Percutaneous Transcatheter Closure of a Stented Patent Ductus Arteriosus in a Patient With Critical Pulmonary Stenosis: A Case Series

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Abstract

INTRODUCTION: TStenting of the ductus is an alternative to surgical shunt in patients with critical pulmonary stenosis who remain ductal dependent following valvuloplasty. Stents close spontaneously after improvement of right ventricular function; however, in rare cases, stents fail to completely endothelialize, resulting in pulmonary overcirculation.

CASE PRESENTATION: The first case was a 3-year-old who had critical pulmonary stenosis at birth, and the second case was a 3-year-old boy who at 5 months of age was managed for severe pulmonary valve stenosis. Both underwent stenting of the duct and percutaneous pulmonary balloon valvuloplasty. On follow-up, the patent ductus arteriosus (PDA) stent of both cases remained patent even after an adequate growth and improvement of the right ventricle, resulting in significant left-to-right shunting and development of increased pulmonary blood flow; hence, percutaneous transcatheter closure of the stented PDA was done.

DISCUSSION: Stenting of the ductus allows significant and balanced growth of the pulmonary artery in patients with duct-dependent congenital heart diseases leading to adequate growth of the right ventricle. Persistence of patency of stent causing significant left-to-right shunting warrants closure of the PDA stent. Closure of the stent is challenging and requires a high degree of technical skill because of the difficulty to enter the partially closed stent.

CONCLUSION: Percutaneous transcatheter closure of a stented PDA in patients with critical pulmonary stenosis can be a technically challenging procedure because of the difficulty to enter the stent due to the presence of intrastent endothelial proliferation.

KEYWORDS: congenital heart disease, critical pulmonary stenosis, patent ductus arteriosus, transcatheter device closure, stent

INTRODUCTION

Critical pulmonary stenosis is a rare type of cyanotic heart disease in neonates where the degree of valvar pulmonary stenosis is severe enough to cause a decrease in right ventricular output.[1] At birth, affected infants are cyanotic and have systemic or suprasystemic right ventricular pressure, and pulmonary blood flow is usually provided by a patent ductus arteriosus (PDA). These infants usually require stabilization and initiation of prostaglandin E_1 (PGE_1) infusion to maintain ductal patency prior to performing balloon valvuloplasty.

Percutaneous balloon pulmonary valvuloplasty is the treatment of choice; however, despite successful relief of pulmonary obstruction, 5% to 10% of these patients were unable to sustain sufficient forward flow through the pulmonary valve to maintain adequate saturations because of their severely hypertrophic, noncompliant, and sometimes hypoplastic right ventricle (RV).[2]

Stenting of the ductus is increasingly accepted alternative to surgical shunt in patients who remain ductal dependent following valvuloplasty. Sometimes, redilatation of PDA stents to prolong patency of the stent and also to allow more time for RV function recovery is performed. Stents usually then close spontaneously as the right ventricular function improves, decrease in the right ventricular hypertrophy, and increase in RV size.

This case report discusses two patients with critical pulmonary stenosis who underwent percutaneous pulmonary balloon valvuloplasty and PDA stenting; however, the PDA stent did not completely endothelialize, and eventually, the patients presented with increased pulmonary blood flow; hence, percutaneous transcatheter closure of the stented PDA was done.

Procedural Details

Right-sided heart catheterization was performed through a right femoral vein percutaneous puncture. A 6F sheath was inserted, and a 5F MP catheter was manipulated under fluoroscopic guidance into the inferior vena cava (IVC), superior vena cava, right atrium (RA), RV, and main pulmonary artery (MPA). Left-sided heart catheterization was performed through a left femoral artery percutaneous puncture. A 5F sheath was inserted, and a 5F pigtail catheter was inserted and manipulated under fluoroscopic guidance to the aorta and into the left ventricle. Oximetry and pressure studies were done in each vessel and chamber entered (Table 1). A descending aortogram at an angle of the right anterior oblique 30° and lateral views was obtained and showed patent PDA stent both measuring 2 mm (Figure 1).

