SVOA Paediatrics
ISSN: 2755-1660

ScienceVolks Open Access

Case Report

Pulmonary Branch Stent Angioplasty in the Post-Surgery of Pulmonary Atresia with VSD

A. Chozas[1]*, A. Gutierrez[2], M. López Daneri[2], D. Cerruti[2], G. Colobraro[2], C. Kreutzer[3], B. Chiostri[3], D. Klinger[3], T. Iolster[4], A. Lugrin[4] and M. Rivarola[1]

[1] *Pediatric Hemodynamic, Hospital Universitario Austral, Argentina.*

[2] *Pediatric Cardiology Department, Hospital Universitario Austral, Argentina.*

[3] *Pediatric Cardiovascular Surgery, Hospital Universitario Austral, Argentina.*

[4] *Pediatric Intensive Care Unit, Hospital Universitario Austral, Argentina.*

***Corresponding Author:** Dr. Chozas Agustin, Pediatric Hemodynamics, Hospital Universitario Austral, Argentina.*

DOI: https://doi.org/10.58624/SVOAPD.2024.03.080

***Received:** July 20, 2024* ***Published:** September 24, 2024*

Abstract

Introduction: Patients with congenital heart diseases that affect the outflow tract of the right ventricle usually develop lesions at both the pulmonary valve and the pulmonary branches. We present a case in which critical stenosis of the right pulmonary branch was treated with a low- profile peripheral stent 7 days after reconstructive surgery.

Method: Retrospective review of the clinical case, pre-surgical condition, surgical technique, complication and resolution during the hospital stay.

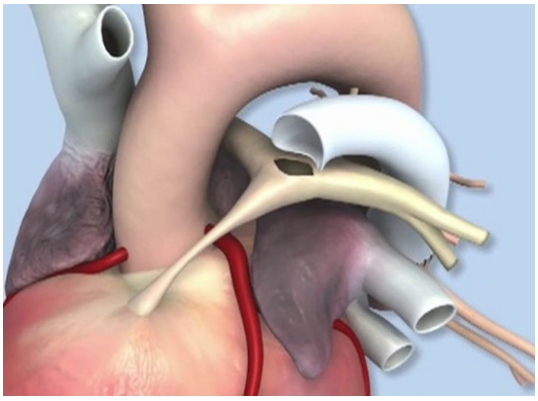
Result: Effective angioplasty of the right pulmonary branch with a low-profile balloon-expandable stent. Subsequent favorable clinical evolution.

Conclusion: Post-surgical injuries must be addressed early both for diagnosis and treatment, in order to obtain better clinical results. For this, the correct functioning of the multidisciplinary team work is necessary.

Keywords: *Pulmonary Atresia; Angioplasty; Congenital heart disease*

Introduction

Approximately 1% of the population is born with congenital heart disease and of these 20% have some complex anomaly of the right ventricular outflow tract[1]. Tetralogy of Fallot, Pulmonary Atresia with intact septum or with ventricular septal defect (VSD), Truncus Arteriosus, Transposition of the Great Vessels, some types of Double Outlet of the Right Ventricle and anomalies of the aortic valve treated, for example, with Ross surgery, are among the heart diseases that require surgical reconstruction of the right ventricular outflow tract (RVOT) using a Trans-Annular Patch (TAP) or some Bioprosthetic conduit (valve or valved tube)[2]. Among the post-surgical complications, we can mention dysfunction at the level of the valve plane or strictures at different levels of the pulmonary territory[3]. These complications depend on several factors (age of intervention, need for re-intervention, underlying heart disease and surgical technique used) and while the degeneration of the bioprosthetic valves or the valved conduits evolve towards Pulmonary Stenosis (PS) or Pulmonary Regurgitation (PR) RVOT reconstruction using TAP does so towards PR with significant dilation[4]. In turn, strictures (stenosis) at different levels of the pulmonary tree can occur in isolation or concomitantly with valve dysfunction. Therefore, multiple re-interventions are required during the life of these patients[5].



