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Case Report

Closing the gap: Navigating the percutaneous path for a 2-month-old infant's perimembranous ventricular septal defect

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ABSTRACT

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Ventricular septal defects (VSDs) represent a substantial portion of congenital heart diseases, with perimembranous VSD (pmVSD) being the most prevalent variant. While surgical closure is conventional, it carries risks such as atrioventricular block, infection, and neurological issues. In contrast, catheter-based interventions have shown promise, particularly for muscular VSDs, but data for pmVSD closure remains limited. We present a case of a 2-month-old male infant referred for heart failure symptoms, characterized by a grade 3/6 pan systolic murmur and mild cardiomegaly on chest X-ray. Echocardiography revealed a 5.5 mm pmVSD with a peak gradient of 32 mm of Hg and pulmonary artery systolic pressure of 10 mm of Hg. Despite de-congestive therapy, the patient showed no symptomatic improvement, prompting consideration of percutaneous device closure due to the infant's low body weight (3 kg) and failure to thrive.

This case report discusses the successful percutaneous closure of a perimembranous ventricular septal defect (pmVSD) in the youngest patient ever recorded. It highlights the effectiveness of transcatheter techniques in managing congenital heart defects in young patients.

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1. Introduction

Ventricular septal defects (VSDs) are prevalent heart conditions, encompassing nearly one-third of all cases. Among the various types, perimembranous VSD (pmVSD) is the most common,¹ presenting unique challenges in management compared to other variants. While surgical closure has been the standard approach, percutaneous device closure has gained traction, particularly for its minimally invasive nature and potential benefits in pediatric patients.²

Catheter-based interventions for muscular VSDs have shown promising results since 1988, with the youngest

patient being 8 months old infant, with increasing application in pediatric populations.³ However, data specific to perimembranous VSD closure, especially in low-body

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weight patients, remains relatively scarce. The feasibility and outcomes of percutaneous device closure in such cases are critical considerations, given the challenges posed by smaller vascular access and anatomical variations.

This report highlights a noteworthy case of successful percutaneous device closure of a pmVSD using the Amplatzer duct occluder device II in a 2-month-old male infant weighing merely 3 kg, who presented with heart failure. "It is the youngest age ever recorded for the closure of such a pmVSD". The procedure exemplifies the evolving role of transcatheter interventions in pediatric cardiology, particularly in addressing the complexities of pmVSDs in neonates and young infants with limited body weight.

2. Case Presentation

A 2-month-old male infant was referred to the cardiology OPD due to increased breathing efforts, increased heart

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rate, and failure to thrive. The baby was born through a cesarean section due to postdated pregnancy, with a birth weight of 2.65 kg, and was currently weighing 3 kg. The parents were consanguineous. His mother noticed that he had dyspnea during feeding and was irritable. A pan systolic murmur of grade 3/6 was found in the left parasternal area by physical examination. Chest X-ray was suggestive of mild cardiomegaly. Echocardiography showed 5.5 mm of perimembranous VSD with peak gradient across VSD of 32 mm of Hg with pulmonary artery systolic pressure of 10 mm of Hg.

Despite the de-congestive therapy, the patient did not have any symptomatic relief and was not gaining weight appropriately. Because of the failure to thrive, the case was discussed with a pediatric cardiovascular surgeon, and the percutaneous closure of the defect was planned.

2.1. Procedure

After well-informed written consent, the patient was sedated and intubated by the anesthesia team. The patient was loaded with a tablet of aspirin 5mg/kg body weight on the prior night. Right femoral artery (RFA) and right femoral vein (RFV) access were obtained with a 4F sheath. A 4 F pigtail catheter was then inserted on PTFE guiding wire through RFA till the ascending aorta and placed inside the Left ventricle (LV) and an LV shoot was taken in the left anterior oblique (LAO) cranial view which showed VSD of size 5.2 mm, followed by removal of the pigtail (Figure 1 A). A 4F JR was inserted through RFA over the guiding wire and placed in the ascending aorta (Figure 1 B). A Terumo wire passed through the JR catheter, VSD crossed and the wire progressed to the right ventricle (RV) to the right atrium (RA) to the inferior vena cava (IVC). A Cocoon 6 F sheath was inserted through RFV, and a terumo wire was snared inside IVC with the help of an amplatz snare catheter. The amplatz duct occluder device (ADO II) of size 6 x 4 MM passed through the delivery sheath across VSD followed by the release of the LV rim of the device under fluoroscopy (Figure 1 C & D). To confirm the proper device position, LV shoot was taken (Figure 1 E), followed by the device's release from the delivery cable (Figure 1 F).

