New Percutaneous Techniques for Perforating the Pulmonary Valve in Pulmonary Atresia with Intact Ventricular Septum

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We report new percutaneous techniques for perforating the pulmonary valve in pulmonary atresia with intact ventricular septum, in 3 newborns who had this birth defect. There was mild to moderate hypoplastic right ventricle, a patent infundibulum, and no coronary-cavitary communications. We succeeded in all cases, and no complications related to the procedure occurred. The new coaxial radiofrequency system was easy to handle, which simplified the procedure. Two patients required an additional source of pulmonary flow (Blalock-Taussig shunt) in the first week after catheterization. All patients had a satisfactory short-term clinical evolution and will undergo recatheterization within 1 year to define the next therapeutic strategy. We conclude that this technique may be safely and efficiently performed, especially when the new coaxial radiofrequency system is used, and it may become the initial treatment of choice in select neonates with pulmonary atresia and intact ventricular septum.

Even though pulmonary atresia with intact ventricular septum is an infrequent defect, accounting for less than 1% of congenital heart defects, it is the 3rd most frequent cyanotic heart defect in the neonatal period [1]. Its morphological spectrum is broad with cases ranging from extremely hypoplastic tricuspid valves and right ventricles to ventricular cavities of almost normal dimensions [2-4]. Ebstein's anomaly of the tricuspid valve and extreme ventricular dilation have also been reported, and they are associated with a poor prognosis [5]. In addition, communication between the right ventricular cavity and the coronary arteries is a relatively common finding, which is sometimes associated with coronary circulation partially or totally dependent on the right

ventricle [6-8]. Due to this anatomic heterogeneity, the therapeutic algorithm should be individualized [6]. The final objective is always to attain biventricular correction with total separation between systemic and pulmonary circulations [6]. However, this is sometimes impossible, and correction with a 1½ ventricle or of the univentricular type (Fontan) is necessary [6-9]. Cardiac transplantation should also be considered for treating cases with severe stenoses or multiple interruptions in the coronary arteries and secondary left ventricular dysfunction [6,8]. The initial therapeutic approach in the neonatal period should, whenever possible (if the coronary circulation pattern allows), open the pulmonary valve to decompress the right ventricle and stimulate its growth [3,6-8]. During the last decade, perforation of the pulmonary valve to establish continuity between the right ventricle and the pulmonary artery with the aid of interventional catheterization became a reality [10-15], even in Brazil [16]. We report 2 percutaneous techniques of valve perforation, which were recently introduced into clinical practice, and their advantages and disadvantages are discussed.

Case report

We report the cases of 3 patients referred to our service from other neonatal units for investigation or treatment of cyanotic congenital heart defects. The clinical, echocardiographic, and hemodynamic data, are listed in tables I, II, and III. It is worth noting that patient 2 had a previous diagnosis of critical pulmonary stenosis. All patients had mild to moderate cyanosis under continuous infusion of prostaglandin; on auscultation, the 2nd cardiac sound was single and low, and was followed by a mild systolic murmur in the dorsum. On chest X-ray, the cardiac silhouette was slightly enlarged, mainly because of the right atrium, and the pulmonary flow was reduced. The electrocardiogram showed sinus rhythm and left ventricular hypertrophy in all patients. QRS axes ranged from 90° to 120°. In regard to the echocardiographic findings, all patients had situs solitus, pulmonary atresia with imperforate pulmonary valve, and intact ventricular septum. The right ventricle was hy-

Table I – Clinical data of the patients

Patient	Age	Sex	Weight	Saturation
1	3 days	Female	3.2kg	75%
2	5 days	Female	3.0kg	85%
3	15 days	Male	2.8kg	75%

Table II – Echocardiographic data

Patient	TV (Z value)	TI	PuV	RV Hypo
1	8.5mm (-1.7)	Mild/Mod	7.0mm	Moderate
2	9.0mm (-1,3)	Mild/Mod	6.8mm	Mild/Mod
3	7.0mm (-2,1)	Mild/Mod	7.0mm	Moderate

