Case report

Bridge to surgery with pulmonary valve perforation and right ventricular outflow tract stenting in an infant with PA-VSD

Abstract

Pulmonary atresia (PA) with ventricular septal defect (VSD) is a rare congenital heart disease. Blood flow to pulmonary arteries is provided by patent ductus arteriosus or major aorto-pulmonary collateral arteries (MAPCAs). Palliative interventions are required to maintain the blood supply to the pulmonary arteries in order to achieve for the complete surgical repair. In this case report, we present a patient who underwent pulmonary valve perforation and right ventricular outflow tract (RVOT) stenting as an alternative to a systemic-pulmonary artery shunt (SPS). Subsequently, surgical repair was performed ten months later.

Key words: Pulmonary valve perforation, Pulmonary atresia, RVOT stenting, Tetralogy of Fallot

1. INTRODUCTION

Pulmonary atresia with ventricular septal defect (PA/VSD) is a variant of tetralogy of Fallot (TOF) and management of this cyanotic congenital heart disease is still challenging. Its difference from TOF is the luminal discontinuity between the right ventricle and pulmonary arteries. Therefore, pulmonary blood flow is provided by extracardiac sources such as ductus arteriosus, aorto-pulmonary collateral, or systemic-pulmonary shunts (SPS). Patients with PA/VSD and severely hypoplastic pulmonary arteries require an intervention in the early infancy period to relieve cyanosis and to maintain blood supply to the pulmonary arteries. Palliative procedures reported to date such as central shunts, modified Blalock-Taussig shunts (mBTs), right ventricular outflow tract (RVOT) reconstruction or stenting and ductal stenting can be associated with complications such as occlusion and distortion of the pulmonary architecture [1,2]. With the development of catheter-based intervention techniques, safer procedures can be performed to provide pulmonary blood flow as an alternative to surgical SPS. Here, we would like to report a case that pulmonary valve perforation and, RVOT stenting could be performed as an alternative intervention to surgical palliative procedures for infants with PA/VSD.

2. CASE PRESENTATION

A 2-month-old infant weighing 2250 g was admitted to our clinic with the diagnosis of PA / VSD. On physical examination, there was a 2/6 continuous murmur heard best in the left second intercostal space on auscultation. Her resting oxygen saturation was 70% and other systems examination findings were within normal limits. Electrocardiography (ECG) revealed signs of right axis deviation and right

ventricular hypertrophy. Telecardiography showed mild cardiomegaly and, decreased pulmonary vascularization. Transthoracic echocardiography revealed malalignment large subarterial VSD, aortic overriding and membranous pulmonary atresia with good-sized confluent pulmonary arteries. Vertical type, medium-width patent ductus arteriosus was the sole supply of the pulmonary blood flow (Fig. 1).

Catheterization was planned to attempt ductal stenting or pulmonary valve perforation. Under general anesthesia, 5F sheath was placed in the femoral vein, and a 4F sheath was placed in the femoral artery with a percutaneous technique. Heparin (75 U/kg) was administered after the sheaths were placed in the groin. A descending thoracic aortogram showed no MAPCAs. With the 5.2F Judkins right (JR) coronary catheter (Cooksmedical, Bloomington, USA), a right ventriculogram was performed in the left anterior oblique with cranial angulation and 90° lateral projection. It was observed that the trabeculated right ventricle was filled, and contrast material passed to the left ventricle and aorta via the VSD. Although stenosis was observed in the RVOT starting from the subvalvular region, the pulmonary valve was atretic, and minimal contrast material passed to the pulmonary artery (Fig. 2 a-b). Hence, the JR was attempted to be passed through the valve with the help of a 0.035" hydrophilic guidewire, but it was unsuccessful. While the JR catheter was positioned in the RV outflow tract and centered on the atretic pulmonary valve membrane the location of the catheter was checked with small hand contrast injections. Simultaneous injections performed from the pulmonic side via a catheter, which was advanced through PDA, might describe the positions of the catheters. When the position of the JR catheter was optimal the pulmonary valve was perforated by pushing forward the Conquest Pro CTO (Asahi Intecc Co. Ltd., Aichi, Japan) guidewire (Fig. 2 c-d). The atretic valve was attempted to be perforated with the help of a CTO wire; in the second attempt, the valve was successfully perforated. A secondary 0.014'' extra stiff wire was sent through the JR catheter and taken forward into the right pulmonary artery. Once the pre-dilatation was performed with a 2x15 mm coronary balloon (Alvimedica Invader™ PTCA) over the congest pro wire (Fig. 2 e-f), followed by an implantation of 6x15 mm renal stent (RX Herculink Elite®) at the RVOT to include the pulmonary valve (Fig. 2 g-h). The stent's diameter was selected to be +/-1 mm of the MPA as measured just above the atretic pulmonary valve.