Table 1. Hemodynamic Studies Prior to Transcatheter Closure of the Stented PDA

	CASE 1	CASE 2
RV pressure (mmHg)	31/6	40/7
PA pressure (mmHg)	25/5 (15)	22/4 (13)
Gradient across the pulmonic valve	6	8
Aortic pressure (mmHg)	105/60 (84)	110/73 (93)
Qp:Qs	1.7:1	1.6:1

PA=pulmonary artery; PDA=patent ductus arteriosus; RV=right ventricular.

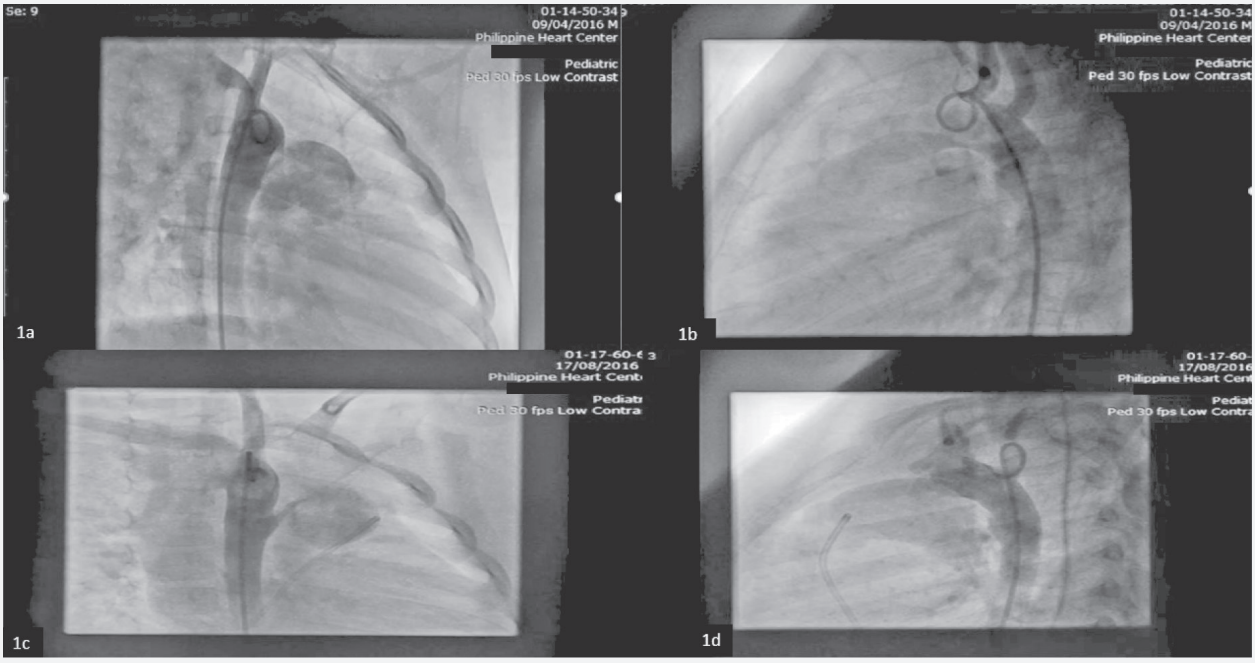

Figure 1. Descending aortogram at an angle of right anterior oblique 30° and lateral views showing patent PDA stent both measuring 2 mm (A, B: case 1; C, D: case 2)

In the placement of a PDA occluder in case 1 (Figure 2), a cut pigtail catheter was advanced at the level of the ampulla of the PDA. A long straight wire was then advanced through the pigtail catheter and was manipulated through the PDA and into the MPA. A snare catheter with a regular J-tip wire was then inserted through the venous access and was advanced to the IVC, RA, RV, and the MPA. The straight wire was then snared out of the venous access completing an arteriovenous (AV) loop. With the AV loop in place, the snare and pigtail catheters were then removed. In case 2 (Figure 3), a JR4 catheter was inserted and manipulated using a guidewire from the descending aorta and then was advanced at the level of the ampulla of the PDA. A long straight wire was then advanced through the JR4 catheter and was manipulated through the PDA stent and into the MPA. An MP catheter was inserted and manipulated through the venous access and was advanced to the IVC, RA, RV, and MPA. Using a kissing technique without using a snare catheter, the long straight wire was then externalized through the venous access, completing the AV loop.

