Tetralogy of Fallot with Pulmonar Valve Imperforation in Extremely Preterm Infants. Prenatal Diagnosis and Neonatal Management: Right Ventricular Outflow Stent Placement as a Bridge To Definitive Repair

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Introduction

Tetralogy of Fallot (TOF) accounts for about 6% of all congenital heart defects. It has a wide spectrum of anatomical presentation that influence differently on clinical and hemodynamic presentation. Right ventricular outflow tract (RVOT) obstruction and pulmonary arteries sizes impact directly on oxygen saturation and clinical symptoms and, only the most severe forms require neonatal intervention[1,2]. Blalock-Taussig (BT) shunt is a palliative operation that has been the standard procedure to increase the pulmonary blood flow in cases of severe hypoxemia, postponing the definitive intervention to a more stable clinical condition. Although it is a simple procedure, prematurity and low birth weight impact significantly in morbidity and mortality, due to difficulties in the balance of pulmonary and systemic blow flows (Qp:Qs), especially if the shunt size is larger than it should be[2-4]. Stenting RVOT has been described as a palliative procedure in newborns and infants with low birth weight and/or preterm babies who need to improve pulmonary blood flow or depend on continuous infusion of E1 prostaglandin[1,5,6].

Case Report

Baby boy diagnosed prenatally with TOF, imperforate pulmonary valve and reverse flow through the arterial duct (Figure 1). Despite the imperforate pulmonary valve, branch pulmonary arteries were confluent and of good size. He was born in the 28th gestational week with extreme low weight (790 grams) due to severe placental insufficiency diagnosed in the 27th week. He was initially managed with continuous infusion of prostaglandin (PGE) and mechanical ventilation until he reached 1.5 kg (65 days of age), when was transferred to the cardiology hospital for interventional treatment.

Postnatal echocardiogram confirmed the diagnosis of TOF with imperforate pulmonary valve and well-developed pulmonary arteries. The right heart chambers were dilated, there was right-to-left shunt at the atrial level and the arterial duct was patent. The pulmonary valve had thickened leaflets with normal sized annulus (Z score = – 0.7) and no antegrade blood flow was noted.

Under general anesthesia, a 4F sheath was inserted in the right femoral vein allowing a successful percutaneous pulmonary valvuloplasty. The procedure was monitored by echocardiography to reduce the use of contrast. Although a pulmonary antegrade flow was noticed immediately after the pulmonary valve dilation, few minutes later it disappeared due to severe infundibular spasm. For this reason, a 4.5 x 12 mm stent ("Springer") was chosen to be placed in the RVOT across the pulmonary valve annulus, reestablishing the antegrade pulmonary flow (Figures 2 and 3). After the procedure, the patient oxygen saturation improved. He was immediately weaned out of the PGE infusion and slowly from mechanical ventilation. There was no complication in the venous access site. After one month of the procedure, he was discharged home weighting 2.4 kg and feeding orally.

The patient returned for surgical repair when he was 4 months old and 4.6 kg. The stent was removed and a transanular monocuspid patch was positioned in the RVOT with excellent anatomical result and clinical outcome (Figure 4).

Discussion

In patients with TOF, the degree of hypoxemia and occurrence of hypoxemic spells depend on the severity of RVOT obstruction (infundibular and valvar stenosis) and the size of pulmonary arteries. Neonatal intervention is unnecessary in the majority of patients but the obstruction tends to evolve with time. However, some cases present with severe pulmonary flow obstruction, identified during fetal life. The diagnostic sign in this setting is that the ductal flow is reverse (from the descending aorta to the pulmonary artery). This finding indicates that, in the neonatal period, the baby will have a pulmonary blood flow dependent on the patency of the arterial duct, and will need continuous infusion of prostaglandin to keep the duct open until the first intervention. Up to the last decade, the palliative procedure of choice was the modified BT shunt, in which a polytetrafluoroethylene graft is interposed between the subclavian artery and the ipsilateral pulmonary artery[7-9]. Although technically simple and with satisfactory results, the presence of prematurity and low birth weight lead to significant morbidity and mortality related to the procedure. A major limitation in preterm newborns is the graft size: even the smallest ones

Keywords

Infant, Extremely premature; Tetralogy of Fallot; Stent; Double outlet right ventricle; Balloon valvuloplasty; Echocardiography.

