Open Access

Case Report

Alternative Approach for Pulmonary Valvuloplasty in Critical Congenital Pulmonary Stenosis

Andaleeb Ara, Khurram Akhtar, Saima Rafique

Armed Forces Institute of Cardiology/National Institute of Heart Diseases (AFIC/NIHD)/National University of Medical Sciences (NUMS) Rawalpindi Pakistan

ABSTRACT

Critical congenital pulmonary stenosis with intact interventricular septum is a cyanotic and potentially lethal neonatal cardiac anomaly. Percutaneous transcatheter balloon pulmonary valvuloplasty for critical pulmonary stenosis often presents technical difficulties due to supra-systemic pressure in right ventricle. Using transjugular approach instead of the femoral vein helps in dealing with this issue. We describe the successful use of right internal jugular vein approach in a case of critical pulmonary stenosis.

Keywords: Critical pulmonary stenosis, pulmonary valvuloplasty, Trans jugular approach.

How to Cite This Article: *Ara A, Akhtar K, Rafique S. Alternative Approach for Pulmonary Valvuloplasty in Critical Congenital Pulmonary Stenosis. Pak Armed Forces Med J 2022; 72(Suppl-3): S648-650. DOI: https://doi.org/10.51253/pafmj.v72iSUPPL-3.9574*

INTRODUCTION

Pulmonary stenosis (PS) accounts for approximately 7% of patients having congenital heart defect.[1] It may occur as an isolated lesion or in association with other cardiac defects like ventricular septal defect, tetralogy of Fallot, tricuspid atresia, double outlet right ventricle (RV), and transposition of great arteries.[2] Critical PS presents with cyanosis in the newborn if the ventricular septum is intact. Right to left shunting at foramen ovale causes cyanosis because of compromised compliance of the RV because of ventricular hypertrophy. In majority of these cases percutaneous balloon pulmonary valvuloplasty is considered the procedure of choice.[3] Studies have described percutaneous balloon valvuloplasty to have immediate success with re-intervention requirement in fewer cases along with reasonable growth of the RV and that of the pulmonary valve in infants with critical pulmonary valve stenosis.[4,5] However, the procedure often presents technical difficulties because of increased pressure in the RV making it difficult to reach the stenotic pulmonary valve. To overcome this issue certain methods have been devised and used around the world like using a catheter with bent hard end of a guide wire and using trans-jugular approach instead of the routinely used femoral vein.[6] However there is reluctance among the operators due to the possibility of complications and little availability of supportive data.[7] We present a case report in which we used the trans-jugular approach using the right IJV with relative ease convenience of the procedure.

Correspondence: Dr Andaleeb Ara, Peads Cardiology Department, Armed Forces Institute of Cardiology/National Institute of Heart Diseases (AFIC/NIHD) Rawalpindi, Pakistan

CASE REPORT

Current case report is of a 6 months old baby boy, weighing 7 kg who presented to us in severe respiratory distress and cyanosis. The baby had been diagnosed with critical PS at 4 months of age but they could not seek timely treatment despite the diagnosis. On examination the infant was cyanosed with facial plethora and was severely dyspnoeic with a respiratory rate of 60 per minute. There was a faint ejection systolic murmur on auscultation. Oxygen saturation (SpO_2) fluctuated between 35% and 40% on arrival. Echocardiography revealed dilated RV, severe tricuspid regurgitation (TR) with pressure gradient of 100 mmHg and minimal flow across a hypoplastic and doming pulmonary valve. The pulmonary valve annulus measured 5mm but there was just a streak of flow on color Doppler, a small patent ductus arteriosus (PDA) shunting left to right and persisting foramen ovale (PFO) shunting right to left. The high grade of TR and direction of shunt at PFO fulfilled the diagnosis of critical PS. Baby was admitted and stabilized by instituting oxygen therapy with bubble continuous positive airway pressure (CPAP) and prepared for emergency transcatheter balloon pulmonary valvotomy. After general anaesthesia, right femoral vein was accessed with 5F short sheath. After multiple attempts (due to high right ventricular pressure) RV was entered with 4F JR and RV angiogram done in LAO 15 CR 30 which revealed severe pulmonary valve stenosis and hypoplastic pulmonary arteries (Figure-1A). However, positioning the catheter for crossing pulmonary valve was the real challenge, as it would keep jumping out of RV cavity. Stiffer 5F JR was then used but did not improve the outcome. Access site was then changed to right internal jugular vein (IJV) and 5F

JR used to successfully and easily enter the RV cavity. Right IJV approach provided better stability in crossing the pulmonary valve as it would hook into the infundibulum and would not jump out easily. Graduated technique was used for ballooning with 7x30mm and 9x20mm VACS II balloons over 0.014" guide wire successfully relieving the obstruction (Figure-1B, C). Post procedure saturations improved from 40% to 90% and baby was discharged the next day.

