent respiratory tract infections. She had previously been diagnosed with ToF but was lost to follow-up.

On physical examination, her vital signs were within normal limits except for an oxygen saturation of 90% in all four extremities. Her height was 151 cm, and her body weight was 45 kg. Cardiac examination revealed a high-pitched ejection systolic murmur at the upper left sternal border. An electrocardiogram (ECG) showed sinus rhythm at 75 beats per minute, left axis deviation, biventricular hypertrophy, and an incomplete left bundle branch block (LBBB) (Figure 1). Chest X-ray revealed cardiomegaly with dilation at the base of the heart, concavity at the region of the pulmonary arteries, a normal broncho-vascular pattern, and a mild left-sided pleural effusion (Figure 2). Laboratory investigations were unremarkable.

Transthoracic echocardiography (TTE) demonstrated situs solitus, concordant atrioventricular connections, pulmonary atresia, and an overriding aorta of approximately 50%. All pulmonary veins drained into the left atrium. There was no atrial septal defect, but a bidirectional shunt was present through a subaortic VSD. The LVEF was decreased at 41.3%, while right ventricular contractility was preserved, with a tricuspid annular plane systolic excursion (TAPSE) of 25 mm. Severe valvular aortic stenosis with mild to moderate aortic regurgitation was also noted, caused by a calcified bicuspid aortic valve with post-stenotic dilation

(Figure 3). The aortic annulus measured 30 mm, and the descending thoracic aorta measured 15.1 mm in diameter. Mild pericardial effusion was also observed.

Pre-procedural TTE and TEE revealed concentric LVH, decreased LV function, valvar aortic stenosis with rudimentary noncoronary cups / NCC (bicuspid physiology), mild aortic regurgitation, overriding 50% of the aorta, with an aortic root size was 30 mm, Pulmonal Atresia, VSD subaortic bidirectional shunt (Figure 4). Given the low LVEF, a decision was made to perform percutaneous BAV using a retrograde transvenous approach. The patient underwent general anesthesia with premedication consisting of fentanyl 100 mcg, propofol 120 mg, midazolam 1 mg, and rocuronium 30 mg. She was intubated using an endotracheal tube (ETT) size 7 and ventilated with FiO_2 30%, resulting in a post-intubation oxygen saturation of 88%.

The right internal jugular vein was punctured, followed by the insertion of a 5F sheath. A 5F bipolar temporary pacemaker lead was introduced and advanced to the right ventricular apex. Pacing threshold was 1 A with an R-wave of 6 mV. The temporary pacemaker was set at a sensitivity of 3 mV, output of 3 A, and a rate of 50 bpm. Defibrillator pads were applied and connected to a defibrillation machine.

A 6F sheath was inserted into the right femoral artery (RFA), and a retrograde transvenous approach for BAV was initiated. A pigtail catheter was advanced into the aorta via the RFA. A 0.035-inch guidewire was positioned across the aortic valve into the left ventricle. Initially, attempts to cross the severely stenotic aortic valve using a JR guiding catheter were unsuccessful due to the tight valve and tortuous anatomy. The guiding catheter was exchanged for a JR diagnostic catheter, and with the help of a 0.035" exchange-length Terumo wire, the catheter was successfully advanced across the VSD into the aorta. An extra-stiff Lunderquist wire was then placed into the distal descending aorta.

A balloon catheter (Tyshak II, 30 mm x 3 cm x 100 cm) was advanced over the stiff wire and positioned across the aortic valve (Figure 5). Rapid right ventricular pacing was performed using the indwelling temporary pacemaker to induce pulseless ventricular tachycardia (pVT) and stabilize balloon position during balloon inflation. The first inflation was suboptimal due to misalignment and incomplete balloon expansion. The temporary pacemaker rate was reduced, and the patient returned to sinus rhythm.

A second balloon inflation was performed with careful repositioning, and this time the balloon fully expanded at 1.5 atm for 10 seconds. Following balloon deflation, the patient developed pVT (Figure 6). A 200 J DC shock was delivered, followed by one minute of cardiopulmonary resuscitation, which successfully restored sinus rhythm with a palpable pulse.