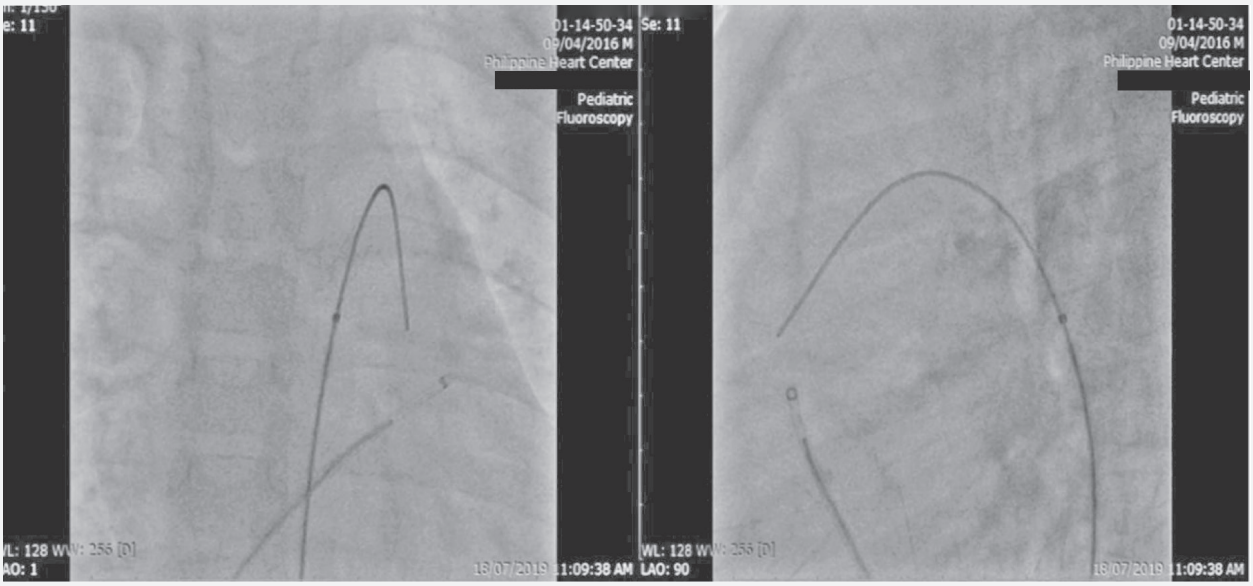

Figure 2. Case 1: the stent was crossed from the aortic end, and then an arteriovenous looping was done via a snare catheter technique