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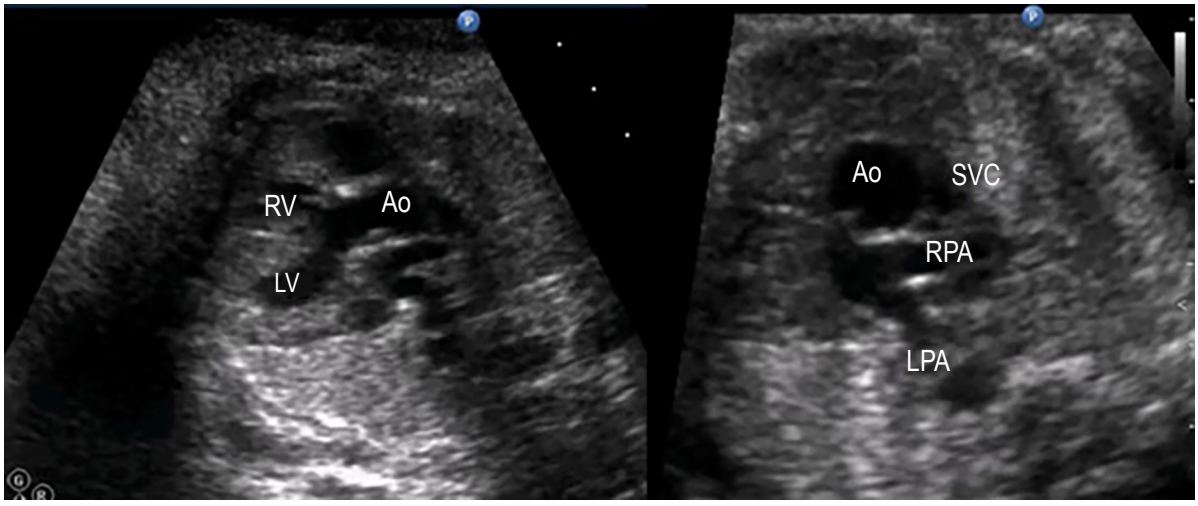

Figure 1 – *Fetal echocardiogram performed in the 24th gestational week: A - Large ventricular septal defect with dextroposed aorta overriding the interventricular septum; B - Three vessels view showing good-sized confluent pulmonary arteries and dilated aorta.*
LV: left ventricle; RV: right ventricle; Ao: aorta; SVC: superior vena cava; RPA: right pulmonary artery; LPA: left pulmonary artery.

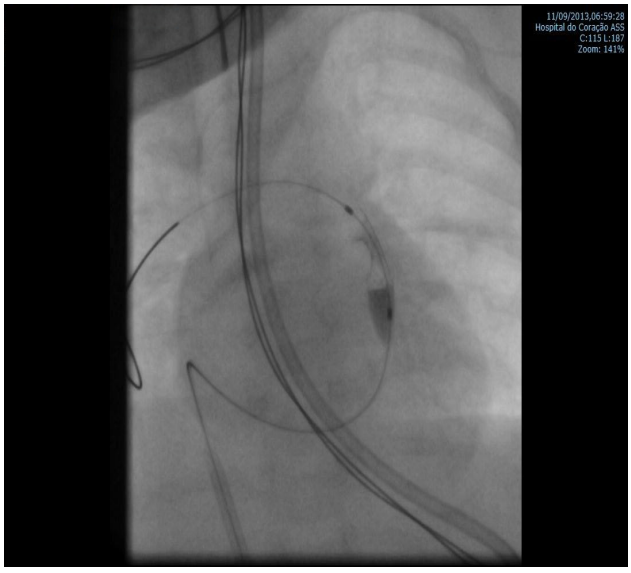

Figure 2 – *Radioscopy that shows the balloon expanding the stent positioned in RVOT.*

used in neonates (3 mm) are too large for preterm patients. Moreover, the oversized shunt may increase considerably the pulmonary blood flow and steal effective systemic blood flow, with risk of renal failure and enterocolitis (unbalanced Qp/Qs). On the other hand, the use of very small grafts are at high risk of thrombosis or inappropriate development and growth of the pulmonary arteries[7,8]. Another concern related to the BT shunt is the high chance for distortions and stenosis in the suture site at the pulmonary artery, that may occlude the upper pulmonary lobe branch[8,9]. Stenting the RVOT has been reported since early 1990s as an alternative treatment for cases similar to the described above, or for infants with hypoxemic spells and unfavorable anatomy for anatomical repair[10,11]. This percutaneous technique provides antegrade and pulsatile pulmonary blood flow allowing normal growth of the pulmonary arteries, and avoids the harms of a procedure with open chest and cardiopulmonary bypass. RVOT stenting in high surgical risk patients has proven to be an excellent alternative to hypoxemic babies, and has low complication rates in experienced hands, postponing the surgical intervention to a more favorable surgical condition[1,5,6,9], especially in very low birth weight babies. The optimal infant outcome observed herein, with oxygen saturations in the high eighties, signs of good thrive (weight Z score of -2.1) and