TV- diameter of the tricuspid valve; TI- degree of tricuspid insufficiency; PuV- diameter of the pulmonary valve; RV Hypo – degree of right ventricular hypoplasia; Mod- moderate

Table III - Data of the catheterization prior to valve perforation

Patient	RV (mmHg)	LV (mmHg)	RV Hypo	Infund. Hypo	RV-Co Communic
1	60/6	41/6	Mild/Mod	Mild	No
2	50/6	50/6	Mild/Mod	Normal	No
3	60/7	50/6	Mod	Mild	No

RV – right ventricle; LV – left ventricle; RV Hypo – degree of right ventricular hypoplasia; Infund Hypo – degree of infundibular hypoplasia; RV-Co Communic - communications between the right ventricle and the coronary arteries; Mod: moderate.

poplastic with a significant reduction in the trabecular zone. The tricuspid and pulmonary valves also had varied degrees of hypoplasia (tab. II). No patient had communication between the right ventricle and the coronary arteries. The pulmonary branches were of a good caliber, and pulmonary circulation was maintained through the arterial canal to the left. The patients were then referred to the catheterization laboratory for diagnostic testing and possible perforation of the pulmonary valve. After a detailed explanation of the possible risks and benefits of the percutaneous procedure, the parents provided written consent. The hemodynamic study showed right ventricular systemic pressure levels equal to or above the systemic levels in all patients (tab. III). Right ventriculography confirmed the echocardiographic findings and absence of communication between the right ventricle and the coronary arteries (figs. 1 and 2). The infundibulum was slightly hypoplastic in patients 1 and 3 and almost normal in patient 2. The technique and results of percutaneous valve perforation are described for each patient.