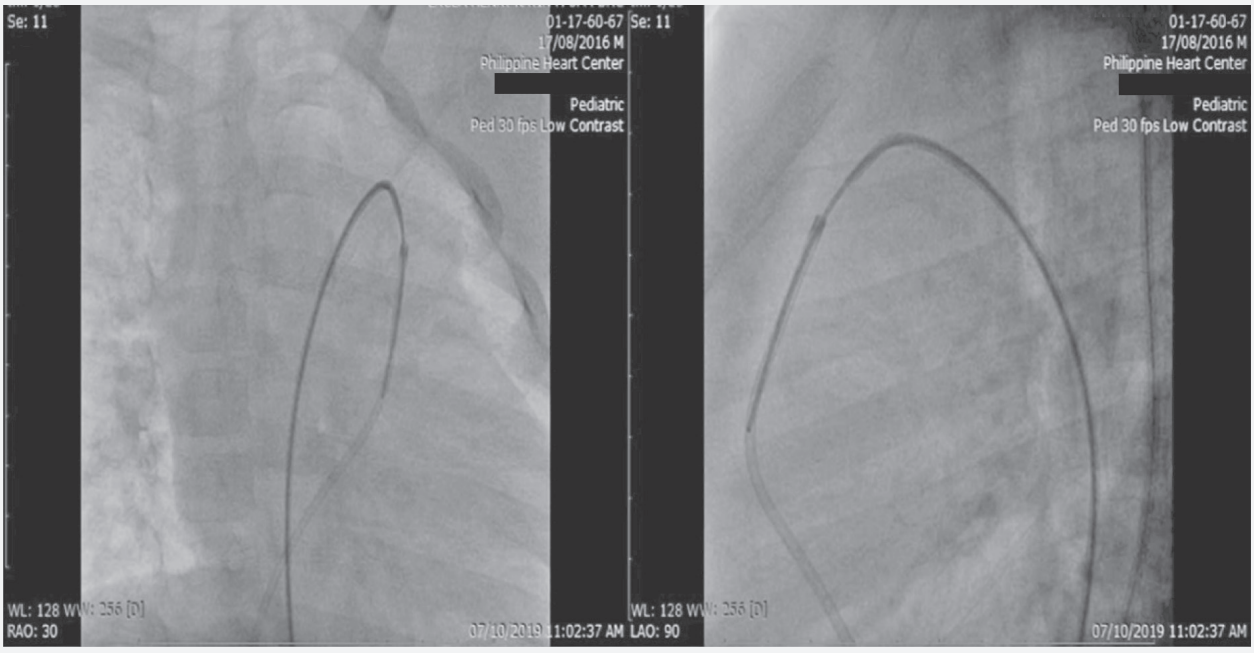

Figure 3. Case 2: the stent was crossed from the aortic end, and then an arteriovenous looping was done via kissing technique without the use of a snare catheter

A 4F delivery sheath was then advanced through the venous access through the IVC, RA, RV, MPA, and PA insertion of the PDA stent. A 6/4-mm LifeTech multifunctional occluder (MFO) device was inserted through the delivery sheath at the ampulla of the PDA and with the distal end at the pulmonic side to occlude the PDA (Figure 4). The device was then deployed.

Angiography was again done to determine the position of the device and showed the device in place and with minimal egress of dye through the PDA device. The device was released, and cineangiography after occlusion of the PDA showed the device positioned within the PDA stent (Figure 5).

