

## CASE REPORT

# Emergency transcatheter closure of a stented PDA in a patient with pulmonary atresia and intact ventricular septum: be ready for the unexpected!

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## Introduction

Pulmonary atresia with intact ventricular septum (PAIVS) is a rare congenital heart disease with a reported prevalence of 4–6 cases per 100,000 live births. It is characterized by marked anatomic heterogeneity including plate-like valvar versus long segment infundibular atresia, a functionally intact ventricular septum, varying degrees of underdevelopment of the right ventricle (RV), RV-coronary artery connections (fistulas or sinusoids), and both functional and structural abnormalities of the tricuspid valve (TV). A patent ductus arteriosus (PDA) usually provides pulmonary flow [1, 2]. Due to its morphological variability, different surgical and transcatheter strategies can be chosen, mainly according to the morphology of the RV and pulmonary valve–infundibulum complex [3]. A percutaneous approach can be reserved for patients without a RV-dependent coronary circulation, consisting of transcatheter perforation of the atretic pulmonary valve (PV) with a wire, laser, or radiofrequency energy, followed by balloon dilation of the valve, with or without stenting of PDA [4]. PDA stenting is commonly performed in patients who have undergone successful

### Key Clinical Message

Patients with congenital heart disease and duct-dependent pulmonary circulation can undergo stenting of the patent ductus arteriosus (PDA). This case shows that, due to the physiological changes occurring after stent implantation, sometimes it is necessary to close the stented PDA rather than to redilate it.

### Keywords

Amplatzer occluder device, cardiac catheterization, ductus arteriosus, patent, pulmonary atresia with intact ventricular septum, stents.

radiofrequency pulmonary valve perforation but who still need pulmonary circulation support, being impossible to wean from prostaglandin infusion [5]. Stents usually then close spontaneously as the RV function improves. However, PDA stents are sometimes redilated during follow-up to lengthen stent life and allow more time for RV function recovery [6]. Here, we report a case where stent redilation was complicated by signs of severely reduced diastolic aortic pressure, myocardial ischemia, and a circular shunt. PDA stent closure was urgently indicated.