Case 1 - A 5 Fr (Cook) right coronary Judkins catheter was carefully positioned right below the valvar plane and monitored through fluoroscopy in the cranial posteroanterior and lateral views (fig. 1 A and B). The hard tip of a 0.014" steerable angioplastic guidewire was manually molded accompanying the curvature of the right ventricular outflow tract, which was defined by angiography in the left
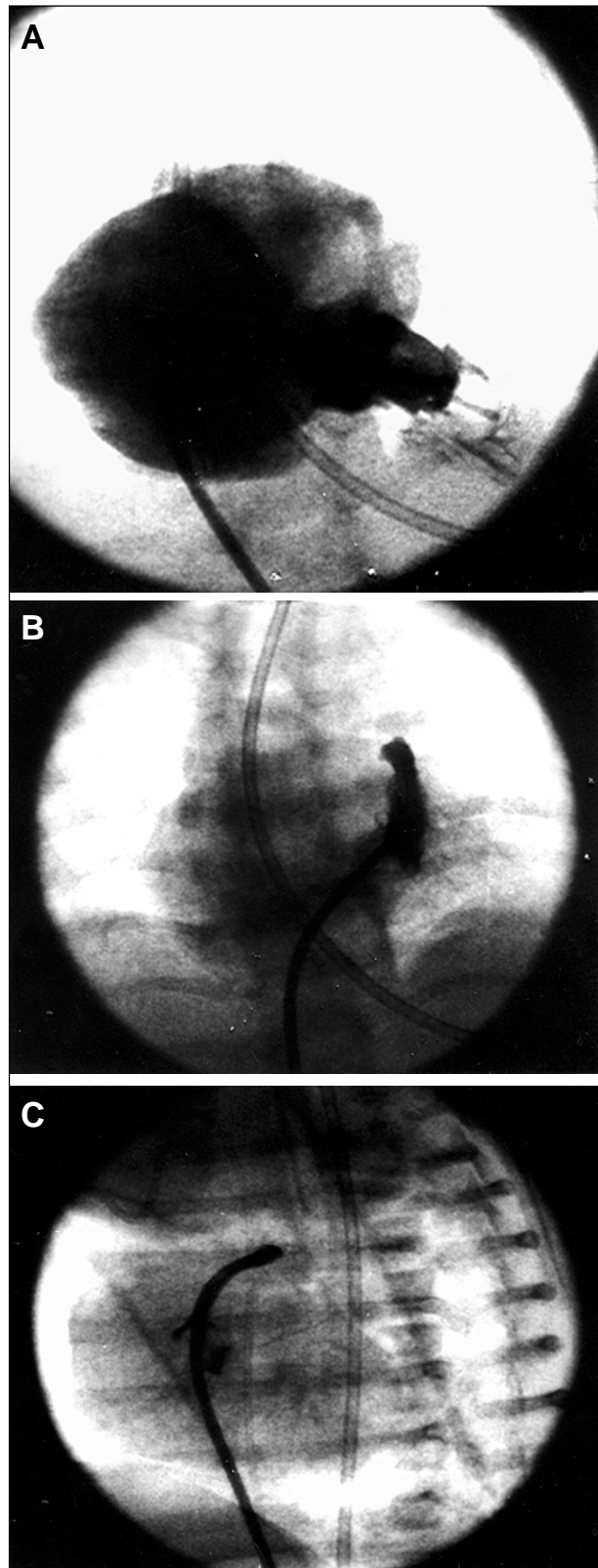
Fig. 1 - Case 1 - A) Right ventriculography in right cranial anterior oblique view. Note the right ventricle with mild to moderate hypoplasia and significant tricuspid insufficiency with mild opacification of the infundibular portion, without communication between the coronary arteries and the right ventricle; B) selective manual injection into the infundibulum with mild hypoplasia showing unquestionable pulmonary atresia; C) infundibulum in left profile view. A minimum anterograde jet exists through the pulmonary valve after perforation with the hard tip of the steerable coronary guidewire. Note how the guidewire and the catheter follow the curvature of the infundibulum, leading to adequate perforation at an angle perpendicular to the valve.
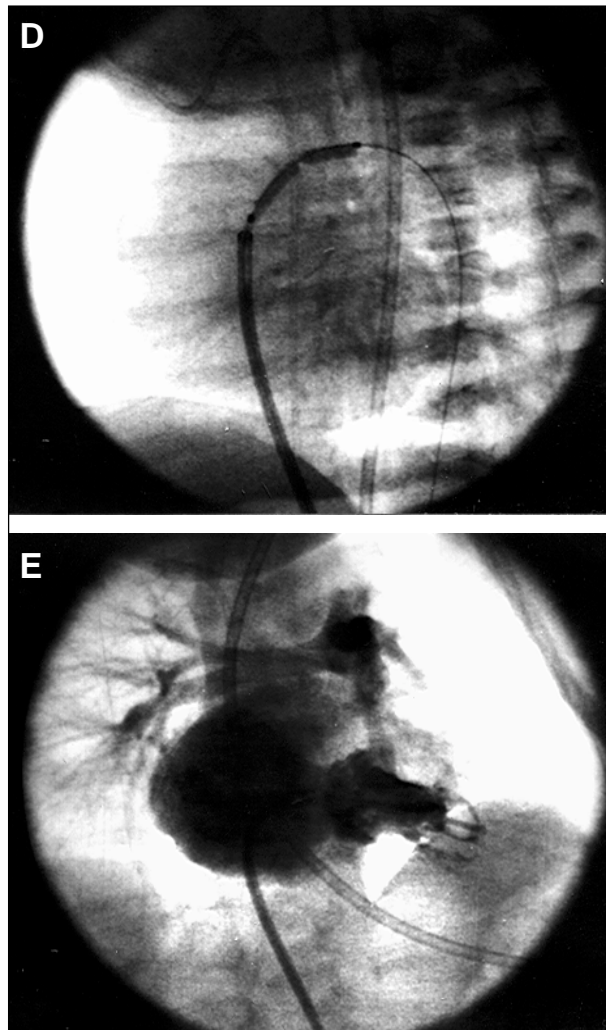
Fig. 1 - Case 1 - D) note the inflated coronary angioplastic balloon at the valvar level. The guidewire was left in the descending aorta through the arterial canal; E) right ventriculography in right cranial anterior oblique view after completion of the pulmonary valvoplasty. Note the anterograde flow through the valve with a reduction in the degree of tricuspid insufficiency. Dynamic infundibular reaction occurs.