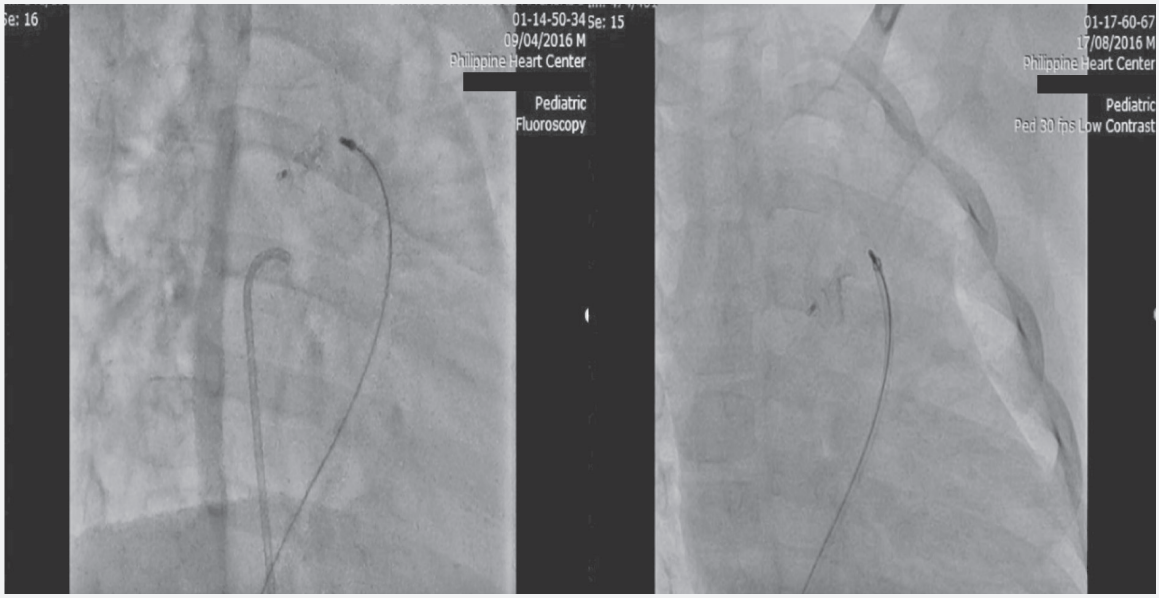

Figure 4. Deployment of the LifeTech multifunctional occluder device size 6/4 mm via antegrade technique

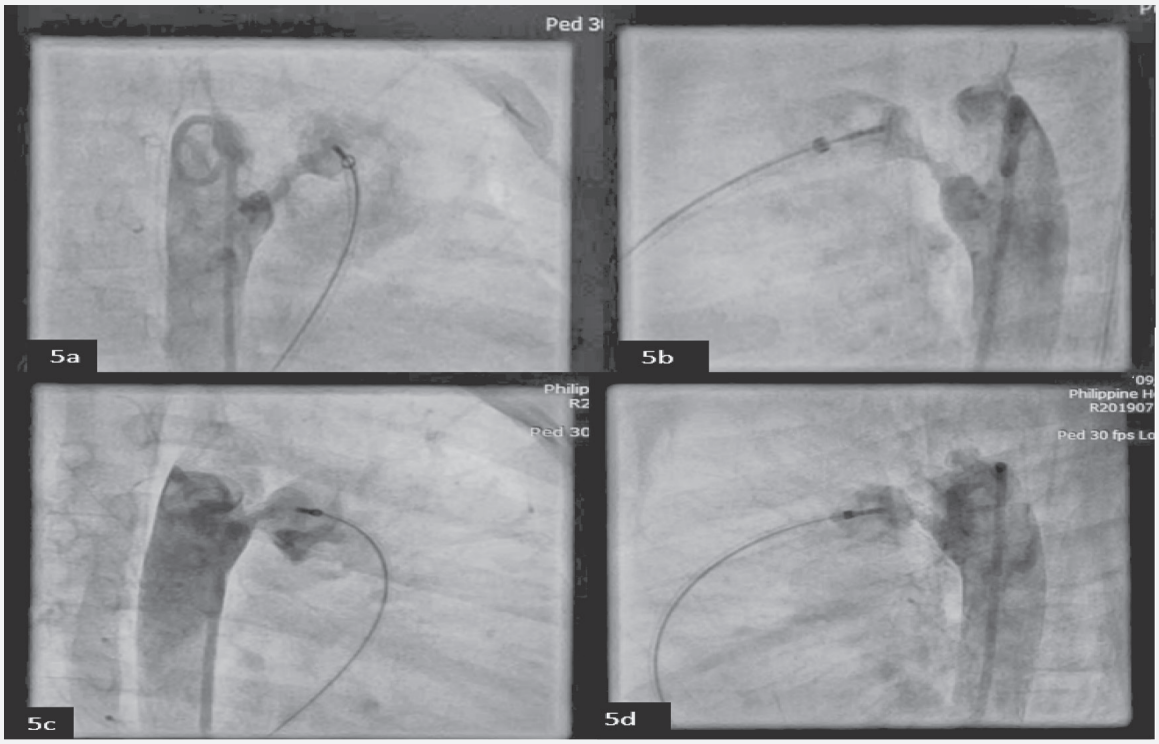

Figure 5. Descending aortogram at right anterior oblique 30° and lateral views showing the device in place and with minimal egress of dye through the device (A, B: case 1; C, D: case 2)

