

Case Report

Double Intervention in A Patient with Large Patent Ductus Arteriosus and Moderate Upper Muscular Ventricular Septal Defect with Bidirectional Shunt with Severe Pulmonary Hypertension

Nurun Nahar Fatema¹, Priyanka Das²

¹Lab Aid Cardiac Hospital, Dhaka, ²Department of Pediatric Cardiology, Bangladesh Specialized Hospital

Abstract:

Patent ductus arteriosus (PDA) and Ventricular septal defect (VSD) are commonly encountered problems and constitute about 40% of all congenital heart lesions. The association of these two conditions in a single patient is not very uncommon. Both these conditions can be treated by placing intracardiac devices. Double interventional closure of PDA and VSD was performed in a gravely ill infant at her 3 months and 5 months of age respectively, who was suffering from recurrent pneumonia and heart failure since birth. These were lifesaving procedure for that patient which led to writing this report.

(*Cardiovasc j* 2024; 16(2): 105-109)

Key Words :
Patent Ductus Arteriosus, Ventricular Septal Defect, Pulmonary Hypertension.

Introduction:

A combination of PDA and VSD can lead to severe symptoms, which can be manifested by shock, and subsequently heart failure. Patent ductus arteriosus is associated with VSD in 1–7% of cases.

Multiple cardiac defects are not rare in children with congenital heart disease (CHD). Historically, these were usually addressed with surgery. Although conventional open-heart repair with cardiopulmonary bypass (CPB) has been considered the standard treatment, recurrent respiratory tract infection and congestive heart failure are still risk factors for an operative procedure, and the long midline incisions might lead to increased postoperative physical and psychological complications.^{1,2} However, CPB scope is limited in infants because of their very young age and low weight. With the advent of newer technologies and expertise, transcatheter treatment has evolved as an alternative option for infants with multiple shunt anomalies.

Case Report:

A 46 days old female baby weighing 2.8 kg, presented with severe respiratory distress, and difficulty in feeding since birth, and was referred to our center for evaluation. Clinical examination showed the child was ill-looking, pale, cyanotic, dehydrated, dyspneic, and tachypneic, with visible bilateral chest indrawing, and clearly audible long systolic murmur.

Chest x-ray showed cardiomegaly, with the plethoric lung fields and electrocardiography (ECG) showed right axis deviation with Right ventricular hypertrophy.

Echocardiography showed, a large sized PDA with bidirectional shunt, Moderate sized VSD with bidirectional shunt, and severe pulmonary hypertension (PAH). So patient was planned for urgent PDA closure first. Case was refused by Cardiac surgeon for low body weight, so the patient was accepted as a high-risk case for PDA device closure.

Address Correspondence: Prof. Dr. Nurun Nahar Fatema, Congenital and structural Interventionist, Lab Aid Cardiac Hospital, Dhaka, Bangladesh. Email: colfatema@hotmail.com

© 2024 authors; licensed and published by International Society of Cardiovascular Ultrasound, Bangladesh Chapter and Bangladesh Society of Geriatric Cardiology. This is an Open Access article distributed under the terms of the CC BY NC 4.0 (<https://creativecommons.org/licenses/by-nc/4.0>)

After admission, the patient was stabilized with high-flow oxygen, her hydration was maintained by Intravenous fluid and relevant investigations were sent. Injectable antibiotics were added to treat infection. After initial resuscitation for 2 weeks, the patient was taken into the Cath lab on 9th December 2022 for PDA device closure.

Procedure:

Hardwires: Standard pediatric drape, Jensini catheter, Pigtail catheter, Bherman balloon catheter, Tyshak-mini balloon, Piccolo occluder, MFO-Konar device, 4F long delivery system, exchange wire, PTCA all-star wire, etc.

Aortogram: The Aortogram showed a large PDA which filling the branch PA's. LV angiogram showed smooth walled poorly contractile posteriorly located LV ejecting to AO. There was a Moderate upper muscular VSD with reverse shunt from right ventricle to left ventricle.

While the exchange wire crossed through the PDA, the patient developed Brady event and desaturated. We changed our strategy and PDA device closure was performed from a retrograde approach with MFO-Konar 08 x 06 mm device.