profile view. This guidewire was then advanced through the Judkins catheter and positioned right below the valvar plane with its tip perpendicularly directed at the pulmonary leaflets. The appropriate positioning was confirmed by manual injections of contrast medium through a Y system connected to the Judkins catheter, commonly used for coronary angioplasty. The pulmonary valve was then mechanically perforated, and the hard tip of the steerable guidewire was advanced. With test injections, perforation and appropriate positioning of the guidewire in the pulmonary trunk were confirmed, and no signs of perforation or contrast medium extravasation, or a combination of the two, were found (fig. 1 C). The guidewire was withdrawn, and the flexible and directional tip of another steerable guidewire (0.014") was advanced through the same catheter, passing beyond the pulmonary valve through the orifice created by the hard tip of the previous guidewire. This guidewire was maneuvered through the arterial duat and positioned in the distal part of the descending aorta at the level of the

bifurcation of the iliac artery. A 3.0X20mm balloon catheter for coronary angioplasty (World-pass, Cordis) was advanced over the guidewire, positioned at the level of the valvar plane, and inflated under pressure monitoring with formation and disappearance of the sand-glass image (fig. 1 D). This catheter was then replaced by a 9X20 mm Tyshak II low-profile balloon catheter (Numed, Corwall, Canada) for completion of the pulmonary valvoplasty. The hemodynamic control revealed an infundibular gradient of 30 mm Hg and the absence of a gradient through the pulmonary valve (tab. IV). Control ventriculography showed reestablishment of right ventricle-pulmonary trunk continuity (fig. 1 E), with a dynamic infundibular reaction. Within the first 4 days after catheterization, we interrupted the infusion of prostaglandin but had to reinstate it due to systemic desaturation and hemodynamic lability. The patient was then referred for a right modified Blalock-Taussig shunt. The postoperative period was uneventful, with oxygen saturation maintained at around 80%-90% (patient extubated), absence of metabolic acidosis, and normal levels of lactate. The patient was discharged after 10 days with an echocardiogram showing anterograde flow through the pulmonary valve and a maximum systolic gradient of around 50 mm Hg, secondary to infundibular reaction. Moderate to severe pulmonary insufficiency and satisfactory flow through the Blalock-Taussig shunt existed. After a 7-month follow-up, the patient was clinically well, gaining weight, and his saturation was around 90%. The echocardiogram showed infundibular hypertrophy regression, a maximum systolic gradient of 15mmHg through the right ventricular outflow tract, severe pulmonary insufficiency, and a normally functioning Blalock-Taussig shunt. The right ventricle and the tricuspid valve developed (Z value = -1). The patient now awaits recatheterization to test occlusion of the shunt and of the oval foramen with new pressure measurements and calculation of cardiac output.

Case 2 – In this patient, we used a new coaxial radiofrequency system (Bayliss Medical Company, Mississauga, ON, Canada), designed by Nykanen, for perforation of the pulmonary valve (figs. 2A and B). As in the previous case, a 5 Fr. (Cook) right coronary Judkins catheter was positioned right below the valvar plane. The Nykanen radiofrequency catheter (external diameter 0.024", length 265cm) was advanced through the Judkins catheter and positioned below the pulmonary valve touching the leaflets. This catheter was connected to a generator source of radiofrequency (BMC) and was programmed to apply 5W of