Finally, a right ventriculogram was performed and it was demonstrated the patency of right ventricular outflow tract. The procedure was terminated without any complications. When the patient was ten months old, catheterization was performed and she was referred to surgery for a complete correction operation and the surgery was done uneventfully (Fig. 3 a-b).

3. DISCUSSION

Management of pediatric cases with PA/VSD is still very challenging. Establishing pulmonary blood flow, to minimize hypoxia and ensure adequate pulmonary arterial growth are crucial in these patients.

Therefore, early intervention is commonly required, especially in patients with hypoplastic pulmonary arteries. The management options and prognosis, depends on the development of the pulmonary artery segments and its branches. The aim in the early period is; providing blood flow to the pulmonary artery in order to ensure their development [3]. For this purpose, surgical or cardiac catheterization intervention may be planned. Surgically; SPS are traditionally used in congenital heart diseases with ductal-dependent pulmonary circulation. At the same time shunt-related complications such as early or late shunt occlusion, surgical adhesions, asymmetrical growth, chylothorax, phrenic and vagal nerve palsy, may increase the complexity of subsequent corrective surgery [4]. Although acute postoperative care and survival indicate good results when performed after the neonatal period, surgical palliation has its own risks in underweight infants [4,5]. Therefore; transcatheter interventions are increasingly used as an alternative to surgery for initial palliation. Ductal stenting has become a good alternative for the initial palliation. But; unfavorable ductal anatomy (long and tortuous; vertical origin from the internal arch) is more common than expected in this subgroup of patients. Usually, unusual arterial access and multiple manipulations are associated with increasing procedural time and complications like ductal spasm, incomplete stenting, etc. [1]. PDA stenting after the neonatal period has not been found to be much effective in underweight infants. In such cases where ductal stenting cannot be achieved, guidewire perforation and balloon dilatation or radiofrequency pulmonary valve perforation may be performed [6]. Membranous pulmonary atresia, as in our case, is the most suitable anatomy for percutaneous perforation with antegrade or retrograde approach. This intervention with/without RVOT stenting presents a more physiological solution for establishing pulmonary flow. The thickness of the atretic valve, RVOT diameter and RVOT length are very important in the patient selection for procedural success.

There are many studies in the literature comparing the efficacy of ductal stenting and surgical shunts. In most of these studies, the efficacy of surgical shunt and ductal stenting for the first palliation were found to be similar, and it was reported that ductal stenting could be the first option due to short hospitalization and safety [7]. In a previous study comparing RVOT stenting with surgical shunts; similar to ductal stenting, RVOT stenting has been shown to be similarly effective and safer than surgical alternatives [8-10]. A meta-analysis including ten studies comparing RVOT stenting and the BT shunt in TOF patients, no significant difference was observed in arterial oxygen saturation increases and mortality rates [8]. Also, the study of Quandt et al. [9], which comparing the BT shunt and RVOT stenting, showed PICU and length of hospital stay were shorter and complete repair was performed earlier in RVOT stenting group compared to the BT shunt group. It has been shown that there is no significant difference in mortality rates. In the same way another study comparing the perforation and RVOT stenting and surgical shunts for palliation of patients with PA/VSD, showed that no significant difference was observed in terms of efficacy, length of hospital stay, increase in oxygen saturation, and operation time in complete repair, but the reintervention rate was shown to be higher in the SPS group [10]. Although the disadvantages of RVOT stent implantation are currently

being discussed nowadays, some recent case series demonstrated similarities about the length of bypass and mortality between stenting and surgical repair [4,10]. In our case, we performed pulmonary valve perforation and RVOT stenting as the first palliation and referred our patient to the total repair operation when she was ten months old.