Schematic image of Pulmonary Atresia

We present a clinical case which presented in the immediate postoperative period a critical stenosis of the right pulmonary branch with resolution by catheterization.

Case Presentation

This is a male patient with a fetal diagnosis of Pulmonary Atresia with VSD and confluent branches, born at 38 weeks with a weight of 3200g. Central venous access was placed for the administration of Prostaglandins 0.01 gamma/kg/min. AngioTomography (AngioCT) was performed to verify the confluence of the central branches and the absence of important collaterals (see Figure 1).

At 17 days of age, corrective surgery was performed: "Through a central sternotomy, 3.5mm central branches are dissected. The anterior surface of the hypoplastic pulmonary trunk is opened to the left pulmonary branch and sutured to a valved polytetrafluoroethylene (PTFE) tube, expanding the origin of both branches. Through ventriculotomy, a wide VSD is closed with a PTFE patch and the proximal end of the tube is sutured, creating RV-PA continuity" (see Figure 2).

The patient progresses critically with high inotropic requirements (Adrenaline, Norepinephrine, Vasopressin and Milrinone). Due to hemodynamic decompensation, sternal re-opening with skin closure is performed. The echocardiogram showed systemic pressure in the RV, severe regurgitation of the RV-PA tube and critical stenosis of the right pulmonary branch (RPB) (see Figure 3).

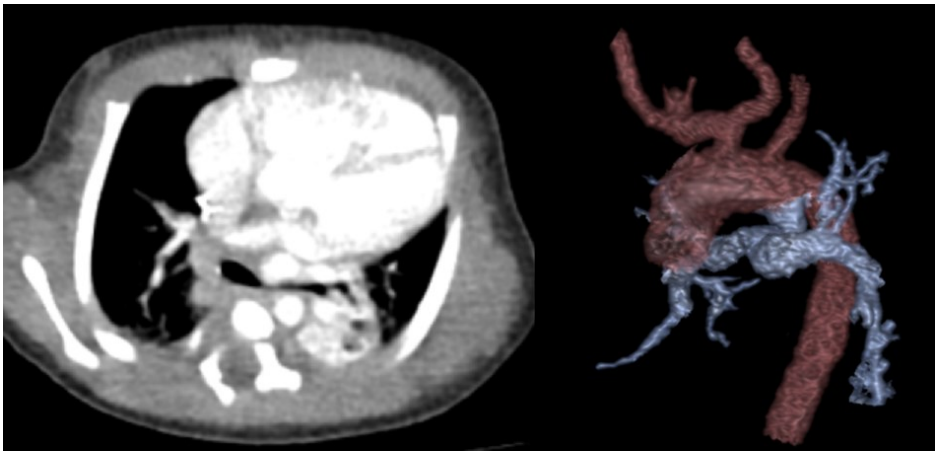


Figure 1: *Tomographic image and 3D reconstruction of pulmonary atresia and Ductus arteriosus (in blue) emerging from the aorta artery (in red).*