After the procedure, the patient remained stable with echocardiography showing a properly placed VSD device in situ with no residual shunt across the device, no evidence of pericardial effusion; or any electrical disturbances on the ECG. However, due to a significant amount of blood loss, the patient received a blood transfusion and was closely monitored in the ICCU for 24 hours before being discharged.

2.2. Follow up

After only four weeks, the child began to gain weight and showed an improvement in oral intake. All of his symptoms had completely resolved, and medications for congestive heart failure were stopped after six weeks. Follow-up echocardiography showed no residual shunt across the device and no evidence of aortic or tricuspid regurgitation, as well as no signs of pericardial effusion. Additionally, the ECG showed no abnormalities in conduction or rhythm.

We have been following the case for the last 2 years and the patient was doing well.

3. Discussion

Transcatheter closure of ventricular septal defects (VSDs) has become a well-established procedure in older pediatric patients, but its application in very young infants, especially those weighing less than 5 kg, remains a challenging frontier. Our case represents a significant milestone as it reports the successful transcatheter closure of a perimembranous VSD (pmVSD) using the Amplatzer duct occluder device II in a 2-month-old male infant weighing only 3 kg, to our knowledge, the youngest reported age for such a procedure.

Percutaneous closure of pmVSDs poses distinct challenges due to the defect's proximity to the aortic valve and the conduction system, as well as the smaller anatomy and fragility in younger patients. Key technical challenges include ensuring appropriate local site puncture, selecting an optimal sheath size, and delivering devices with precision to avoid complications such as residual shunts or device embolization.

As in our case, transcatheter retrograde closure of VSD with ADO II has been mentioned in the literature previously.⁴

Percutaneous closure of ventricular septal defects (VSDs) has been reported in the literature for children as small as 3.2 kg.⁵ However, in cases where the child weighs less than 5 kg, Literature reports highlight varying success rates and complications associated with transcatheter VSD closure in infants, emphasizing the critical role of operator expertise and careful patient selection. While our case encountered minimal procedural complications, such as local site bleeding necessitating blood transfusion, the absence of major device-related issues underscores the importance of meticulous procedural planning and execution.

Zartener et al. reported transvascular closure of single and multiple muscular ventricular septal defects in neonates and infants < 20 kg in 17 patients, with a success rate of 88%, reducing the inter-ventricular shunt and improving the hemodynamic situation in 14 patients. A patient with the lowest weight of 2.2 kg had developed an acute AV block which led to the immediate removal of the device. One Amplatzer muscular VSD occluder could not be delivered due to the sharp bending of the delivery sheath in 2004. ⁶ Device closure of pmVSD is more challenging due to its proximity to the aortic valve and the conduction system, as



Figure 1: A-F: Cine fluoroscopic images of steps ofpmVSD device closure.

well as the device's lesser stability. It is more challenging to achieve successful closure at younger ages and lower weights.

According to a report by Diandong et al., the most common abnormality in heart rhythm and conduction after perimembranous VSD device closure is incomplete right bundle-branch block. The incidence of persistent abnormality in rhythm or conduction is 4.5%. Although complete AV block is the most serious complication, its occurrence has gradually decreased to 1% in recent years.⁷ The occurrence of complete LBBB is of significance because it can cause abnormal contraction of the left ventricle, leading to deterioration of its function and potentially causing heart failure.⁸

Patients need to be monitored for recurrence and late onset of complete AV block and LBBB. In our case, the patient showed no signs of such complications during the follow-up.

4. Conclusions

Transcatheter therapy offers a safer alternative to surgery for selected infants with symptomatic perimembranous ventricular septal defects (pmVSDs) and low body weight. It reduces risks like morbidity and mortality associated with surgery. Key considerations include careful vascular access, choosing appropriate device sizes, and monitoring for heart rhythm issues post-procedure. Our case demonstrates the feasibility and benefits of this approach, emphasizing ongoing improvements in techniques and devices. Further research is needed to refine outcomes and expand its use in pediatric cardiology.

5. Ethical Approval

Not applicable.

We declare that the paper is not under consideration elsewhere and none of the paper's contents have been published previously.

6. Consent to Participate

Well-informed written consent was obtained from the patient and relatives before the procedure.

7. Source of Funding

No funding was received.

8. Conflict of Interest Statement

The authors declare no conflict of interest.

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