A post-procedural pressure pullback from the ascending aorta to the right ventricle showed no significant residual pressure gradient (Figure 7). The criteria for a successful BAV—reduction of more than 50% in the mean transaortic pressure gradient—were achieved, with the gradient reduced from 55 mmHg to 21 mmHg. The patient was transferred to the ICU for monitoring and was sedated overnight.

The following day, the patient was fully awake without any neurological deficits. She showed significant clinical improvement. A follow-up TTE revealed improved LV function with LVEF increased to 66.1%, a reduced mean aortic gradient to 21 mmHg, no worsening of aortic regurgitation, and no increase in pericardial effusion (Figure 8). The patient was extubated and subsequently discharged in stable condition.

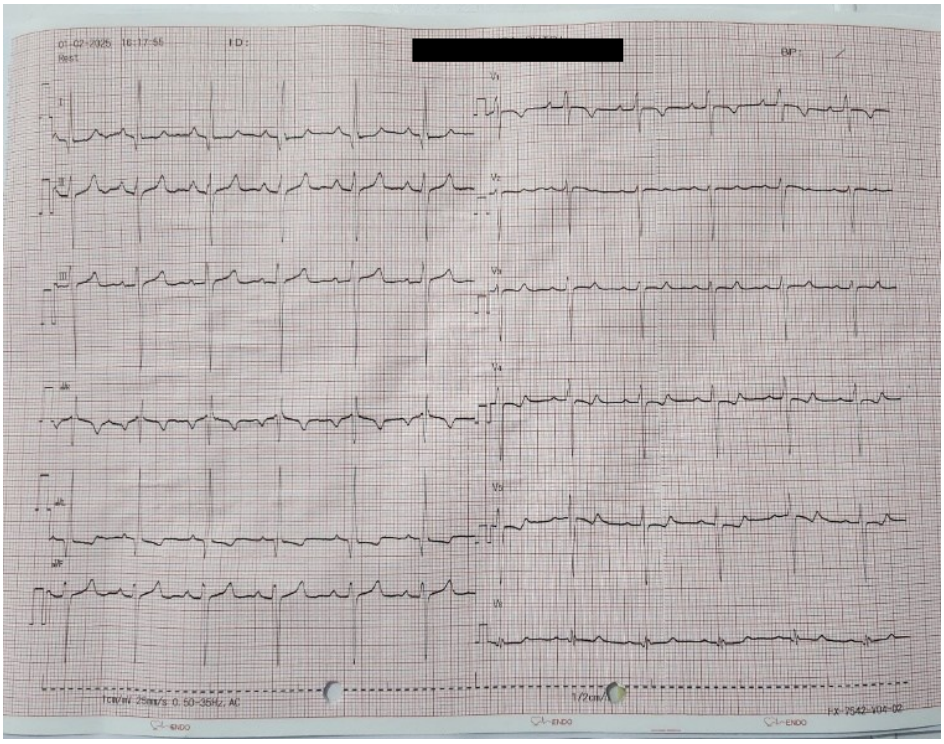
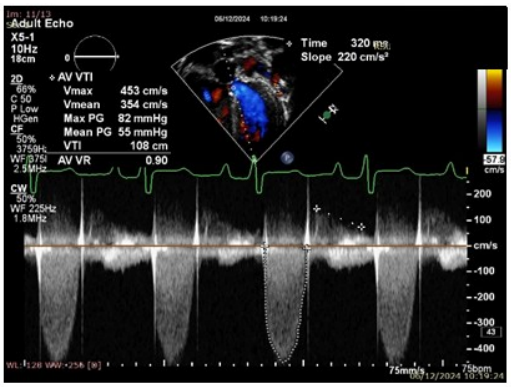


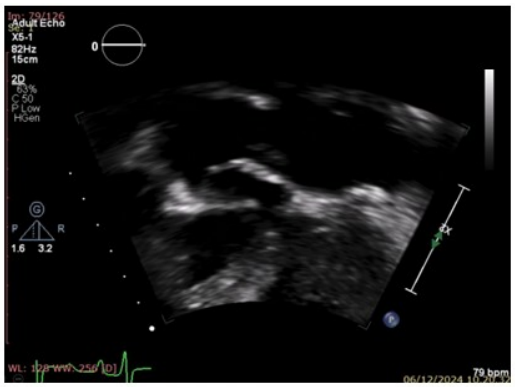
Figure 1. Electrocardiogram (ECG) showing sinus rhythm with biventricular hypertrophy and incomplete left bundle branch block (LBBB)