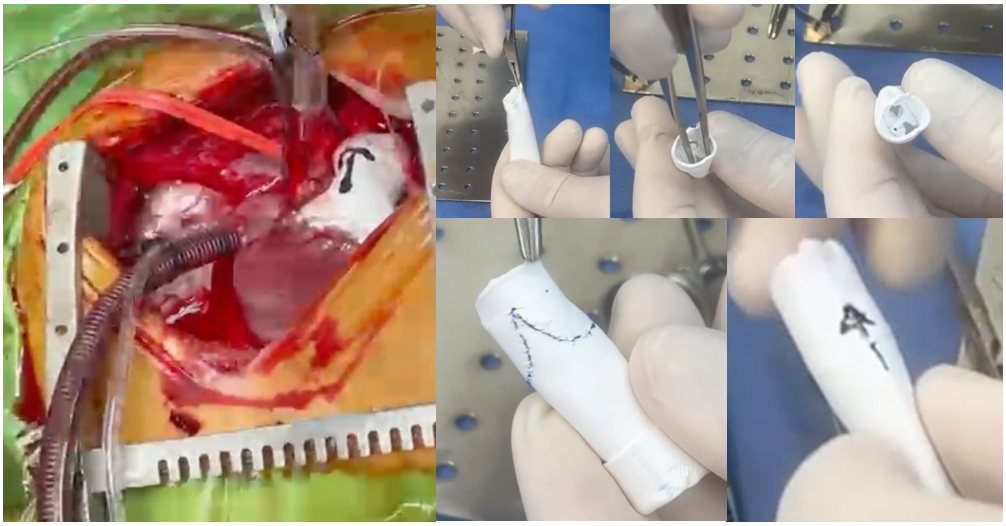


Figure 2: *Surgical view of the RV-PA tube implantation (left), manual assembly of the valved tube with PTFE (right). Courtesy Dr. Chiostri B.*

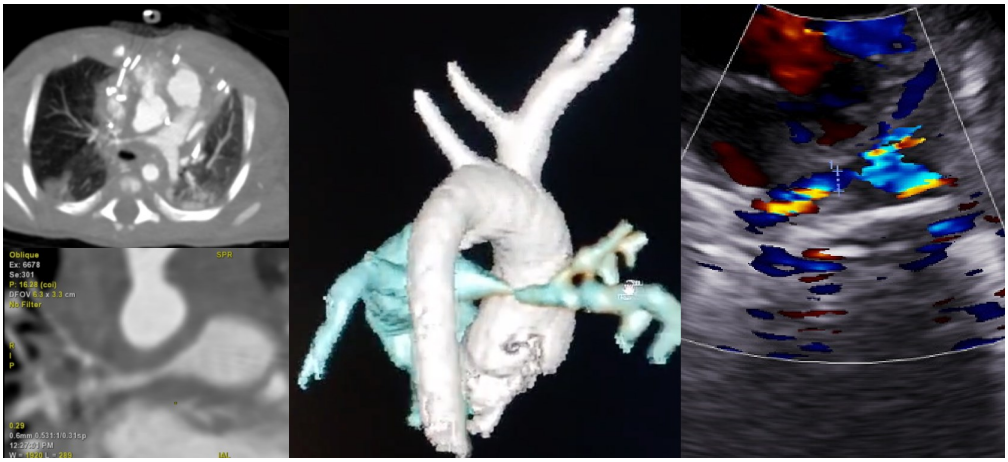


Figure 3: *Tomographic image of critical RPB stenosis (left), 3D reconstruction (center) of the lesion and pulmonary tree in blue and aorta artery in white, in posterior view. Transthoracic echocardiogram (right).*

At 24 days of life (7 days post-surgery) it was decided to enter the catheterization lab: "Stand-by ECMO, accessed by puncture of the left femoral vein, Heparinized with 100 IU/Kg. Systemic pressure in RV; oximetry samples are taken that reveal QP/QS 1.5; a pressure gradient of 40 mmHg between RV-AP; RPB which has at its origin a diameter of 4.6mm, a passage through the stenosis of 1mm and distally 3.8mm by angiography. It was possible to pass a 0.018" hydrophilic wire to the lower right branch and advance the Radix2 5x12 stent (CID S.p.a. - Alvimedica Group - Italy) rapid Exchange expandable balloon. It is insufflated with a pressurized syringe up to its nominal value of 8 atm and improvement in the expansion of the lesion is checked. Central vascular access is placed and the procedure is completed without complications" (see Figure 4).

At 28 days of life, chest closure and reduction of inotropes are achieved. Echocardiogram: Fixed VSD patch with 2.2mm residual shunt, mild RV dysfunction, severe PR with RV pressure 50% of the systemic pressure, no pericardial effusion, 4.2mm RPB through the stent (see Figure 5).