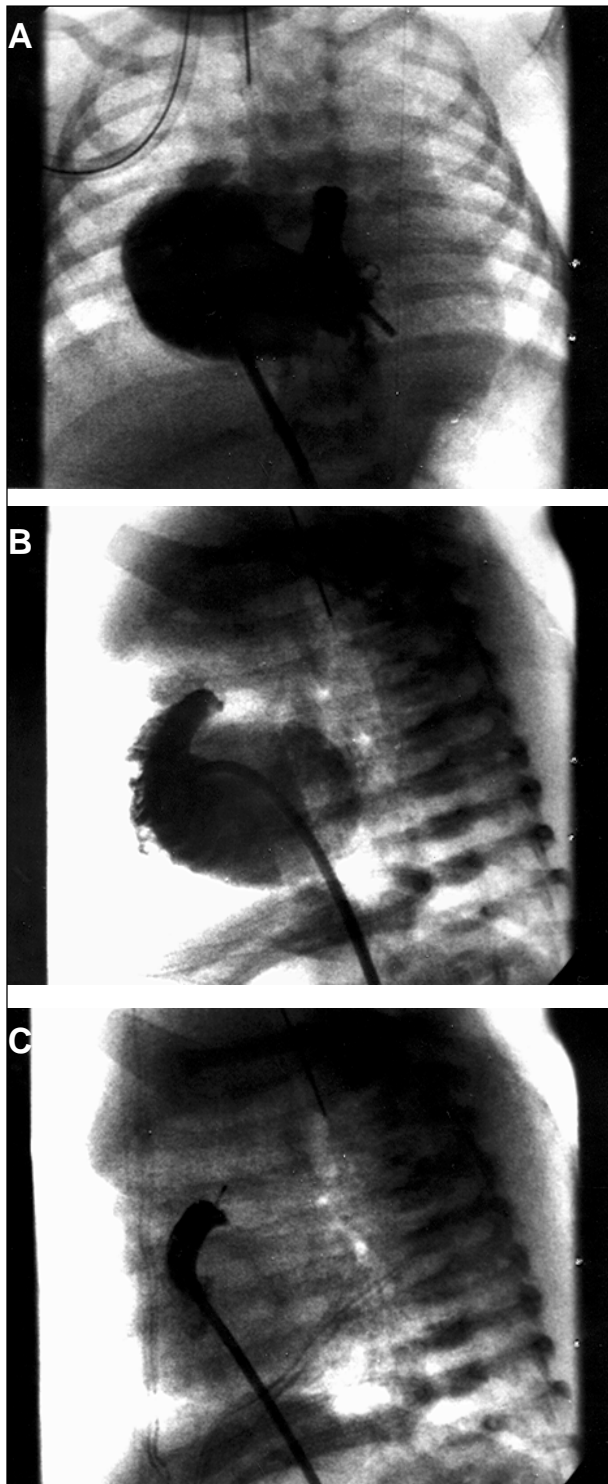| Table IV – Hemodynamic data (in mm Hg) after pulmonary valve perforation			
Patient	PuA	Infund	RV
1	20-8	20-8	50-8
2	35-15	35-8	40-8
3	26-12	32-8	62-8
PuA- pulmonary artery; Infund- infundibulum; RV- right ventricle			

Fig. 2 - Case 2 - A and B) right ventriculography in cranial posteroanterior and left profile views: Note mild to moderate right ventricular hypoplasia with attenuation of the trabecular zone and well-formed infundibulum. Significant tricuspid insufficiency exists; B and C) in left profile view, note the radiofrequency catheter located right below the valvar plane, with immediate progression to the pulmonary trunk after application of radiofrequency energy.