Figure-1 (A-C): A): Angiogram in LAO 15 Cr 30, showing severe Pulmonary valvular stenosis and Hypoplastic Pulmonary arteries. B) i. Vacs ii Balloon (7x30 mm) over 0.014" guide wire showing waist at the level of pulmonary valve. C): Ballooning repeated with a larger balloon (9x20 mm) with disappearance of waist

DISCUSSION

Critical PS in newborns presents with marked cyanosis and early intervention is required to ensure pulmonary blood flow.[8] Prostaglandin E1 infusion is administered for initial stabilization once the diagnosis is established on 2D echocardiography.[7] Percutaneous transcatheter balloon pulmonary valvuloplasty is considered to be the treatment of choice these days which can be done in the first week of life.[9]

Balloon pulmonary valvuloplasty in critical PS is technically challenging because of supra systemic RV pressures.[10,11] The presence of severe tricuspid regurgitation and dilated right heart make catheter handling difficult. The main challenge during the procedure is accessing the RV cavity as the tricuspid valve is very difficult to cross due to high RV pressures. To achieve this various methods have been devised and used around the world like using a catheter with bent hard end of a guide wire or adopting a trans-jugular approach instead of the femoral vein. Hoetama *et al.* described transjugular approach used in 8 neonates for balloon pulmonary valvuloplasty and found it safe to use in cases of critical PS.[6] Right ventricular/systemic pressure ratio is generally around 150%. Once the tricuspid valve is crossed, the next challenge is to cross the stenotic pulmonary, which needs stable positioning of the catheter in the right ventricular outflow tract (RVOT). In our setup we use 4F/5F JR catheters, but even the slightest movement combined with high RV pressures expels the catheter out when right femoral venous access is used. To cater for that, the right internal jugular approach is used, which allows the JR to hook into the RVOT, and thus having to cross the tricuspid repeatedly is taken out of the equation, substantially decreasing procedural and fluoroscopy times. Transjugular approach reduced the procedure and fluoroscopy times because the route from superior vena cava to RV is relatively straighter as compared to the route from the inferior vena cava (IVC) to RV. Also only one curve is needed to cross pulmonary valve (RV to PA) in transjugular approach, whereas in transfemoral approach, making two curves are essential i.e. from IVC to RV and from RV to PA. This was confirmed by Hoetama *et al* who reported a significant decrease in procedural time and fluoroscopy time in the transjugular approach as compared to transfemoral approach (65±8 vs. 108±17.8 min, $p<0.05$ & 29±13 vs. 67±35 min, $p<0.05$).[6]

CONCLUSION

Transjugular approach provides a useful and more efficient alternative to transfemoral approach for balloon pulmonary valvuloplasty in critical PS.

ACKNOWLEDGEMENT

I am deeply grateful to my supervisor for his guidance, patience and support who provided insight and expertise that greatly assisted my research project. I also want to share my gratitude for COMDT Exec Dir AFIC/NIHD and HOD R&D for their support and contribution in completion of the research paper

Conflict of Interest: None.

Author's Contribution

Following authors have made substantial contributions to the manuscript as under:

AA: Manuscript writing, concept and manuscript drafting

KA: Intellectual contribution, concept and final approval

SR: Proof reading, Intellectual contribution, final approval

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

REFERENCES

1. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS. AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. J Am Coll Cardiol 2019; 73(12): e81-e192.

2. Coffey S, Rayner J, Newton J, Prendergast B. Right-sided valve disease. Int J Clin Pract 2014; 68(10): 1221-1226.
3. Latson LA. Critical pulmonary stenosis. J Interv Cardiol 2001; 14(3): 345-350.
4. Saad MH, Roushdy AM, Elsayed MH. Immediate-and medium-term effects of balloon pulmonary valvuloplasty in infants with critical pulmonary stenoses during the first year of life: A prospective single center study. J Saudi Heart Assoc 2010; 22(4): 195-201.
5. Yin D, Wu X, Xiang P, Zhang Y, Tian J, Lv T, et al. Outcomes of percutaneous balloon pulmonary valvuloplasty in congenital pulmonary valve stenosis. Clin Case Rep 2021; 9(9): e04705.
6. Hoetama E, Prakoso R, Roebiono PS, Sakidjan I, Kurniawati Y, Siagian SN, et al. Balloon pulmonary valvuloplasty in neonates with critical pulmonary stenosis: Jugular or femoral. Ann Pediatr Cardiol 2020; 13(1): 11-15.
7. Mortezaeian H. Percutaneous balloon pulmonary valvuloplasty of critical pulmonary stenosis and severe pulmonary stenosis in neonates and early infancy: a challenge in the cyanotic. J Cardiovasc Thorac Res 2021; 13(2): 156-160.
8. Khalil M, Jux C, Rueblinger L, Behrje J, Esmaeili A, Schranz D. Acute therapy of newborns with critical congenital heart disease. Transl Pediatr 2019; 8(2): 114-118.
9. Zeevi B, Keane JF, Fellows KE, Lock JE. Balloon dilation of critical pulmonary stenosis in the first week of life. J Am Coll Cardiol 1988; 11(4): 821-824.
10. Guccione P, Gagliardi MG. Cardiac catheterization through the internal jugular vein in pediatric patients: an alternative to the usual femoral vein access. Chest 1992; 101(6): 1512-1514.
11. Joseph G, Kumar KP, George PV, Dhanawade S. Right internal jugular vein approach as an alternative in balloon pulmonary valvuloplasty. Catheter Cardiovasc Interv 1999; 46(1): 425-429.