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Case Report

How to Recognize and Overcome Pulmonary Hypertension Crisis During Patent Ductus Arteriosus Closure by Device in Adult

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ABSTRACT

Background: Managing a patent ductus arteriosus (PDA) with severe pulmonary hypertension (PH) is challenging since closing the PDA can result in reduced cardiac output and right ventricular (RV) failure. The latest guideline for Adult Congenital Heart Disease (ACHD) stated it is harmful to close the defect in patients with pulmonary vascular resistance (PVR) ≤5 WU and flow ratio (FR) <1.5.
Case presentation: A 37-year-old female was referred with cough, fever, low saturation, and murmur findings. After serial examination, she was diagnosed with large PDA and severe PH her peripheral saturation (SaO_2) was 88%-89% and non-responder to acute vasoreactivity test. After a year of PH therapy, vasoreactivity showed a response to the vasoreactivity test and improved on clinical presentation with SaO_2 91%-92%. Patient fell in to PH crisis condition during the procedure of device closure, prostacyclin analogue intravenous (IV) and phosphodiesterase inhibitors inhalation was administered, and the procedure was carried on. The device was successfully implanted, and the patient had SaO_2 97% in all four extremities before discharge.
Conclusion: With established PH therapy, PDA with severe PH can underwent PDA closure by device with satisfying outcome.

1. Introduction

The main challenge in treating a large PDA is that it might cause volume and pressure overload on the pulmonary vascular bed. Although PH can be reversed with prompt shunt removal, PH may become irreversible if PDA is not identified early on. To protect the pulmonary vascular bed from prolonged pressure and volume overload and prevent Eisenmenger syndrome from developing, shunt closure is essential. When PH is both high-flow and high-resistance, closing the PDA may result in right ventricular failure and poor cardiac output which can lead to PH crisis. Closure of the defect should wait until PH treatment has been established. Device closure is the preferred methods as it has low risk of complications in most cases. One way to help during device closure procedure determine whether to move on with defect closure is to use an occlusion test before removing the device.

2. Case Report

A 37-year-old female with cough, fever, low saturation, and murmur findings. During her childhood, she had poor weight gain but was otherwise healthy. She was underweight with a Body Mass Index (BMI) of 15.6 kg/m2. Her SaO_2 was 88%-89% and blood pressure (BP) of 144/67/96 mmHg. Physical examination revealed loud and palpable second heart sound with pansystolic murmur at left infraclavicular area grade 3/6. Echocardiography showed a PDA with bidirectional shunting dominant Left (L) to Right (R) and mild tricuspid regurgitation with high probability of PH, biventricular systolic function was normal and no other anatomical anomalies were identified. Cardiac catheterization revealed a significant PH with non-responder to vasoreactivity test. It was decided not to close the PDA instead, the patient was started on medication for PH, which included sildenafil, spironolactone, and bisoprolol. After one year of medical treatment the patient's symptoms improved, especially in terms of increased tolerance to exercise. Her SaO_2 was improved to 91%-92%, and was reactive of vasoreactivity testing with 100% FiO2. (see Table 1.) She was then registered for PDA Closure by Device.

Intervention Procedure

Using femoral access with local anesthesia, measurements of intracardiac pressure and saturation was taken. Baseline Aorta (Ao) pressure before the procedure was 120/62/87 mmHg and PA pressure was 102/45/69 mmHg with PA saturation of 66% and Ao saturation of 90%. Angiography showed a large PDA Krichenko type A, with ampulla measuring 14.3 mm and the narrowest point measuring 12.1 mm. PDA Occluder Memopart (Lepu) 18-20 mm was selected. To deliver the occluder we use a 10F delivery seath and through the Amplatz wire, crossing from the PA to the descending aorta (AoD) over the PDA. As the Amplatz wire crossed the PDA, PA pressure abruptly increased to 117/54/80 mmHg, the patient complaints of shortness of breath, desaturation with SaO_2 83%, hypotension, and bradycardia. The patient fell on PH crisis and cardiac arrest. Chest compression was performed and after the return of spontaneous circulation, 100% FiO2 with non-rebreathing mask, milrinone iv and iloprost inhalation was delivered as

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Table 1. Hemodynamic data.

Parameter	1st RHC (November 2021)	1st RHC + FiO2 100%	2nd RHC (March 2023)	2nd RHC + FiO2 100%
Hemoglobin (g/dL)	14.3	14.3	14.2	14.2
RV Pressure	145/2	138/7	104/14	N/A
LV Pressure	136/4	145/2	137/12	108/17
Art Pressure	140/67/95	138/64/96	122/61/87	113/51/79
LPA Pressure	137/59/92	135/60/87	118/55/83	107/46/68
PCWP (mean) (mmHg)	4	2	12	17
Arterial SpO2 (%)	84	97	91	97
Qp:Qs	0.58	2.83	1.1	3.0
PVR index (WU/m2)	37.4	15.3	12.3	6.7
Rp:Rs	1.46	0.34	0.47	0.2
Event during procedure	PVC and NSVT		PVC	

Art: arterial; Dia: diastolic; LV: left ventricular; LPA: left pulmonary artery; PVR index: Pulmonary vascular resistance index; PCWP: pulmonary capillary wedge pressure; RV: right ventricle; Sys: systolic; RHC: Right Heart Catheterization.