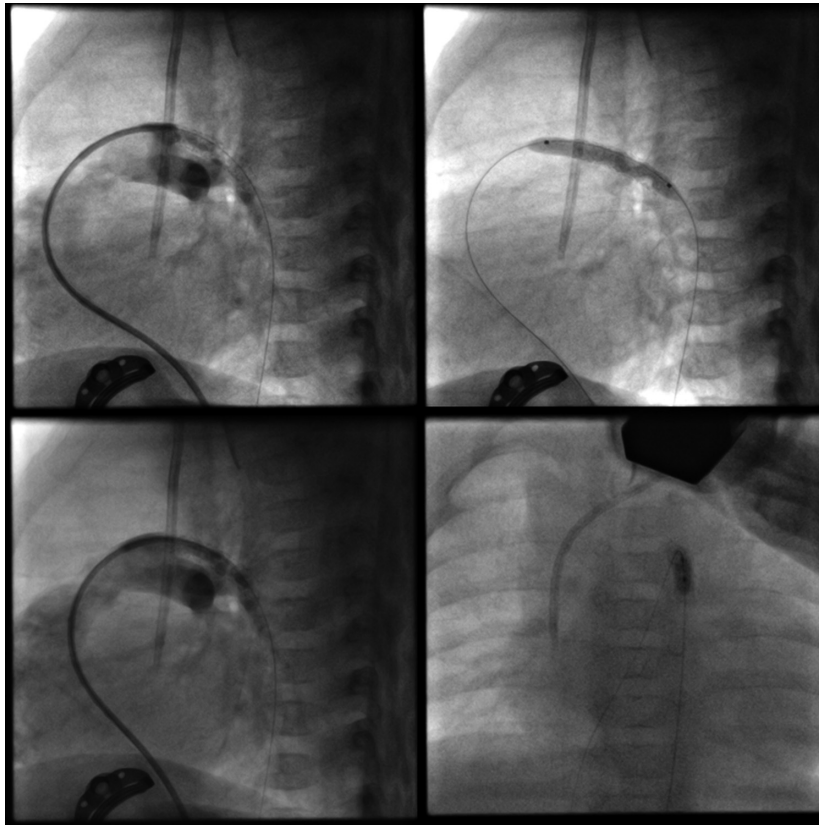
## Case Report

A baby with a prenatal diagnosis of PAIVS, born at the 38 + 4 weeks of gestation, underwent PV perforation with radiofrequency and subsequent balloon dilation of the PV on the second day of life. At angiography, the annulus was 7.2 mm and a 3 × 20 mm coronary balloon as well as two Tyshak Mini balloons (NuMED Canada Inc.), 6 × 20 mm and 8 × 20 mm, respectively, were used to dilate the PV. The residual gradient across the PV was 10 mmHg. Due to prostaglandin dependence and persistent cyanosis, after 1 week a Pro-Kinetic Energy

Biotronik  $4 \times 18$  mm bare metal stent was implanted in the PDA, through right venous femoral access. Oxygen saturations were 93% and the patient was discharged home 10 days after the procedure. During follow-up, the patient showed a progressive reduction in systemic oxygen saturation down to 82% and echocardiographic evidence suggestive of high systolic pressures in the RV (100 mmHg) with a high pressure gradient measured across the PV, together with moderate-to-severe tricuspid regurgitation was seen. Furthermore, the flow across PDA looked smaller compared to that at implantation. After having acquired informed consent from the baby's parents, a further cardiac catheterization was performed when the patient was 2 months old under general anesthesia and orotracheal intubation. Full heparinization (100 UI/kg) and antibiotics were given intravenously. Left femoral venous access was obtained. The PV appeared severely dysplastic, with poor opening of the cusps and an annulus diameter of 8 mm, moderate tricuspid regurgitation and no communication between RV and coronary arteries evident on RV angiography (Video S1). A

peak-to-peak gradient between the RV and the pulmonary artery of 35 mmHg and an RV systolic pressure equal to the 79% of the systolic systemic pressure were measured. RV pressures were 50/8/22 mmHg. A pulmonary valvuloplasty was then performed with a Tyshak Mini balloon (NuMED Canada Inc.)  $8 \times 20$  mm and with a Tyshak Mini balloon (NuMED Canada Inc.)  $10 \times 20$  mm (Video S2). A final peak-to-peak gradient of 7 mmHg with moderate PV insufficiency and a RV systolic pressure equal to the 38% of the systolic systemic pressure were obtained. However, oxygen saturations were still at around 84%. It was therefore decided to perform PDA stent redilation. In fact, the PDA stent showed mild stenosis due to neo-intimal proliferation (Fig. 1, Videos S3 and S4). Progressive dilation with coronary balloons (Accuforce Terumo, Terumo Corporation, Tokyo, Japan)  $3 \times 8$  mm and  $3.5 \times 8$  mm was performed (Fig. 1, Video S5).

Soon after PDA stent redilation, aortic pressure tracings showed a low diastolic aortic pressure of 20 mmHg. Signs of severe ischemia occurred with ST depression in the ECG that then turned into ST elevation and the



**Figure 1.** Fluoroscopy and angiography in lateral view showing: PDA stent mild stenosis due to neo-intimal peeling (left upper); balloon redilation of the stent in the PDA (right upper); angiography after redilation showing a significantly improved shunt across the PDA and relevant pulmonary and tricuspid regurgitation (left bottom); a  $3.5 \times 8$  mm coronary balloon inflated inside the stent while the wire was still in place to perform a balloon occlusion test of the PDA and reverse hemodynamic instability (right bottom).