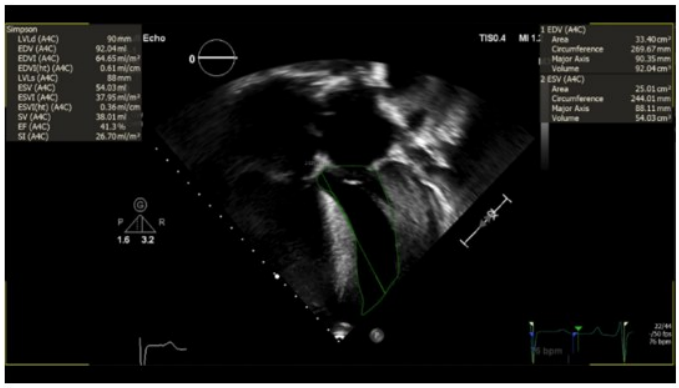
Figure 2. Chest X-ray demonstrating cardiomegaly with dilatation of the base of the heart and a concave region of the pulmonary arteries



3.1



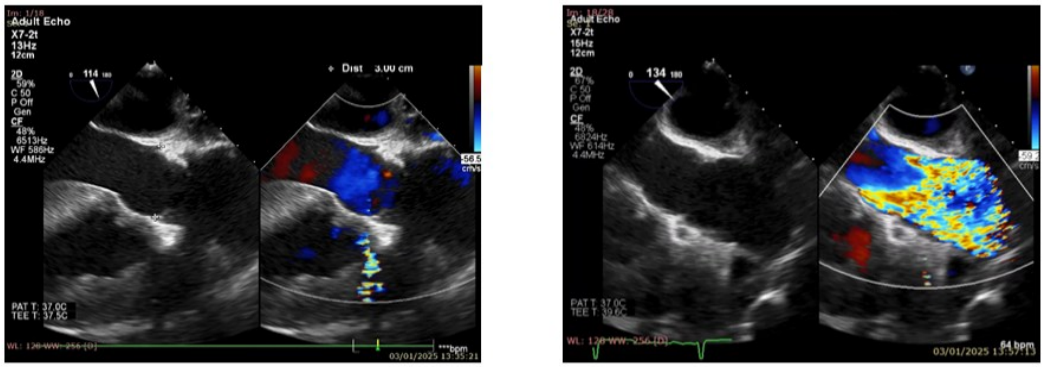
3.2



3.3

Figure 3. Transthoracic echocardiography (TTE) showed severe bicuspid aortic stenosis with mild to moderate AR and a low LVEF

3.1 The image displays the velocity-time integral (VTI) and pressure gradients, indicating significant aortic stenosis with a maximum velocity of 453 cm/s and a mean pressure gradient of 55 mmHg.
3.2 Two-dimensional echocardiographic image showing the left ventricular outflow tract (LVOT) and aortic valve
3.3 Key parameters include left ventricular end-diastolic volume (EDV) of 92.04 ml, end-systolic volume (ESV) of 54.03 ml, and an ejection fraction (EF) of 41.3%.



4.1 4.2

Figure 4. Pre-procedure TTE and TEE showed severe domming of the aortic valve

4.1 The left panel displays the anatomical structure, while the right panel illustrates the flow dynamics with a distance measurement of 3.00 cm.

4.2 The left panel displays the anatomical structure, while the right panel illustrates significant flow dynamics with a peak velocity of 59.2 cm/s.