Follow-up: Following PDA device closure the baby was kept in NICU for further management, few hours after PDA device closure, the patient's distress went down and gradually improved in 2 days, she was discharged on the 3rd post-procedure day with anti-failure and pulmonary vasodilators.

Six days later she was admitted again to the hospital with severe respiratory distress. On examination, the child was ill-looking, pale, cyanotic, dehydrated, dyspneic, and tachypneic, with visible bilateral chest indrawing.

Chest x-ray showed –Sign of aspiration pneumonia
Echocardiography showed, a PDA device in Situ and Moderate sized Ventricular Septal Defect (VSD) with reverse shunt, and severe pulmonary hypertension (PAH).

The patient was resuscitated and stabilized in PICU and treated with broad-spectrum antibiotics, fluid electrolyte glucose balance was maintained, and nutrition was maintained through NGT with high-calorie milk. Within a few

days, the patient's condition improved and discharged successfully.

She again got admitted after 2 weeks with aspiration pneumonia and respiratory arrest. Again she was resuscitated; she was admitted and was kept on observation for weight gain, through NG tube feeding. We planned for VSD device closure at her 5 months of age. She was taken into the Cath lab on 16.02.23 for VSD device closure.

Hardware: Standard pediatric drape, pigtail catheter, Gensini catheter, Multifunctional occlude (MFO-Konar), JR guide catheter, Terumo exchange wire.

LV angiogram: Showed moderate upper muscular VSD.

Procedure: An 8×6 MFO-Konar device was implanted through a retrograde approach.

The patient was discharged after 2 days of the procedure and advised to come for a follow-up after 1 month with CXR, ECG, and Echo.

So far, a 6-month follow-up of the patient is completed and she is doing fine, her pulmonary artery pressure is still at a mildly raised level of 35mmHg. She is gaining weight and episodes of respiratory infection are minimum in last few weeks.



Figure 1: Baby M before intervention.



Figure 2: Baby M at 6 month follow up.



Figure 3: CXR after aspiration pneumonia.

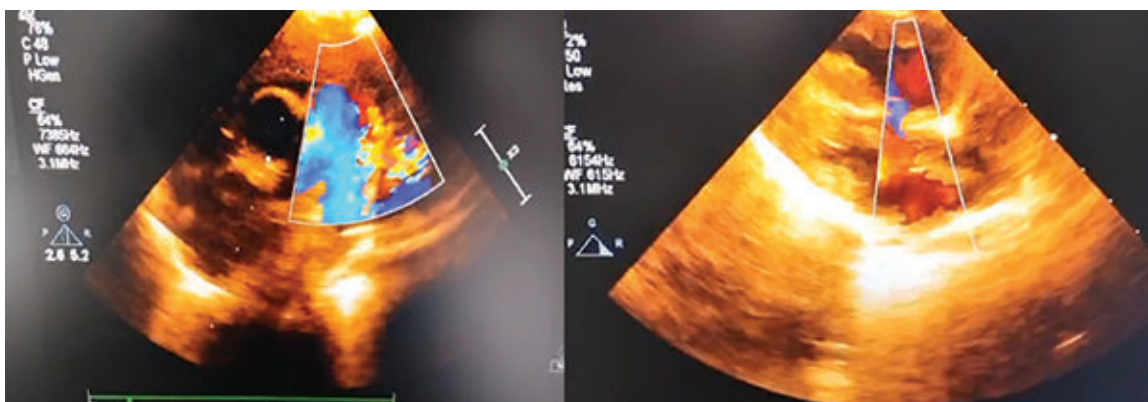


Figure 4: Echocardiography showing PDA and VSD.

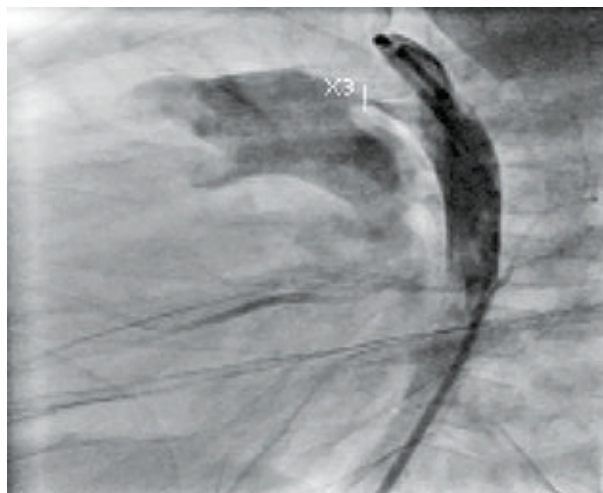


Figure 5: Aortogram showing large Tubular PDA.