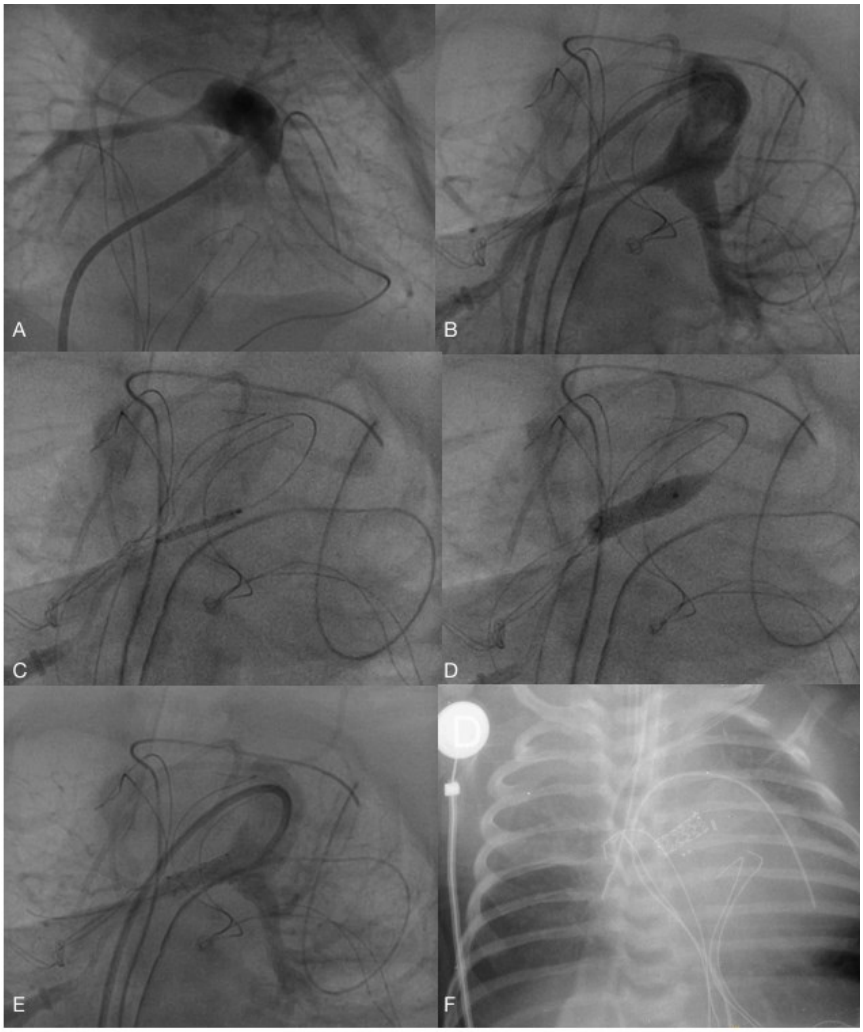


Figure 4: *(A) Angiography in cranial position; (B) Angiography in caudal position; (C) Stent positioning; (D) Balloon dilation and stent implantation; (E) Control angiography same projection as image B; (F) Anteroposterior X-ray control.*