Case 1

The first case was a 3-year-old boy who at birth was diagnosed as a case of congenital heart disease and critical pulmonary stenosis (Table 2), who at the third day of life presented with desaturation and ductal restriction that existed despite PGE_1 infusion; hence, immediate PDA stenting using Boston Omega coronary stent size 4 × 8 mm was initially done. After securing the patency of the ductus arteriosus, percutaneous pulmonary balloon valvuloplasty was done using an 8 × 20-mm TMP-PED balloon catheter with residual gradient across the pulmonic valve of 13-mm Hg post–percutaneous pulmonary balloon valvuloplasty (PPBV). Oxygen saturation improved up to 93%; the patient was discharged, improved 2 weeks after the procedure, and was maintained on aspirin. On subsequent follow-ups, patient initially was thriving well, asymptomatic, and fully saturated, and serial chest radiography showed the PDA stent in place, normal vascularity, and normal heart size. However, at 2 years of age, patient started to have frequent lower respiratory tract infection, and on physical examination, the patient was acyanotic; vital signs were within normal limits. Oxygen saturation was 98% on all extremities, with adynamic precordium, apex beat at the fourth intercostal space left midclavicular line, S1 normal, S2 physiologically split, normal P2, and grade 3/6 continuous murmur at the left upper sternal border. Chest radiography revealed increased pulmonary blood flow and left ventricular prominence, and the electrocardiogram showed left ventricular hypertrophy. Echocardiographic findings showed a gradient across the pulmonary valve of 14 mm Hg, a patent PDA shunt with continuous Doppler signal of 79 mm Hg, Qp/Qs of 1.7:1, and left ventricular enlargement. The patient underwent hemodynamic studies and percutaneous transcatheter closure of the stented PDA. The patient made an uneventful course following the procedure. Twenty-four hours after the procedure, chest radiography revealed the PDA device in place, decreased vascularity, and decreased left ventricular hypertrophy (Figure 6). Echocardiography revealed the PDA device in place, no leak, normal chamber sizes, and good ventricular function.

Case 2

This is the case of a 3-year-old boy who was diagnosed as a case of congenital heart disease, pulmonary stenosis valvar, severe, PDA, and patent foramen ovale at 5 months old, who was admitted due to severe cyanosis. On echocardiography (Table 2), the RV was noted to be hypertrophied and noncompliant, unable to sustain sufficient forward flow through the pulmonary valve. Descending aortogram showed a ductal restriction at the pulmonic end; hence, stenting of the PDA implantation using a 4 × 12-mm Medtronic Resolute integrity coronary stent was initially done to maintain adequate saturation. Eventually, the patient underwent percutaneous pulmonary balloon valvuloplasty using a 12 × 20-mm TMP-PED balloon with residual gradient across the pulmonic valve post-PPBV of 10 mm Hg. O_2 saturation improved up to 96%, and 24 hours after the procedure, chest radiography showed increased pulmonary vascularity; hence, diuretics were started. Patient was then discharged and improved 10 days after the procedure and maintained on antiplatelet and diuretics. On immediate follow-up, there were clinical symptoms of pulmonary overflow, and chest radiography showed increased pulmonary vascularity; hence, diuretic was maintained. Serial echocardiography revealed mild residual pulmonary stenosis, patent PDA stent with continuous Doppler signal, patent foramen ovale with left-to-right shunting, and improving right ventricular function. On subsequent follow-ups, clinical improvement was noted, and pertinent physical examination showed the patient was acyanotic, vital signs were within normal limits. Oxygen saturation was 99% on all extremities, with adynamic precordium, apex beat at the fourth intercostal space left midclavicular line, S1 normal, S2 physiologically split, normal P2, and grade 3/6 systolic ejection murmur at the left lower sternal border. However, on chest radiography, there was a persistence of increased pulmonary blood flow and left ventricular prominence; latest echocardiography revealed patency of the PDA stent with continuous Doppler signal of 46 mm Hg, Qp/Qs of 1.6:1, and gradient across the pulmonary valve of 23 mm Hg. The patient underwent hemodynamic studies and percutaneous transcatheter closure of the stented PDA. The patient made an uneventful course following the procedure. Twenty-four hours after the procedure, chest radiography revealed the PDA device in place, decreased vascularity, and normal chamber sizes (Figure 6). Echocardiography showed the PDA device in place, no leak, normal chamber sizes, and good ventricular function.