guide catheter [inner diameter 0.024", outer diameter 0.035", and length 145 cm (BMC)] was advanced to the pulmonary trunk, with confirmation of its position by manual injections of the contrast medium through the it (fig. 2 D). The radiofrequency catheter was then replaced by a 0.014" steerable coronary guidewire that was advanced through the arterial duat and positioned in the distal part of the descending aorta. The coaxial injectable catheter was then withdrawn, and a 3.0X20 mm angioplastic balloon catheter (World-Pass, Cordis) followed by a 9X20mm Tyshak II low-profile balloon catheter (Numed) completed the pulmonary valvoplasty (fig. 2 E). The hemodynamic control revealed an infundibular gradient of 5 mm Hg and the absence of a gradient through the pulmonary valve (tab. IV). Control ventriculography showed the establishment of right ventricle-pulmonary trunk continuity (figs. 2 F and G), with a minimum dynamic infundibular reaction. The postcatheterization evolution was uneventful. Prostaglandin was suspended
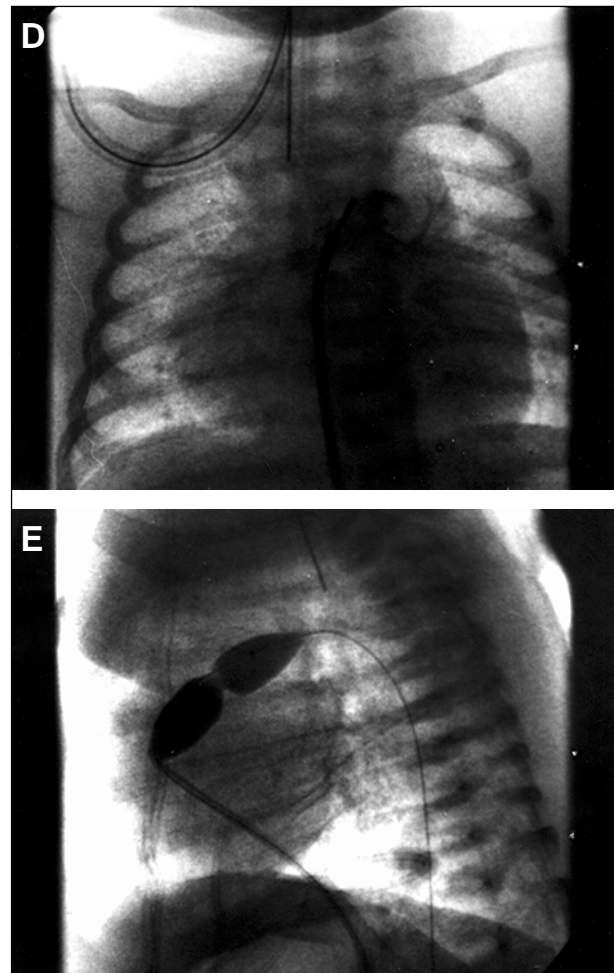

Fig. 2 - Case 2 - D) pulmonary trunk in posteroanterior view. The injectable guide catheter was advanced towards the pulmonary trunk over the Nykanen radiofrequency catheter, and its position was confirmed through a small injection of contrast medium, with mild opacification of the pulmonary arteries; E) after exchanging the radiofrequency catheter for a coronary guidewire and its progression to the descending aorta through the arterial canal, pulmonary valvoplasty was performed in a progressive way. Note the 9x20 mm Tyshak balloon being inflated at the level of the pulmonary valve for completion of the dilation.

energy for 2s at most. The valve was perforated at the first attempt, with progression of the catheter to the pulmonary trunk with no need to advance it manually (fig. 2 C). Over the Nykanen radiofrequency catheter, a coaxial injectable

after 4 days, saturation was maintained at around 80% (patient extubated), no metabolic acidosis was observed, and lactate levels were normal. The patient was discharged after 8 days. His echocardiogram showed a nonobstructive anterograde flow through the pulmonary valve and a maximum systolic gradient of around 15mmHg secondary to a mild infundibular reaction. Moderate to severe pulmonary insufficiency and minimum flow through the arterial duct existed. After a 3-month follow-up, the patient is clinically well, is gaining weight, and saturation is around 85%. The patient awaits a new echocardiogram and catheterization to define further management.