4. CONCLUSION

In selected cases with PA/VSD, catheter based pulmonary valve perforation followed by RVOT stenting may be an effective and safe method compared to surgical shunt options in cases where ductal stenting is not feasible.

CONSENT

Written informed consent was obtained from the patient's family for the publication of the case report and the accompanying images.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

REFERENCES

1. Santoro G, Palladino MT, Capozzi G, Iacono C, Russo MG, Calabrò R. Pulmonary artery growth following arterial duct stenting in congenital heart disease with duct-dependent pulmonary circulation. Catheter Cardiovasc Interv. 2009 Dec 1;74(7):1072-6. doi: 10.1002/ccd.22191. PMID: 19642190.
2. Ahmad U, Fatimi SH, Naqvi I, et al. Modified Blalock-Taussig shunt: immediate and short-term follow-up results in neonates. Heart Lung Circ. 2008 Feb;17(1):54-8. doi: 10.1016/j.hlc.2007.06.003. Epub 2007 Aug 1. PMID: 17683985.
3. Marshall AC, Love BA, Lang P, et al. Staged repair of tetralogy of Fallot and diminutive pulmonary arteries with a fenestrated septal defect patch. J Thorac Cardiovasc Surg 2003; 126: 1427–1433.
4. Sandoval JP, Chaturvedi RR, BChir MB, et al. Right ventricular outflow tract stenting in tetralogy of Fallot infants with risk factors for early primary repair. Circ Cardiovasc Interv 2016; 9(12): e003979.
5. Arsdell GSV, Maharaj GS, Tom J, et al. What is the optimal age for repair of tetralogy of fallot? Circulation. 2000;102(suppl_3). Iii-123-Iii-9.
6. Walsh MA, Lee KJ, Chaturvedi R, et al. Radiofrequency perforation of the right ventricular outflow tract as a palliative strategy for pulmonary atresia with ventricular septal defect. Catheter Cardiovasc Interv 2007; 69: 1015–1020.

7. Mallula K, Vaughn G, El- Said H, et al. Comparison of ductal stenting versus surgical shunts for palliation of patients with pulmonary atresia and intact ventricular septum. Catheter Cardiovasc Interv 2015;85(7), 1196-1202.

8. Ghaderian M, Ahmadi A, Sabri MR, et al. Clinical outcome of right ventricular outflow tract stenting versus Blalock Taussig shunt in tetralogy of Fallot: a systematic review and meta-analysis. Curr Probl Cardiol 2020; 46: 100643.

9. Quandt D, Ramchandani B, Penford G, et al. Right ventricular outflow tract stent versus BT shunt palliation in Tetralogy of Fallot. Heart 2017;103(24), 1985-1991.

10. Aurigemma D, Moore JW, Vaughn G, et al. Perforation and right ventricular outflow tract stenting: Alternative palliation for infants with pulmonary atresia/ventricular septal defect. Congenit Heart Dis 2018; 13(2), 226-231.

