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Case Report

Overcoming critical aortic stenosis: a rare case report on successful balloon valvotomy at 16 hours of life

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ABSTRACT

Congenital aortic stenosis is a significant cardiac malformation, comprising 5% of cardiac disorders in childhood, and is more prevalent in males. The most common type is bicuspid aortic valve and the stenosis can lead to a mortality rate of 40-50% if left untreated. Initial presentations often require urgent hemodynamic, respiratory, and metabolic resuscitation. Effective management aims to maintain systemic blood flow, typically through balloon dilation or surgical valvotomy. In this case, we present a neonate born to a 39-year-old G6P5L5 mother via normal vaginal delivery, with a birth weight of 2.7 kg. Although the mother had no antenatal risk factors, an anomaly scan revealed aortic stenosis with left ventricular hypertrophy. The infant cried immediately after birth, but an ejection systolic murmur was detected in the right second and third intercostal spaces on auscultation. Postnatal echocardiography confirmed severe aortic stenosis, significant left ventricular systolic dysfunction, mild to moderate mitral regurgitation. At 16 hours of life, the neonate underwent successful percutaneous balloon aortic valvotomy via right femoral access, leading to improved hemodynamics. The infant was discharged on day 5 and is now thriving at one year of age on anti-failure medications. This case underscores the vital role of early detection and intervention in congenital heart disease. Timely management significantly improves outcomes and mitigates the risks associated with congenital aortic stenosis. The integration of prenatal screening and immediate postnatal evaluation is crucial for optimizing care in affected infants, highlighting advancements in paediatric cardiology that enhance prognoses for these conditions.

Keywords: Congenital aortic stenosis, Balloon dilatation, Early intervention

INTRODUCTION

Neonatal aortic stenosis is a significant congenital heart defect that contributes to 3-6% of all cardiac malformations.[1] It arises from an obstruction of the left ventricular outflow tract at the aortic valve, often due to structural anomalies such as bicuspid or unicuspid valves, or fused or malformed cusps. These malformations restrict blood flow from the left ventricle to the aorta, leading to critical symptoms within the first few weeks of life. The condition typically requires immediate intervention to stabilize the infant's cardiovascular, respiratory, and metabolic systems. Once stabilization is achieved, the primary goal is to improve systemic blood flow, typically through procedures like balloon aortic valvuloplasty or surgical valvotomy, which relieve the obstruction by enlarging the aortic valve opening.[2] Neonatal aortic stenosis differs markedly from the condition seen in older children, as the severity and timing of the obstruction in utero can significantly affect left ventricular development.

During fetal life, if the obstruction is moderate to severe and develops early, it can drastically reduce left ventricular output, causing rapid thickening of the ventricular wall. This thickening impairs the ventricle's ability to fill properly, reducing blood flow through the foramen ovale. In severe cases, this process leads to a hypoplastic left ventricle due to the decreased cavity size. However, in less severe or later-onset cases, although the left ventricle still thickens, the impact on its cavity size is less pronounced. After birth, the physiological changes that occur such as

decreased pulmonary vascular resistance and closure of the ductus arteriosus further stress the left ventricle. In severe cases, the left ventricle may be unable to sustain systemic blood flow adequately, making the ductus arteriosus essential for maintaining circulation temporarily.[3]

The clinical presentation of neonatal aortic stenosis varies depending on the severity of the obstruction. In milder cases, symptoms may include poor feeding, failure to thrive, and mild respiratory distress. Severe cases can present with signs of heart failure, such as poor peripheral perfusion, cool extremities, hepatomegaly, and edema. Lethargy or irritability may also occur due to decreased cardiac output. Prompt diagnosis and intervention are crucial; as early treatment can significantly improve outcomes. We discuss about a particularly urgent case involved a neonate diagnosed prenatally with suspected aortic stenosis, which was confirmed postnatally through echocardiography. The infant underwent a successful surgical intervention at just 16 hours of life, underscoring the critical need for timely management in such severe cases.

CASE REPORT

This was the case of a term female newborn baby delivered via normal vaginal delivery with a birth weight of 2.72 kg. She was born to 39-year-old G6P5L5 mother with no antenatal risk factors. Anomaly scan showed aortic stenosis with left ventricular hypertrophy. Baby cried immediately after birth and was shifted to neonatal intensive care unit (NICU) for further management. Baby had tachypnea at admission and ejection systolic murmur was heard in the right second and third intercostal space in the aortic area. Postnatal ECHO done within few hours of birth showed congenital bicuspid aortic valve, severe aortic stenosis, severe LV systolic dysfunction, mild-moderate mitral regurgitation, with no pulmonary arterial hypertension.

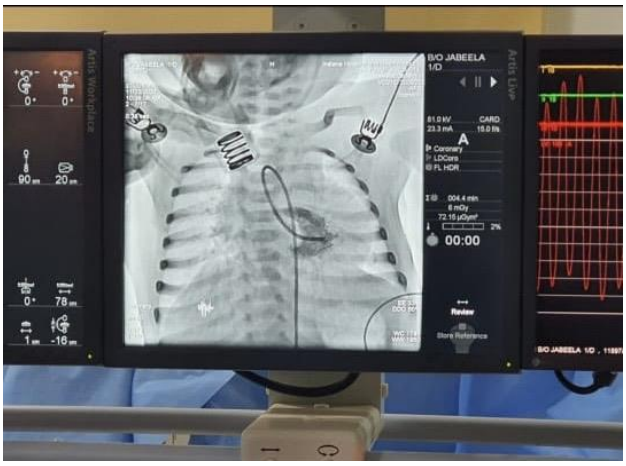

Figure 1: Image of balloon aortic valvotomy for the baby done at 16 hours of life via right femoral access.