Case 3 – In this case, the procedure was performed in the same way as in the previous patient. However, 2 applications of 5W of energy during 2s were required for valvar perforation. A 2.5X20mm angioplasty balloon (World Pass, Cordis) was used, being followed by a 9X30mm Tyshak II low-profile balloon catheter (Numed) for completion of the pulmonary valvoplasty. Hemodynamic control revealed an infundibular gradient of 30mmHg and a valvar gradient of 6mmHg (tab. I). Control ventriculography showed the es-

tablishment of right ventricle-pulmonary trunk continuity, with an intense dynamic infundibular reaction, which led to the administration of 1mg/kg/day of propranolol. Five days after the procedure, we suspended prostaglandin, but had to reinstate it. A right Blalock-Taussig shunt was indicated. The patient HAO sepsis caused by *Staphylococcus aureus*, which responded well to antibiotics, and convulsive crises with the suspicion of paradoxical embolism secondary to deep venous thrombosis due to the presence of a central venous catheter in the femoral vein. The postoperative echocardiogram on the 30th day after catheterization revealed regression of the subpulmonary stenosis, a residual gradient of 9mmHg through the right ventricular outflow tract, and severe pulmonary insufficiency. The right ventricle developed, and the tricuspid ring diameter increased to 11mm (Z value =0). Despite the satisfactory flow through the surgical anastomosis, mild stenosis was detected in the right pulmonary artery at its insertion. Saturation in the postoperative period ranged from 80% to 90%.

Discussion

Opening of the right ventricular outflow tract through interventional catheterization is gaining acceptance as the initial therapeutic modality for patients with pulmonary atresia and intact ventricular septum [10-17]. In patients who have tripartite right ventricle [4], with a patent infundibulum [18], moderate ventricular hypoplasia at most, and coronary circulation not depending on the right ventricle [6-8], this technique promotes efficient decompression of the ventricular cavity, stimulating its growth [14,19]. Therefore, some potential surgical complications, which may occur in this type of disease, may be avoided. Right heart failure with systolic-diastolic dysfunction is not infrequent after opening of the right ventricular outflow tract with a transannular patch + ventriculotomy, and it may hinder the anterograde flow to the lungs, worsening the right-to-left shunt through the oval foramen or the atrial septal defect [14,20,21]. In addition, reperfusion lesions usually observed after the use of extracorporeal circulation may be extremely noxious in these patients with potential or real histological anomalies in the coronary arteries [1,14].

The percutaneous techniques already described in the literature for establishing the right ventricle-pulmonary trunk continuity include the use of laser or radiofrequency energy and mechanical perforation [10-17]. Laser energy, despite being the first to be used, especially by British groups [11,12], was gradually abandoned due to its high cost, risks to the staff of the hemodynamics laboratory, and the difficulty of transporting it [17].

Mechanical perforation with the hard tip of a coronary guidewire was reported for the first time by Latson in 1991 [10], and since then occasional reports have been published [14]. Even though the low cost is an obvious advantage, this technique has several potential problems. The catheter positioned in the infundibulum right below the pulmonary valve tends to have its position modified due to rigidity of the

Fig. 2 - Case 2 -F and G) right ventriculography in posteroanterior and left profile views after the procedure: Note establishment of the right ventricle-pulmonary artery continuity with minimum sub-pulmonary reaction and reduction in tricuspid insufficiency.

hard tip of the guidewire when the latter is advanced [14]. In addition, manual modeling of the hard tip may not be precise, leading to errors at the site of the transvalvular puncture [14]. In our first case, the pulmonary leaflets were very thin and offered no resistance to the progression of the hard tip of the guidewire. Thicker fibrotic valves may require greater strength for perforation, increasing the possibility of accidents and complications. In the event of accidental perforation of the muscular portion of the right ventricular outflow tract, or even of the pulmonary trunk, the occurrence of hemopericardium is a possibility. However, due to the reduced inner diameter of the guidewire (0.014"), significant bleeding is unlikely [14]. Another limitation of this technique is the need to withdraw the hard tip of the guidewire after the initial perforation, a new passage through the diminished orifice created being mandatory. This maneuver requires patience of the operator and is not always effective.