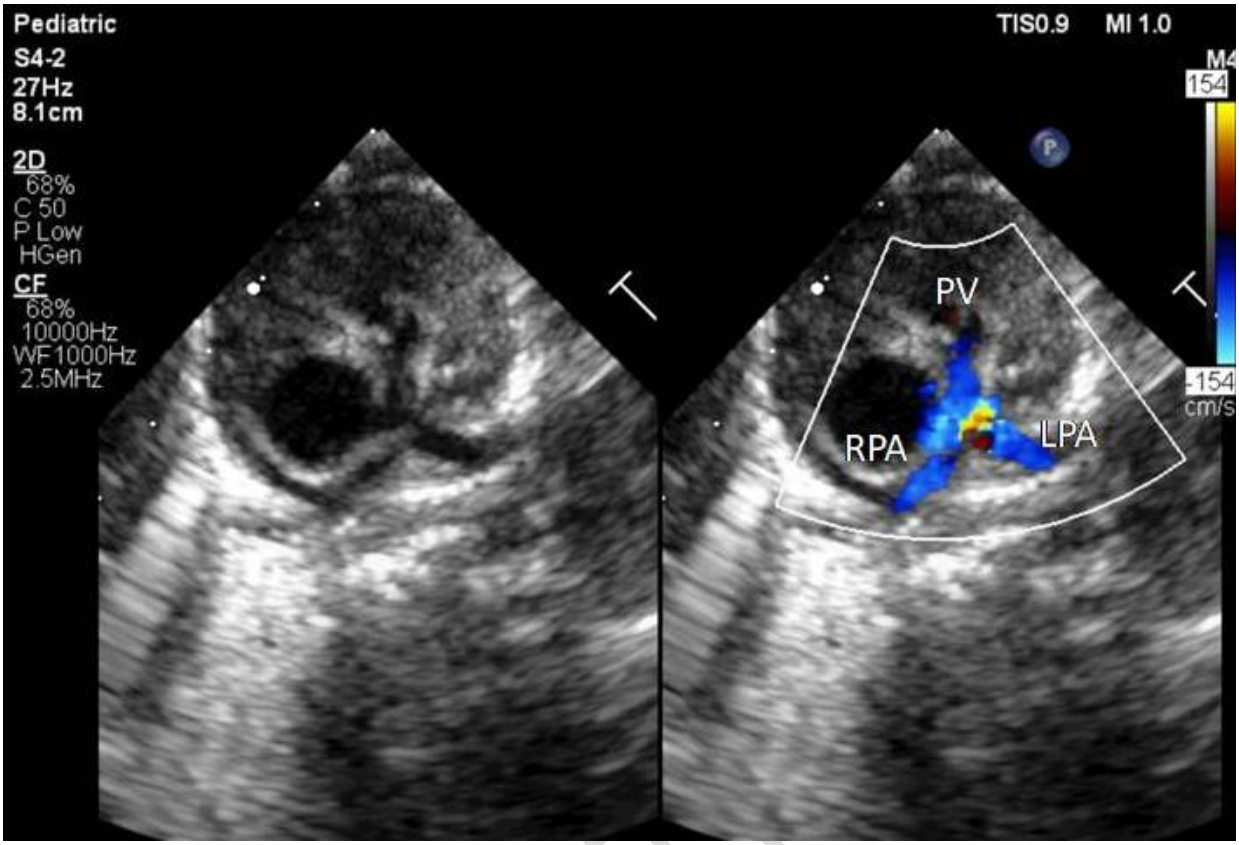

Fig. 1. Echocardiography showing membranous pulmonary atresia with ventricular septal defect, confluent and good-sized branch PA's.

LPA: left pulmonary artery, PV: atretic pulmonary valve, RPA: right pulmonary artery