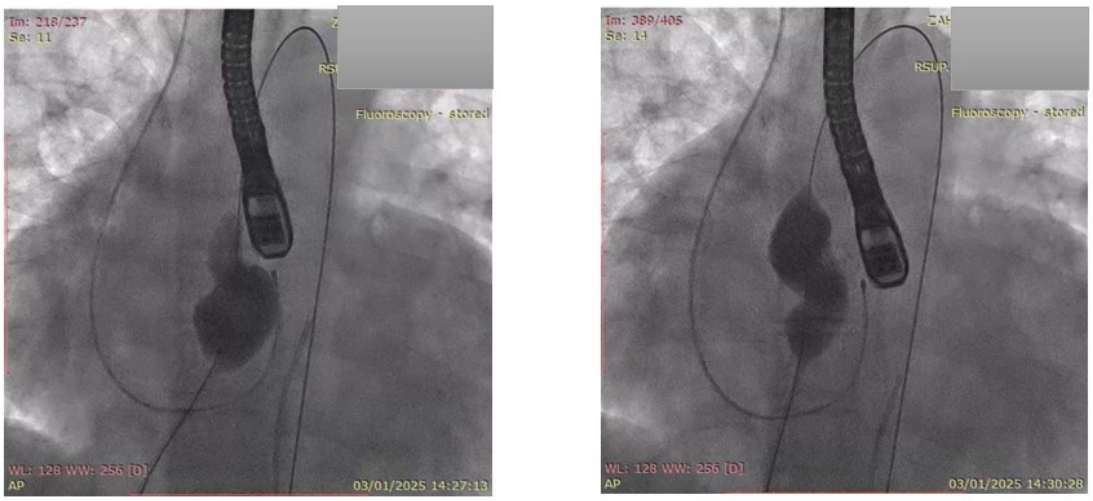


Figure 5. Tyshak II balloon 30 mm x 3 cm x 100 cm inflated

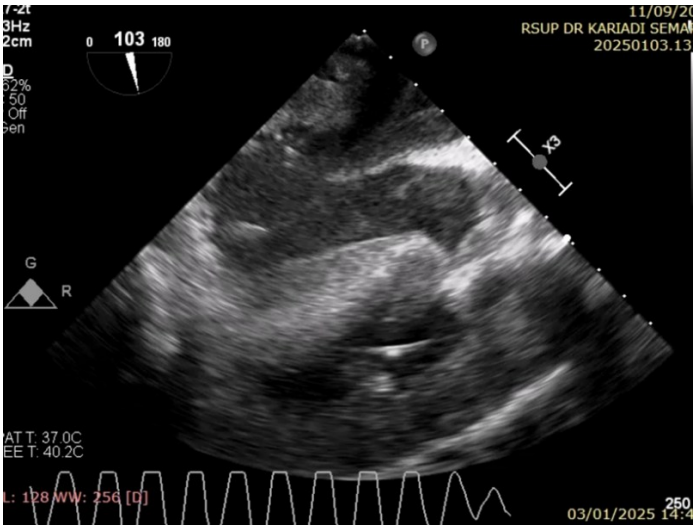


Figure 6. TEE showed pulseless-heart with VT rhythm during balloon inflation

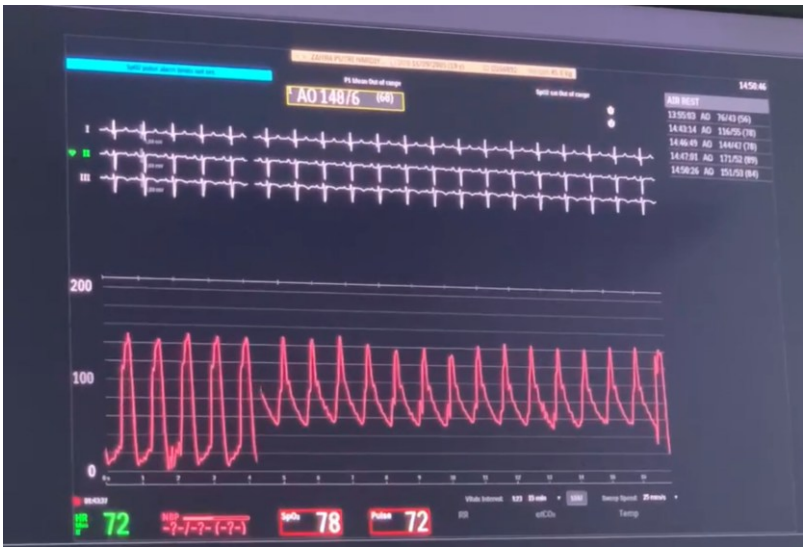
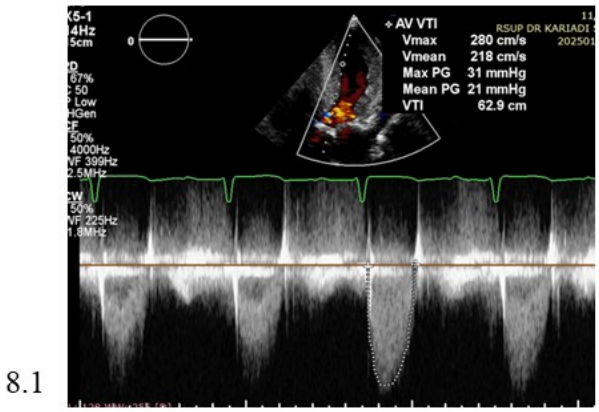
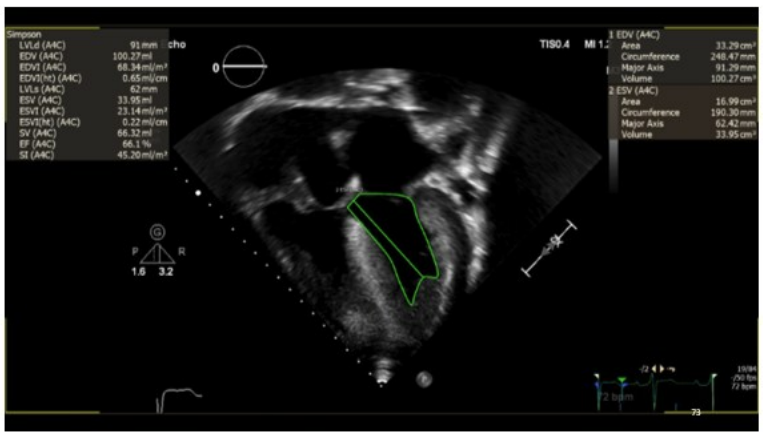


Figure 7. Ao-RV pullback shows there was nearly no pressure gradient between Ao and RV



8.1



8.2

Figure 8. TTE evaluation showed mild aortic stenosis with mild to moderate aortic regurgitation and an improved LVEF

8.1 The parameters include a maximum velocity (Vmax) of 280 cm/s, a mean pressure gradient (Mean PG) of 21 mmHg, and a velocity-time integral (VTI) of 62.9 cm.
8.2 Key parameters include left ventricular end-diastolic volume (EDV) of 100.27 ml, end-systolic volume (ESV) of 33.95 ml, and an ejection fraction (EF) of 66.1%

DISCUSSION

The present case report details a successful percutaneous balloon aortic valvuloplasty (PBAV) performed on a woman with pulmonary atresia, VSD, and severe bicuspid aortic stenosis, accompanied by diminished LV function.[5] As noted by Olasińska-Wiśniewska et al.[6], while the significance of the disease is similar in children and adults, the etiology, course, and forms of treatment, including interventional cardiology methods, differ with the patient's age. This highlights the distinct considerations for managing aortic stenosis in the pediatric population compared to adults.

Low LVEF in late-presenting ToF patients is usually caused by hypoxia; however, in this patient, the oxygen saturation was measured at 90% in all four extremities.[7] Thus, hypoxia is not suspected to be the cause of the low LVEF. The presence of severe bicuspid aortic stenosis results in high afterload that must be overcome by the ventricles. Signs of this overload can be observed through increased thickness of the left ventricular muscle. When the ventricles fail to compensate for this pressure overload, LVEF decreases.[8]

The procedural success in this specific case, demonstrated by the reduction in the aortic gradient (Figure 7) and the subsequent improvement in left ventricular function with a reduced mean pressure gradient on follow-up TTE (Figure 8), is consistent with the findings of other studies on PBAV for congenital aortic stenosis. For instance, a retrospective study by Pan et al.[5] reported that the peak transaortic gradient significantly decreased immediately post-PBAV in patients with congenital aortic stenosis under echocardiographic guidance. Similarly, Charafeddine et al.[9] described their experience at a tertiary center, demonstrating the feasibility of balloon valvuloplasty for congenital aortic stenosis with high immediate success rates.