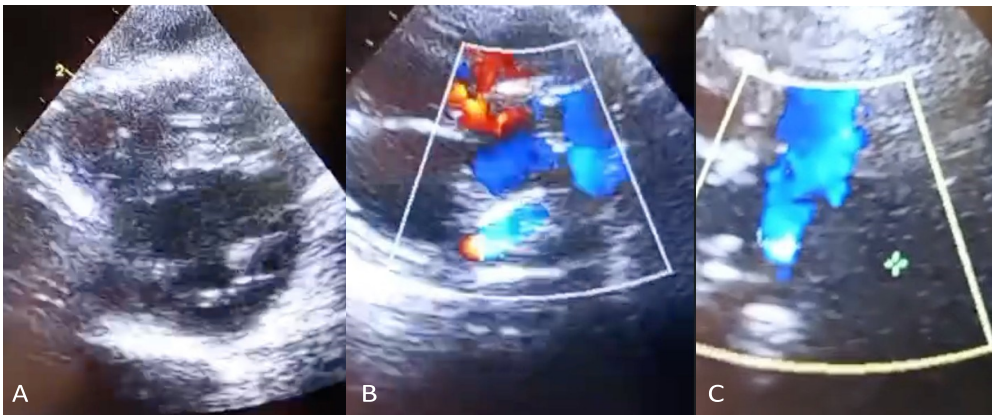


Figure 5: *Control echocardiogram. (A) LV short axis; (B) Flow through the stent in RPB; (C) VD-AP tube and connection with RPB.*

Discussion

In the post-surgical evolution of complex heart diseases that affect the RVOT, strict monitoring of residual lesions that patients may present is necessary. Obstructive lesions are usually not well tolerated in patients with ventricular dysfunction, so early diagnosis and effective treatment can change the course of the disease[6].

The indication for Intervention is usually accepted when: RV pressure >50% systemic pressure, cyanosis due to the presence of right-to-left shunt, dysfunction of the subpulmonary ventricle, pulmonary flow imbalance (20-25% of the flow to one lung), severe lung failure[7].

Angioplasty is the treatment of choice for stenoses and can be performed with low- or high- pressure balloons with or without stent implantation. In those cases where the injuries are rigid, cutting balloon can be used.

Especially in the cases of small patients with stenosis of pulmonary branches, we find ourselves with the difficulty of having dedicated material, either due to the low profile often necessary due to the weight of the patients as well as the need to redilate the stents without losing its structure. It is important when deciding to treat a critical stenosis with a stent in an emergency situation, to take into account that some time after angioplasty, either due to somatic growth of the patient or due to restenosis, re-expansion may be necessary. There are redilatable stents on the market, such as Genesis (Cordis, Santa Clara, CA, USA), Valeo (Bard, Murray Hill, NJ, USA) and Formula (Cook Medical, Bloomington, IN, USA), which require 6F introducers onwards. In our case, we used the Radix2 peripheral stent, which is sold in three versions (5, 6 and 7mm), has a low profile (5F and supports a 0.018" guide) and has the possibility of being reissued up to 2mm more without distorting its architecture offering the possibility of obtaining progressively larger diameters.

Angioplasty is considered effective when an increase in the diameter of the lesion greater than 50% and/or a decrease greater than 20% in the ratio of RV systolic pressure and systemic pressure is achieved.

In the case of elastic lesions, the use of stents would be indicated, which offer the following benefits: greater final lumen, lower incidence of restenosis, and the possibility of redilatation[8]. Complications that may occur after angioplasty are: restenosis, aneurysms, vascular rupture, unilateral or bilateral pulmonary edema, thrombosis, hemoptysis[9].

Conclusion

Both early diagnosis and therapy in post-surgical injuries that affect the RVOT can change the evolution of critically ill patients; for them, joint work between cardiology, critical care, cardiovascular surgery and pediatric hemodynamics is necessary. In accordance with what we find in the bibliography, we consider that it is necessary to have more devices that allow them to be introduced through low-profile sheaths and that tolerate over expansions without altering their architecture and radial force, accompanying the somatic growth of small patients.

Conflict of Interest

The authors declare they have no potential conflicts of interest to disclose.

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Citation: Chozas A, Gutierrez A, López Daneri M, Cerruti D, Colobraro G, Kreutzer C, Chiostri B, Klinger D, Iolster T, Lugrin A, Rivarola M. Pulmonary Branch Stent Angioplasty in the Post-Surgery of Pulmonary Atresia with VSD. *SVOA Paediatrics* 2024, 3:4, 141-146. doi:10.58624/SVOAPD.2024.03.080