At 16 hours of life, percutaneous balloon aortic valvotomy was done via right femoral access under local anesthesia. Baby withstood the procedure well. Pre- procedure LV-Ao gradient was 75 which dropped to less than 20 post procedure. Repeat ECHO showed improving LV function with no evidence of aortic stenosis. Baby was observed in NICU for 48 hours and was discharged by day 5 of life. Now, she is one-year-old and is thriving well on anti-failure medications.

DISCUSSION

Congenital aortic stenosis (VAS) presents with different valve types: unicuspid, bicuspid, tricuspid, and quadricuspid.[4] The unicuspid valve, often seen in severe cases, has a small orifice with one or no commissures. Its single thickened leaflet opens minimally, causing significant obstruction and making balloon valvuloplasty challenging. The bicuspid aortic valve is the most common, affecting about 2% of the population.[5]

Bicuspid aortic valve is classified into three types based on the number of raphes: type 0 (no raphe), type 1 (one raphe, the most common form in 90% of cases), and type 2 (two raphes). Valvular aortic stenosis can develop when the valve leaflets are abnormally short, dysplastic, or have fused commissures. In infants and fetuses, this stenosis is often linked to underdeveloped left heart structures, left ventricular dysfunction, and endocardial fibroelastosis. Children born with bicuspid aortic valve also tend to have larger ascending aortas, which dilate more rapidly than those with tricuspid aortic valves, likely due to abnormal neural crest cell development.[4]

Congenital aortic stenosis, whether valvar or otherwise, frequently remains undiagnosed during the neonatal period, particularly in asymptomatic patients who might be identified only through incidental findings such as an ejection click or systolic murmur. These infants often present with minimal abnormalities on electrocardiography (ECG) and chest X-ray, and echocardiography. Doppler studies typically reveal good ventricular function with a transvalvular gradient of less than 50-60 mmHg. This group represents a different disease spectrum compared to symptomatic newborns presenting with aortic stenosis within the first month of life. For asymptomatic patients, a conservative management strategy is recommended, which includes regular echocardiographic monitoring. Conversely, symptomatic infants represent a medical and surgical emergency, exhibiting signs of tachypnea, congestive heart failure, and, in some instances, hepatomegaly and edema. Unlike older patients who usually present with exercise intolerance or angina, neonates often show rapid respiration, dyspnea, and tachycardia, with pallor and diminished peripheral pulses emerging as cardiac output declines.

Physical examination findings in symptomatic neonates reveal distinct features, including a loud first heart sound and a narrowed second sound, indicative of a poorly

mobile valve. An ejection click may be absent, and a grade 2-3/6 ejection systolic murmur is generally heard at the right upper sternal border, though atypical murmurs can occur elsewhere. In critically ill infants, a quiet murmur may accompany an apical gallop rhythm, while signs of metabolic acidosis, significant hepatomegaly, and reduced peripheral pulses correlate with increased surgical risk. Electrocardiographic findings may show left ventricular hypertrophy in about 60% of cases, and it is noteworthy that 10-50% of neonates may exhibit right ventricular hypertrophy or combined ventricular hypertrophy. Timely recognition and intervention are vital for improving outcomes; asymptomatic patients benefit from regular monitoring, while symptomatic neonates require immediate medical intervention to manage the underlying obstruction effectively.[6]

The treatment of critical aortic stenosis (CAS) in neonates presents significant challenges due to the complexity of the condition and the unstable clinical status of the patients. Various approaches have been explored, including surgical aortic valvotomy (SAV) and percutaneous balloon aortic valvuloplasty (BAV), both of which are lifesaving but have notable limitations.[7]

SAV, while offering a more direct intervention, is associated with high risks such as postoperative aortic insufficiency, left ventricular failure, and the need for reintervention. The complexity of congenital aortic stenosis, with its heterogeneity and potential for additional cardiac anomalies like left ventricular fibroelastosis and mitral valve abnormalities, further complicates the outcomes of SAV. Aggressive valvotomy can exacerbate left ventricular damage due to myocardial ischemia during the procedure.[8]

BAV, on the other hand, offers a less invasive approach, avoiding general anesthesia, cardiopulmonary bypass, and the complications of open-heart surgery.[9] It achieves reduction in transvalvular gradients through valve tissue stretching and commissural fusion rupture. However, it carries its own risks, including femoral artery injury, cusp perforation, and severe aortic insufficiency.[9,10] Despite advancements in catheter size and technique (e.g., using the umbilical artery approach), BAV remains prone to complications. SAV and BAV share comparable efficacy and mortality rates. Zeevi et al, along with several other studies, have demonstrated that left ventricular size is strongly correlated with outcomes in neonates with critical aortic stenosis, regardless of the procedure chosen. Poor left ventricle output are often linked to worse prognoses, highlighting the critical role of ventricular size in determining long-term success.[11]

CONCLUSION

This case exemplifies the critical role of early diagnosis and intervention in the management of neonatal aortic stenosis, a condition with potentially life-threatening implications. The successful balloon valvotomy performed at 16 hours of life not only alleviated the severe obstruction but also improved left ventricular function, demonstrating the effectiveness of less invasive techniques in this vulnerable population. As advancements in prenatal screening and neonatal cardiology continue to evolve, timely interventions can significantly enhance survival rates and long-term health outcomes. This case reinforces the necessity of a collaborative, multidisciplinary approach to congenital heart disease, highlighting how prompt and appropriate treatment strategies can transform prognosis and quality of life for affected infants.

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