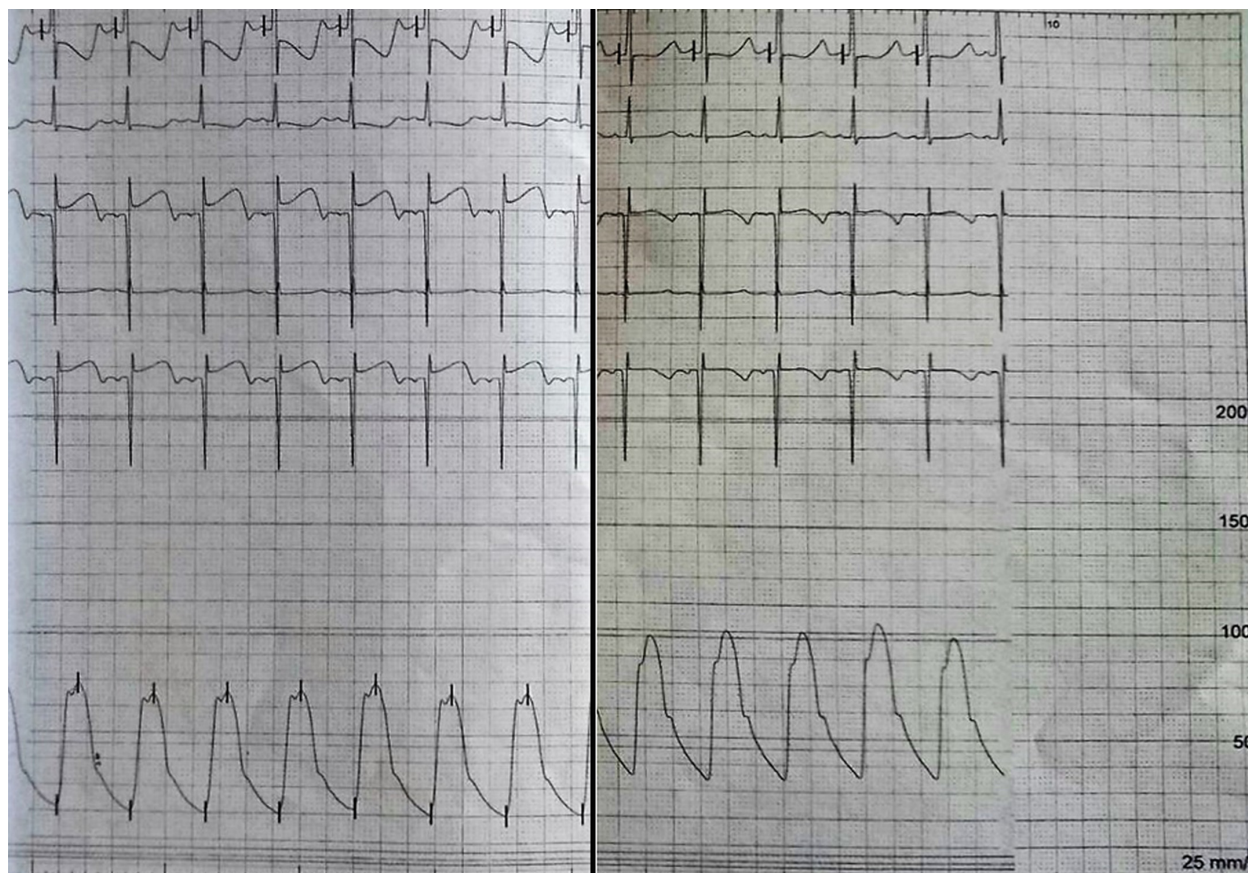
persistence of low cardiac output despite inotrope administration (Fig. 2). At echocardiographic examination, no pericardial effusion or right ventricular outflow tract obstruction was found, but a significant left-to-right shunt across the PDA with severe pulmonary and tricuspid valve regurgitation was shown on angiographic evaluation (Fig. 1, Video S6). A balloon occlusion test of the PDA was then performed using a coronary balloon (Accuforce Terumo, Terumo Corporation, Tokyo, Japan)  $3.5 \times 8$  mm while the wire was still in place (Fig. 1, Video S7). An immediate and significant improvement of the clinical situation occurred: the repolarization abnormalities disappeared and systemic diastolic pressure increased (40 mmHg) with acceptable blood oxygen saturations (88%; Fig. 2). Balloon-assisted advancement of a long sheath (Amplatzer TorqVue LP 5Fr, AGA Medical Corporation, Plymouth, MN, USA) inside the stent was performed (Video S8), and an Amplatzer Duct Occluder (ADO) II AS 4–3 mm was used to close the PDA (Fig. 3, Video S9). At the angiographic and echocardiographic control views, no shunt across the PDA was visible

(Fig. 3, Video S10). The device was unscrewed and it appeared stable inside the PDA stent (Fig. 3, Video S11).