The BAV procedure can be performed using two approaches: the antegrade transvenous approach and the retrograde transarterial approach. Both methods have their own advantages and disadvantages. The antegrade transvenous approach typically requires a transseptal puncture; therefore, the retrograde transarterial approach is generally preferred. However, crossing the severely narrowed aortic cusps can sometimes be challenging. In this case, the antegrade transvenous approach was chosen due to the presence of a subaortic VSD with an overriding aorta, which allowed easier access to the left ventricular outflow tract. The size of the balloon should be carefully selected to maximize the benefits of BAV while minimizing potential risks. A balloon–to–annulus ratio of 0.9 : 1 to 1 : 1 is commonly used to guide balloon sizing. In this case, the aortic annulus measured 30 mm, which is considered quite large. Accordingly, a semi-compliant cylindrical Tyshak II balloon, size 30 mm, was selected to achieve a balloon-to-annulus ratio of 1:1. While hourglass-shaped balloons, such as the Nucleus balloon, are now preferred for improved positioning and stability during inflation, their availability and size options remain limited.[10]

A reduction in the mean transaortic valvular gradient of 50 mmHg or 40–50% from the baseline is considered a criterion for successful BAV. In this patient, the criteria were met, with the initial mean gradient decreasing from 55 mmHg to 21 mmHg, representing a

reduction of more than 50%. The patient's LVEF improved, from 41.3% to 66.1%.

Olasińska-Wiśniewska et al.[6] emphasize that treatment strategies differ with age, suggesting that managing aortic stenosis in conjunction with other critical congenital heart defects requires a specialized and multidisciplinary approach. The ACC/AHA guidelines on perioperative cardiovascular evaluation and management also underscore the importance of considering underlying cardiac conditions in the context of procedures.[11]

The utilization of echocardiographic guidance was crucial in this case, as evidenced by the pre-procedural assessments using TTE and TEE (Figures 3 and 4) and intra-procedural monitoring (Figure 6). This is in line with the increasing adoption of echocardiography as the sole or primary guidance modality for PBAV in congenital aortic stenosis, as demonstrated by Pan et al.[5], who reported a high success and low complication rate using Doppler echocardiography alone.

While BAV in adults with severe aortic stenosis is often considered a palliative measure or a bridge to definitive surgical, its role in congenital aortic stenosis, particularly in complex cases like ours, is often focused on palliating the stenosis, improving hemodynamics, and potentially delaying or avoiding early surgical intervention.[12] The long-term outcomes of BAV for congenital aortic stenosis often involve the need for reintervention, and the progression of aortic regurgitation is a known concern.

Considering the broader context of aortic stenosis management, the ESC/EACTS and ACC/AHA guidelines primarily focus on the management of aortic stenosis in adults.[13] While these guidelines emphasize the importance of a Heart Team approach and risk assessment[14,15], the specific nuances of managing complex congenital heart disease in patients undergoing BAV are not their primary focus. However, the underlying principle of a multidisciplinary team evaluating the risks and benefits of intervention certainly applies to our case.

This case underscores the feasibility and immediate benefits of PBAV in a patient with severe congenital aortic stenosis complicated by pulmonary atresia and a VSD. The successful reduction of the aortic gradient and improvement in left ventricular function highlight the potential of BAV to provide effective palliation in this challenging clinical scenario. Last but not least, it is very important to have great teamwork. Delegate properly to every team member with an exact and clear job description before starting the procedure. Every wrong decision could lead to a worse and fatal outcome.

CONCLUSION

Percutaneous BAV in a patient with uncorrected PA-VSD, severe aortic stenosis, and low LVEF is a challenging and high-risk procedure. However, with careful consideration and a collaborative team approach, the procedure can be performed successfully, resulting in significant hemodynamic and clinical improvement as a bridge to definitive therapy.

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