Figure 6: LV angiogram showing Moderate upper muscular VSD.

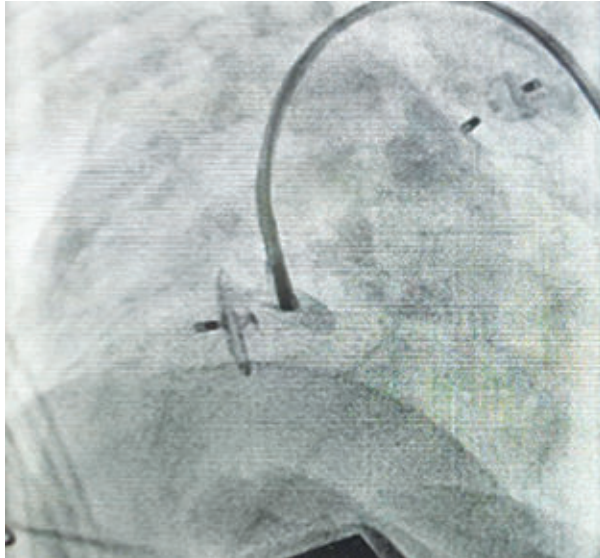


Figure 7: Both PDA and VSD devices are visible.

Discussion:

By now transcatheter closure for PDA has been established as the first choice therapy in many centers.³ Despite the advancement of the procedure, there are limitations in the closure of small children. The procedure was initially recommended only for children weighing over seven to eight kilograms because the size of the femoral vessels was relatively smaller than the delivery system.^{4,5} Few studies investigated transcatheter closure in preterm, neonates and in children less than 10 kg.⁶ Transcatheter closure of PDA is a minimally invasive therapy associated with a low rate of adverse effects in the neonate and early infancy.⁷ In premature infants it was never tried earlier for fragility and concern regarding arterial/venous access. Suitable devices appropriate for neonatal PDA anatomy were also not available.⁸ But recently it has been established that it may be performed in newborns too.⁹ A study showed the incidence of PDA in preterm is 16 times more than in full-term infants.¹⁰ Delayed closure of hemodynamically significant PDA in neonates leads to many complications.¹¹ Transcatheter closure is a better option for such cases and surgical risk can be avoided.

Ventricular septal defect is the most common congenital heart disease accounting for up to 30% of all CHD.^{12,13} Moderate to large VSD presents with features of increased pulmonary blood flow, failure to thrive, and left heart dilatation during

infancy. The large VSD with Pulmonary hypertension is a common indication for surgical intervention. The recent success of the transcatheter intervention for VSD with different types of devices has expanded the scope for the device closure. The device closure of per membranous VSD in young children is challenging and is prone to complications. Recently the Konar-MF occluder (Lifetech Inc., Shenzhen, PRC) was introduced as a newer, softer, double disc soft device. It can be deployed from both sides.^{14,15}

Large VSD in the early infancy is treated by surgical closure. The last two decades saw the growing popularity of device closure of the VSD in the relatively bigger kids. The traditional indication for the device closure of VSD is for larger patients with a weight of around 10 kg. However, some studies show that VSD can be closed in infancy when it is very large causing failure to thrive, heart failure, and other complications.^{14,15} The presence of hemodynamically significant PDA and VSD imposes a double threat of heart failure, and pulmonary hypertension to the patient. Recent studies showed excellent outcome in young infants after device closure.^{16,17}

In this case patient had recurrent pneumonia, failure to thrive, heart failure and severe pulmonary hypertension due to Large PDA and Moderate VSD. Ultimately both PDA and VSD were closed and the patient was cured completely.

Conclusion:

Combination of VSD and PDA is a common finding in early cases. If these defects are hemodynamically significant, they need to be closed. Surgical closure is of very high risk in young infants due to recurrent pneumonia, lower body weight, severe pulmonary hypertension, and heart failure. So, percutaneous closure is a better option which has been proved in this critical case. Conflict of interest: None.