DISCUSSION

Percutaneous balloon pulmonary valvuloplasty is indicated for a patient with critical valvar pulmonary stenosis defined as pulmonary stenosis present at birth with cyanosis and evidence of PDA dependency according to American Heart Association recommendation .[3]

Uncommonly, poor RV compliance due to severe right ventricular hypertrophy may cause persistence of severe hypoxia and dependence on PGE_1 despite an adequate relief of pulmonary obstruction. In this regard, the patient should be given the minimum possible amount of PGE_1 to provide pulmonary blood flow with a target oxygen saturation of approximately 90%. In some cases, a surgically modified Blalock-Taussig shunt or PDA stenting would be subsequently required.[4] A modified Blalock-Taussig shunt has been associated with complications, such as phrenic or vagal nerve paralysis, chylothorax, distorted pulmonary artery growth, pulmonary branch stenosis, and surgical adhesions with reported mortality rate of 7% to 14%.[5] Considering the high mortality and morbidity associated with BTS, stent implantation to maintain ductus arteriosus patency has been used in diseases with duct-dependent systemic blood flow or pulmonary blood flow.

Stenting of the ductus allows significant and balanced growth of the pulmonary artery in patients with duct-dependent congenital heart diseases, leading to adequate growth of the RV.[6,7] The success of stent implantation in PDA varies between 80% and 95%, depending on the type of congenital heart disease. Possible complications of stenting of the ductus are acute thrombosis, spasm of the ductus arteriosus, stent migration, and pulmonary overcirculation.[7,8] In both cases, there is a persistence of desaturations due to restriction of the pulmonic end of the

Table 2. Echocardiographic Characteristics Prior to PPBV and PDA Stenting

	CASE 1	CASE 2
Pulmonary valve annulus (mm), z-value	6 mm, z-3	9 mm, z 0
Gradient across the pulmonic valve (mm)	64	85
TV annulus (mm), z-value	13 mm, z 0	13.4 mm, z-1
RV characteristics	tripartite	tripartite
Tricuspid regurgitation	severe, jet 84 mmHg	severe, jet 92 mmHg
RVFAC	34%	40%
Patent foramen ovale	0.4 cm, right to left shunting	0.3 cm, right to left shunting
PDA size (mm)	1.5 mm, systolic doppler signal	2 mm, systolic doppler signal

PDA=patent ductus arteriosus; PPBV=percutaneous pulmonary balloon valvuloplasty; RV=right ventricle; RVFAC=right ventricular fractional area change; TV=tricuspid valve.