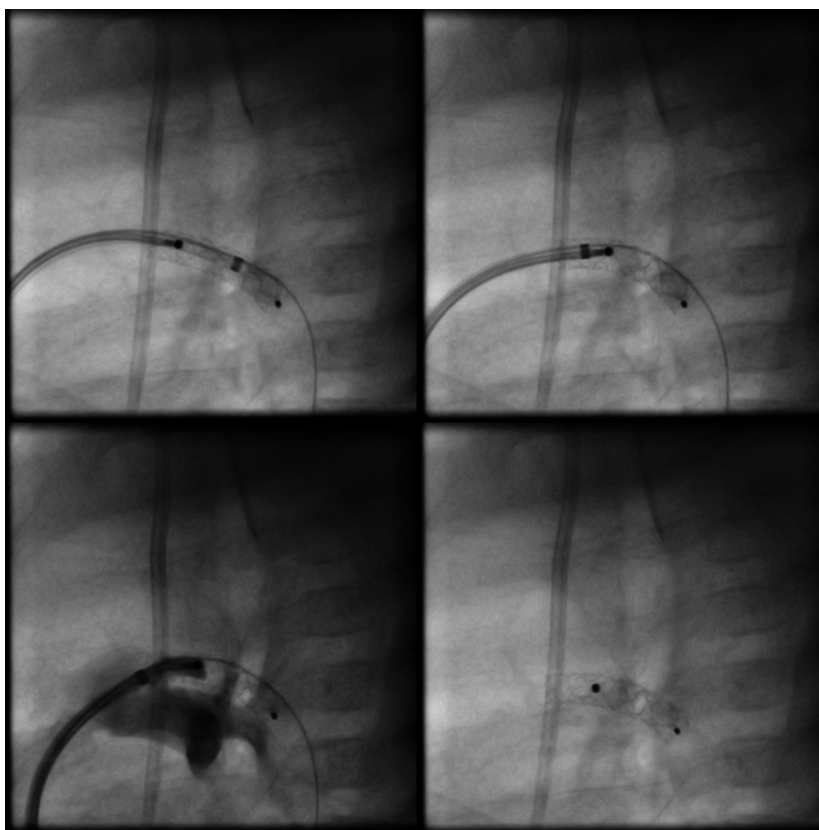
At the end of the procedure, the ECG and systemic pressures were normal and the pulmonary and tricuspid valve regurgitation was mild to moderate.

The baby underwent 24-hr monitoring in the intensive care unit and was then transferred to the pediatric cardiology ward. She remained hemodynamically stable, in sinus rhythm, with nonspecific repolarization abnormalities and negative T waves in V1 in the ECG. Oxygen saturations were 88%, and echocardiographic examination performed prior to discharge showed a mild to moderate left ventricular systolic dysfunction with hypokinesis of the inferior and lateral walls, maximum systolic gradient across the PV of 25 mmHg with mild insufficiency, and no shunt across the stent implanted in the PDA. She was discharged 4 days after the procedure.

At three month follow-up, the patient was growing well, oxygen saturations were 91% and there were no significant differences seen compared to discharge echo.



**Figure 2.** Left: low diastolic aortic pressure and ST elevation post stent redilation. Right: ECG normalization and diastolic aortic pressure increase during the occlusion test.



**Figure 3.** Fluoroscopy and angiography in lateral view showing: Amplatzer Duct Occluder II AS 4–3 mm deployment across the PDA stent. A buddy wire is left in place (left and right upper); angiography from the pulmonary side showing no residual shunting through the PDA (left bottom); Amplatzer Duct Occluder stable inside the PDA stent (right bottom).