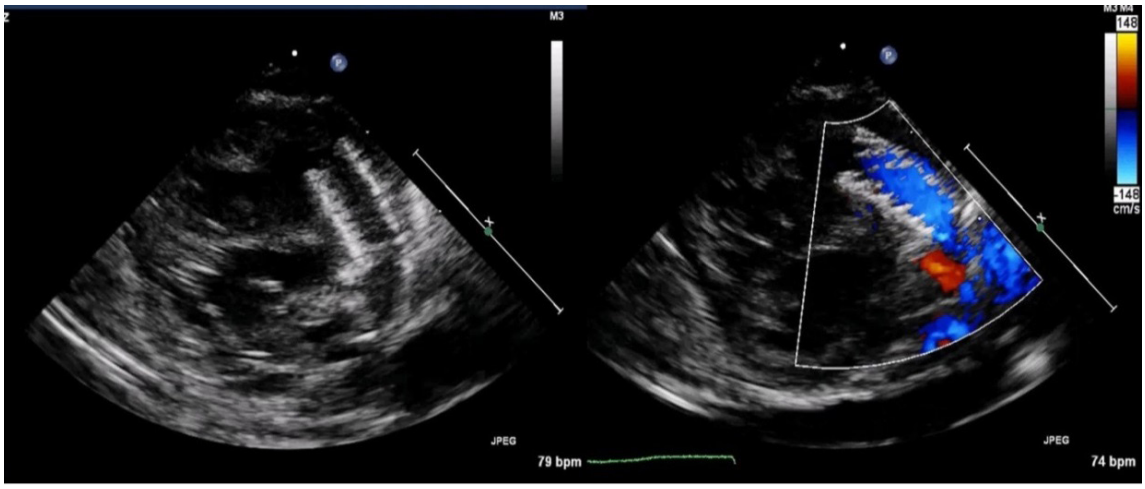

Figure 3 – *Echocardiogram performed in the catheterization laboratory interrogating the of stent placement. A: Free outflow tract; B: Free pulmonary antegrade flow.*

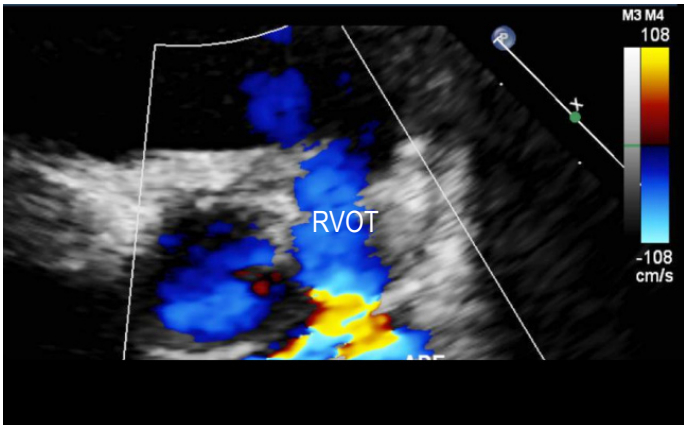

Figure 4 – *Echocardiogram showing free flow through RVOT after transannular patching. RVOT: right ventricular outflow tract.*

normal pulmonary artery growth, corroborate that it was the right choice for the management of such a high-risk patient. In this case, the collaboration of echocardiography and interventional cardiology in the catheterization laboratory must be pointed out. Likewise percutaneous closure of ventricular and atrial septal defects, echocardiography may contribute to the success of the intervention, reduce the use of contrast and x-ray exposure. In this specific case, it monitored the pulmonary valve perforation and dilation, and precipitated the need of RVOT stenting due to severe infundibular stenosis, minimizing the use of contrast in a 1.5 kg baby at risk for renal failure. It is of note that, although less invasive than an open heart surgery, percutaneous procedures poses hazard related to vascular access (very low weight is an important risk factor), cardiac perforation and tamponade, stent sub optimal positioning and rhythm disturbances. For these reason, a team experienced in neonatal percutaneous therapeutic catheterization should perform this type of procedure.

Conclusion

Pulmonary valve perforation and dilation followed by RVOT stenting in extreme preterm infant with TOF and imperforate pulmonary valve seems to be an excellent choice of management of such a challenged situation. Prenatal diagnosis may have impact in the optimal treatment of neonatal heart disease because it allows therapeutic strategy planning even before the patient is born, and better resolution of very unfavorable and risky circumstances.

Authors' contributions

Research creation and design: Cosentino CM; Shiraishi KS; Pedra SP. Data collection: Cosentino CM; Shiraishi KS; Pedra SP; Ponce LL. Data analysis and interpretation: Cosentino CM; Pedra SP; Ribeiro MS; Costa RN. Manuscript drafting: Cosentino CM; Pedra SP.

Critical revision of the manuscript for important intellectual content: Cosentino CM; Martins TC; Pedra CAC; Pedra SP.

Potential Conflicts of Interest

No relevant potential conflicts of interest.

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Academic Association

This study is not associated with any graduate program.

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