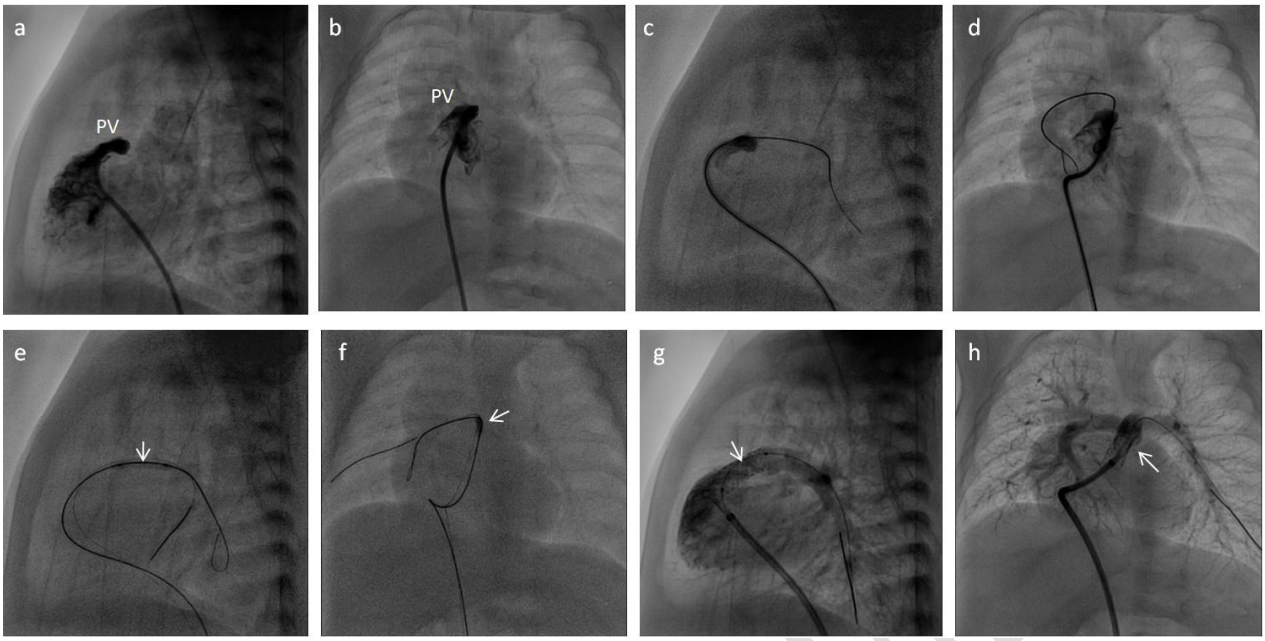

Fig. 2. Catheterization images. Image of the atretic pulmonary valve (PV), lateral (a) and anteroposterior (b) view. Perforation of the pulmonary valve using the Conquest Pro CTO guidewire, lateral (c) and anteroposterior (d) view. Predilatation with coronary balloon (arrow), lateral (e) and anteroposterior (f) view. Image of the adequate flow in the branch pulmonary arteries after the RVOT stent (arrow), lateral (g) and anteroposterior (h) view.

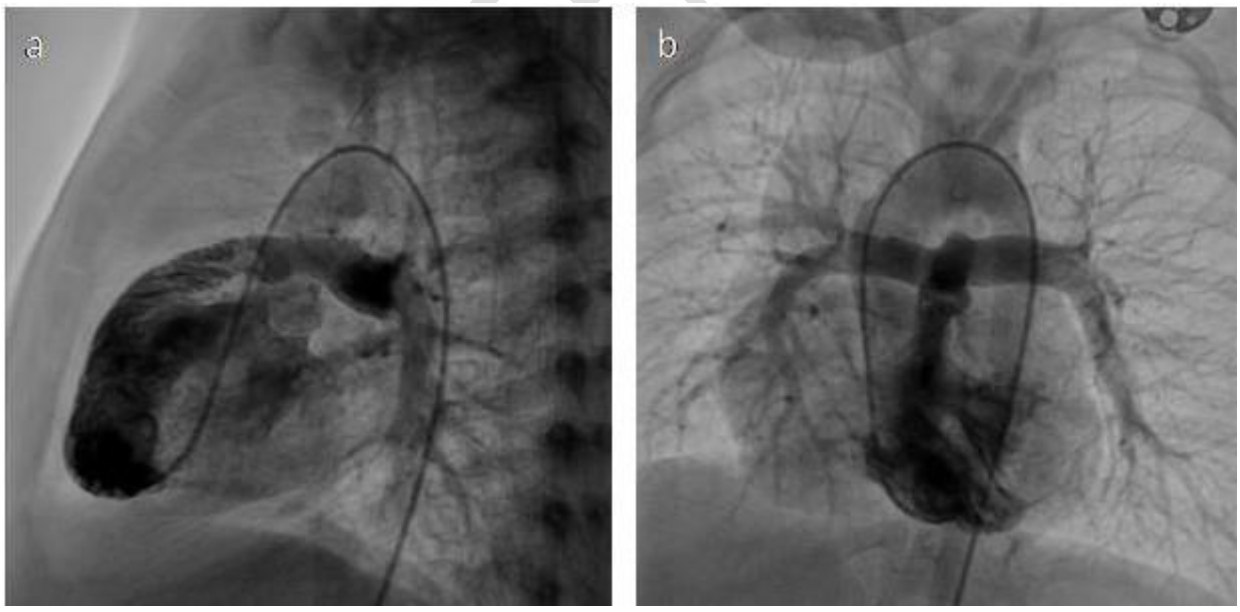

Fig. 3. Image of the increased pulmonary artery flow following stenting of the RVOT prior to the surgical correction. Both pulmonary arteries appear to be well developed, lateral (a) and anteroposterior (b) view.