## Discussion

Palliative PDA stenting is an alternative to a surgical modified Blalock–Taussig shunt in neonates and young infants who have heart disease with duct-dependent pulmonary circulation. It is usually indicated for patients at high risk if undergoing conventional surgical palliation, patients with anticipated need of a short-term support to the pulmonary circulation, and in low-risk neonates where early surgical repair may be planned as elective alternative to a systemic-to-pulmonary artery shunt. It is commonly contraindicated in patients allergic to contrast medium, in cases of active endocarditis and in the presence of pulmonary artery stenosis at the site of duct insertion. Relative contraindication may also be extreme ductal tortuosity [5]. PDA stenting has been associated with significant and balanced pulmonary arterial growth in congenital heart disease with completely duct-dependent pulmonary circulation over a short-term follow-up [7], allowing also adequate growth of the RV [8]. The reported success prevalence of stent implantation in PDA varies between 80 and 95%, dependent on the congenital

heart disease, with better results in PAIVS in comparison with defects with univentricular physiology [9]. Possible complications are acute thrombosis, spasm of the ductus arteriosus, and stent migration [10]. It seems that these stents undergo complete endothelialization by 8 weeks and intimal proliferation by 9 months [11]. When the reduction in flow across the PDA occurs too early, it can be possible to redilate stents during follow-up in order to lengthen stent life. Data in the literature are extremely heterogeneous however, reporting a redilation rate between 9 and 47% [6, 11–14]. To our knowledge, there are no reports on the deleterious effects of PDA redilation during follow-up.

In our case, we performed redilation of a stent in a PDA that presented with mild stenosis after PV valvuloplasty. After the stent redilation, myocardial ischemia occurred and it ceased after stent occlusion. The stent was even dilated at a lower diameter than it was during the first procedure, during which no signs of “severe aortic regurgitation physiology” had occurred.

We hypothesize that, as the diastolic RV function improved compared to the first days of life, the PDA

stent redilation created an “aortic regurgitation pathophysiology.” Furthermore, concomitant pulmonary and tricuspid valve regurgitation contributed to create an ineffective circulation with blood flowing across the stent toward the RV, then to the right atrium and across the atrial septum to the left atrium, in a circular manner that contributed to create coronary blood steal. In fact, temporary PDA stent closure significantly improved the clinical condition of our patient.

There are several technical issues that need to be underscored. First of all, it was crucial to keep the guide wire in place across the PDA stent: it allowed quick advancement of the coronary balloon to test occlude the PDA. In this way, it was possible to reverse the significant “aortic regurgitation type physiology” and test if PDA closure was compatible with patient’s clinical situation. Systemic oxygen saturation improved after PDA closure and RV diastolic pressures remained low. Secondly, it was important to use a balloon-assisted technique to help the progression of the long sheath across the PDA stent. In fact, when closing an abnormal vessel, it is mandatory to use a long sheath advanced beyond the target lesion, a rather not straightforward procedure when a stent is already in place. Finally, it was fundamental to select a device that perfectly fitted into the stent: the ADOII.

## Conclusion

Redilation of PDA stents is a widely accepted practice in patients with duct-dependent circulation. However, due to the physiological changes occurring after stent implantation, unexpected responses can happen and it could be necessary to change the interventional strategy from reopening to closing the PDA. We must always be ready for the unexpected.

## Conflict of Interest

The authors have no conflicts of interest to declare.

## Authorship

AF: collected data and wrote the paper; GB: performed the procedures, designed the project and revised the paper.

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## Supporting Information

Additional Supporting Information may be found online in the supporting information tab for this article:

**Video S1.** Right ventricular angiography in lateral view showing severely dysplastic pulmonary valve.

**Video S2.** Pulmonary valvuloplasty in lateral view.

**Video S3.** Angiography in lateral view showing PDA stent mild stenosis due to neointimal peeling.

**Video S4.** Aortography acquired via an antegrade approach showing PDA stent mild stenosis due to neointimal peeling.

**Video S5.** Balloon redilation of the stent in the PDA by using a 3.5 mm coronary balloon.

**Video S6.** Angiography post re-dilation showing a significantly improved shunt across the PDA and a relevant pulmonary and tricuspid regurgitation.

**Video S7.** A 3.5 × 8 mm coronary balloon was inflated inside the stent while the wire was still in place to perform a balloon occlusion test of the PDA and reverse hemodynamic instability.

**Video S8.** A balloon-assisted advancement of a long sheath inside the stent was performed.

**Video S9.** Amplatzer Duct Occluder II AS 4–3 mm deployment across the PDA stent. A buddy wire is left in place.

**Video S10.** Angiography from the pulmonary side showing no residual shunting through the PDA.

**Video S11.** Amplatzer Duct Occluder II AS is released and it looks stable inside the PDA stent.