inotropic and vasodilator. The patient was stabilized with SaO_2 100%, BP 114/65/83 mHg, HR 70 bpm, and PA pressure decreasaed to 65/26/45 mmHg, the procedure was then continued. The device was successfully deployed with no residual shunt and no obstruction at the AoD after evaluation with aortography. Occlusion test was performed to ensure no negative hemodynamic side effects. As we monitored, PA Pressure was decreased to 64/24/42 mmHg below the Aorta Pressure of 129/80/99 mmHg. The patient remains stable with a SaO_2 of 91%-92% and no complaint of shortness of breath or faintness. The patient was then monitored for 2 days at the intensive cardiac unit. PH therapy was continued with milrinone iv continuous drip, iloprost inhalation 3 mcg every 4 hours, oral Sildenafile 3 x 20 mg, Beraprost 2 x 20 mg, and spironolactone 1 x 25 mg. The patient was discharged with SaO_2 of 97%, stable hemodynamics, no cyanosis, and no complaints of dyspnea or residual murmur. The patient remained asymptomatic with SaO_2 of 98%–99% at the 6-month follow-up. This result, along with the data previously discussed, supported our conclusion that, in PDA patients with severe pulmonary hypertension, device closure after established PH management can result in favorable patient outcomes.

3. Discussion

Studies have highlighted the harmful effects of PH on the mortality of ACHD. A comprehensive approach involves multiple stages of evaluation and management, integrating medical treatment, echocardiography, and invasive cardiac catheterization to address PH with a large PDA.[1-3] This approach not only assesses the safety of ductal closure but also uses medical therapy to ensure a secure PDA occlusion. Hemodynamic parameters, including PVR and ratio of the pulmonary-to-systemic flow (Qp: Qs), are also considered. In our patient's case, with a PVR ≥5 WU and Qp: Qs >1.5, a cautious individualized decision is necessary, particularly in expert centres, with a class IIb recommendation from European guidelines of ACHD 2020. Device closure is the preferred method in adults and can be safely performed with a low risk of complications in most cases, even when concurrent cardiac procedures are needed. Surgical intervention is reserved for a select few patients with ducts too large for device closure or anatomical complications like aneurysm development.[4–6] Initially, this patient exhibited no significant decrease on mPAP after vasoreactivity testing. After a year of medical intervention, the clinical presentation improved, with better tolerance to activity, SaO_2 91%, and responsive reaction to vasoreactivity testing with 100% FiO2 was noted. However, despite the success of PH medication in regulating PVR, mPAP remained systemic before the closure of the large PDA. Mortality perioperative was primarily attributed to the frequent occurrence of PH crisis. This condition arises when the mPAP suddenly exceeds the mSAP, causing abrupt pulmonary vasoconstriction, systemic hypotension, and right heart failure, potentially leading to fatal tissue hypoxia.[2,7] The development of PH crisis is associated with endothelial cell damage, systemic inflammatory response syndrome, elevated endothelin levels, and suppression of nitric oxide (NO) generation. To address acidosis and hypoxia, meticulous management of inotropes, vasopressors, adequate fluid balance, and maintaining sinus rhythm and atrioventricular synchrony are critical.[7,8] PH crisis often requires an aggressive combination of therapies for RV failure. Inhaled nitric oxide (iNO) is a standard therapy for post-operative control of PH and prevention of PH crisis. iNO is initiated at doses of 5-20 ppm, with the possibility of increasing to a maximum of 80 ppm within minutes. Its crucial to administer the lowest effective dose to minimize toxicity and gradual withdrawal from iNO are essential to enhance the utility of iNO and avoid the risk of rebound pulmonary hypertension. While iNO may have limitations, such as cost, complex delivery systems, and delays in administration during acute crises, alternative therapies should be considered.[7,9] Prostacyclin and its analogs, including alprostadil, epoprostenol, treprostinil, and iloprost, represent newer pulmonary-specific vasodilators. Iloprost, in particular, reducing the risk of systemic vasodilation by selectively targets ventilated lung segments. Inhaled iloprost may offer a favorable safety profile compared to iNO, as it can be easily administered by inhalation without requiring a complex delivery system. Another therapeutic alternative includes endothelin receptor antagonists (ERAs), which target endothelins contributing to PAH pathobiology through interactions with endothelin receptors (ETRs). [7,8]

4. Conclusion

When faced with severe PH in ACHD, a thorough assessment must be done and measurements to prevent a PH crisis must be taken. Closure of CHD shunts should be done under the umbrella of established oral PAH therapy. PH crisis usually presents with worsening of shortness of breath, cyanosis, desaturation, and hypotension or shock condition. Early recognition of this condition is paramount as well as administration of PH crisis therapy including oxygenation, addressing acidosis, adequate fluid balance, and maintenance of sinus rhythm and atrioventricular synchrony. With established PAH therapy before closure and occlusion test during the device closure procedure, satisfying outcome can be achieved.

5. Declaration

5.1 Ethics Approval and Consent to participate
Patient has provided written informed consent prior to involvement in the study.

5.2. Consent for publication
Not applicable.

5.3 Availibility of data and materials
Data used in our study were presented in the main text.

5.4 Competing interests
Not applicable.

5.5 Funding Source
Not applicable.

5.6 Authors contributions

Idea/concept: HM. Design: HM. Control/supervision: HM. Data collection/processing: LUK Analysis/interpretation: LUK. Literature review: HM. Writing the article: LUK. Critical review: HM. All authors have critically reviewed and approved the final draft and are possible for the content and similarity index of the manuscript.

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