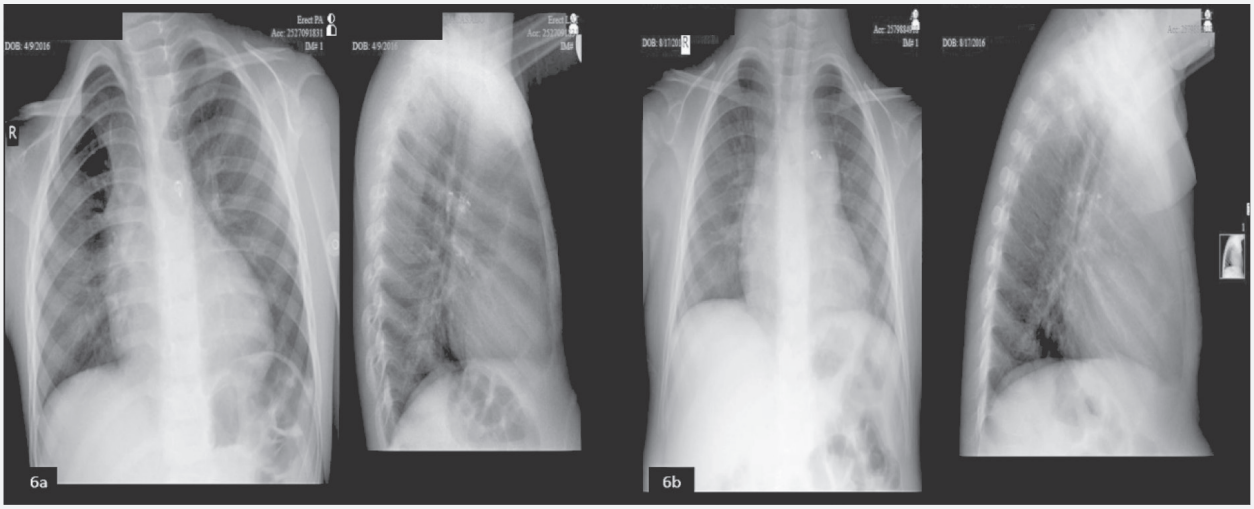

Figure 6. Chest radiography (A: case 1; B: case 2) revealed the PDA device in place, decreased in vascularity and chamber sizes

PDA and a poorly compliant and hypertrophic RV, which results in insufficient antegrade flow across the pulmonic valve; hence, an immediate PDA stenting was done prior to PPBV. The procedure was tolerated by both patients with no recorded complications in the immediate outcome.

The ductal stent has been observed to completely endothelialize at 30 days.[9] In a study by Schneider et al,[10] stent patency decreases over time, and stent was completely occluded between 4.5 and 17 months, with a mean of 10 months. There are cases when reduction in flow across the PDA or acute thrombosis occurs too early; hence, redilatation of the stent is performed in order to prolong stent life. In both cases, the PDA stent remained to be patent even after an adequate growth and improvement of the RV, resulting in significant left-to-right shunting and development of increased pulmonary blood flow.

According to the 2011 American Heart Association guidelines,[3] transcatheter PDA occlusion for the treatment of PDA with left-to-right shunt resulting in pulmonary overcirculation and left ventricle enlargement is a class I indication (level of evidence: B), hence the procedure on both patients. However, transcatheter closure of the stented PDA poses unique challenges. It may be difficult to access the PDA stent with a routine antegrade technique due to stenosis at the pulmonic end secondary to the presence of intrastent endothelial proliferation. In both cases, initial attempts to cross from the pulmonary end of the PDA stent with a multipurpose catheter in antegrade position failed. Hence, AV looping was done via a snare catheter in case 1 and via kissing technique in the other case. An MFO, which is a soft, woven-mesh, self-expanding device with two discs joined by an articulated waist, was used in both cases. Each disc of the MFO has a hub on the external surface; thus, the device can be in either retrograde or anterograde position. An MFO was used because it is self-expanding and thus can accommodate the entire length of the PDA stent (8–12 mm), from the entrance to the exit of the stent. Both patients made an uneventful course following the procedure and were discharged 24 hours after

the procedure. On immediate follow-ups, both patients were asymptomatic, and chest radiography revealed the PDA device in place, decreased vascularity, and normal chamber sizes. Echocardiography revealed the PDA device in place, no leak, no obstruction to adjacent vessels, normal chamber sizes, and good ventricular function.

CONCLUSION

Percutaneous transcatheter closure of a stented PDA in patients with critical pulmonary stenosis is challenging and requires a high degree of technical skill because of the difficulty to enter the stent due to the presence of intrastent endothelial proliferation.

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