References:

1. Rose EA. Off-pump coronary-artery bypass surgery. *N Engl J Med.* 2003;348(5):379-380. doi:10.1056/NEJMp020171
2. Gessler P, Schmitt B, Pre'tre R, et al. Inflammatory response and neurodevelopmental outcome after open heart surgery in children. *Pediatr Cardiol* 2009; 30: 301-305. doi: 10.1007/s00246-008-9354-5

3. Ewert P. Challenges encountered during closure of patent ductus arteriosus. *Pediatr Cardiol.* 2005;26(3):224-229. doi:10.1007/s00246-005-1010-8
4. Butera G, De Rosa G, Chessa M, et al. Transcatheter closure of persistent ductus arteriosus with the Amplatzer duct occluder in very young symptomatic children. *Heart.* 2004;90(12):1467-1470. doi:10.1136/hrt.2003.025122
5. NNF Begum, AU Ahmed. Acute Intra Vascular Haemolysis after Coil Occlusion of Patent Ductus Arteriosus by Detachable Coil - A Case Report. *Journal of BCPS;* 2003;21:151-153.
6. N N Fatema. Device Closure of a Large Patent Ductus Arteriosus with Severe Pulmonary Hypertension: A Case Report. *Bangladesh Heart J* 2005;20(2):73-76.
7. Krichenko A, Benson LN, Burrows P, Möes CA, McLaughlin P, Freedom RM. Angiographic classification of the isolated, persistently patent ductus arteriosus and implications for percutaneous catheter occlusion. *Am J Cardiol.* 1989;63(12):877-880. doi:10.1016/0002-9149(89)90064-7
8. Kumar SM, Subramanian V, Bijulal S, Krishnamoorthy KM, Sivasankaran S, Tharakan JA. Percutaneous closure of a moderate to large tubular or elongated patent ductus arteriosus in children younger than 3 years: is the ADO II appropriate?. *Pediatr Cardiol.* 2013;34(7):1661-1667. doi:10.1007/s00246-013-0700-x
9. Park YA, Kim NK, Park SJ, Yun BS, Choi JY, Sul JH. Clinical outcome of transcatheter closure of patent ductus arteriosus in small children weighing 10 kg or less. *Korean J Pediatr.* 2010;53(12):1012-1017. doi:10.3345/kjp.2010.53.12.1012
10. Jang GY, Son CS, Lee JW, Lee JY, Kim SJ. Complications after transcatheter closure of patent ductus arteriosus. *J Korean Med Sci.* 2007;22(3):484-490. doi:10.3346/jkms.2007.22.3.484
11. Pruetz JD, Carroll C, Trento LU, et al. Outcomes of critical congenital heart disease requiring emergent neonatal cardiac intervention. *Prenat Diagn.* 2014;34(12):1127-1132. doi:10.1002/pd.4438
12. Hoffman JI, Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol.* 2002;39(12):1890-1900. doi:10.1016/s0735-1097(02)01886-7
13. Tanidir IC, Baspinar O, Saygi M, Kervancioglu M, Guzeltas A, Odemis E. Use of Lifetech™ Konar-MF, a device for both perimembranous and muscular ventricular septal defects: A multicentre study. *Int J Cardiol.* 2020;310:43-50. doi:10.1016/j.ijcard.2020.02.056
14. Tao K, Lin K, Shi Y, et al. Periventricular device closure of perimembranous ventricular septal defects in 61 young children: early and midterm follow-up results. *J Thorac Cardiovasc Surg.* 2010;140(4):864-870. doi:10.1016/j.jtcvs.2010.05.013
15. Thakkar B, Patel N, Shah S, et al. Periventricular device closure of isolated muscular ventricular septal defect in infants: a single centre experience. *Indian Heart J.* 2012;64(6):559-567. doi:10.1016/j.ihj.2012.09.006
16. Nurun Nahar Fatema. Transcatheter closure of ventricular septal defect from retrograde transarterial approach: immediate and long-term outcome. *International journal of contemporary pediatrics.* 2020;7(9):1830-1830. doi:https://doi.org/10.18203/2349-3291.ijcp20203639
17. NN Fatema. Device Closure of Large Patent Ductus Arteriosus (PDA) in Young infants. Analysis of Cases in a Bangladeshi Centre. *Insight in pediatric cardiology* 2020; S (1): 1. DOI: 10.36648/IPIPC.6.4.35