The use of radiofrequency energy for perforation of atretic pulmonary valves became a more popular method among pediatric interventionists because of its availability and more accessible costs in large centers [13-16]. Even though the most commonly used system is the Osypka [14], the Nykanen system reported about in this article offers several advantages. First, it is a radiofrequency system specifically designed to perforate biological tissues, and it releases a high amount of energy in an extremely localized and specific location, therefore, against great impedance. Perforation is not caused by an increase in local temperature, but secondary to an alteration in intracellular electrical charges, resulting in a very well localized cellular necrosis [22,23]. The Osypka system was not originally designed for this purpose. Because it is used for ablation of arrhythmias, the system functions with low impedance, causing tissue lesions due to the generation of local heat. Consequently, the lesion is more superficial and extensive, which may be an advantage in cases of ablation, but not when the major objective is to obtain a localized perforation. Even though our team has already successfully used the Osypka system for pulmonary valve perforation in case 1 [16], in another case (unpublished data), we could not pass beyond the valve with the guidewire after the initial perforation. Because the Nykanen system is coaxial, such a limitation does not exist. After an initial perforation, the radiofrequency catheter serves as a support for the advancement of the coaxial injectable guide catheter to the pulmonary trunk. Then the radiofrequency catheter itself may be maneuvered to pass beyond the arterial ducts and reach the descending aorta, or it may be replaced by a steerable coronary guidewire, as performed in both cases. This provides more control, safety, and rapidity to the procedure, avoiding the need to pass beyond the pulmonary valve again after the initial perforation, which is a fundamental advantage in the management of a cyanotic and critically ill newborn infant. In addition, the system is easy to use and handle for the professional used to interventions in pediatric cardiology. The cost of the catheters and system may be a limiting factor in our environment.

After percutaneous valvar opening, the next step in the therapeutic algorithm of this defect is the discontinuation of prostaglandin, which should be tried within the first week after catheterization [14]. Systemic saturation above 75%-80% and the absence of metabolic acidosis with normal levels of lactates usually indicate satisfactory anterograde pulmonary flow, which depends directly on improvement in right ventricular compliance. This phenomenon takes variable amounts of time to establish, and reliable predictors that this moment occurs or actually will occur do not exist [19]. Theoretically, significant hypoplastic ventricles are less compliant and do not manage to maintain an effective anterograde pulmonary flow. This sometimes is not confirmed in clinical practice, and to our surprise, suspension of prostaglandins is possible even under unfavorable anatomic conditions. This fact suggests that other mechanisms, in addition to the degree of ventricular hypoplasia, are implicated in diastolic dysfunction, including subclinical ischemia and myofibrillar disarray intrinsic to the underlying defect [1-3]. In addition, defining the degree of right ventricular hypoplasia is difficult in clinical practice. To this end, some authors use the Z value of the tricuspid valve [6], while others use a mere subjective estimate [17]. If no success is obtained in the initial suspension of prostaglandin, theoretically the prolonged administration of this drug, including its oral administration, is an alternative, but not feasible in our environment. Others suggest stent implantation in the arterial canal in the initial valve perforation procedure [15]. From our point of view, if the patient does not respond well to the attempt to suspend prostaglandins after 1 week, the Blalock-Taussig shunt is indicated because it is simpler and has more predictable postoperative results. This type of therapeutic algorithm in stages reflects another advantage of the initial approach by the percutaneous route in these patients. Surgery would only be indicated after failure in suspending the infusion of prostaglandin. In centers where surgical opening of the right ventricular outflow tract is the initial therapeutical option, the medical team faces the clinical dilemma of defining, beforehand, which patient will require an additional source of pulmonary flow in the same surgical stage. They risk performing an unnecessary shunt or having to perform it in a second stage, increasing the morbidity and mortality inherent in performing 2 surgical procedures in a critically ill patient.

Both percutaneous and surgical valvar openings, mainly when the latter is accomplished by transannular patch, result in severe pulmonary insufficiency as observed in our patients. This finding has always been classically considered benign in nature. However, recent evidence suggests that pulmonary insufficiency may be extremely noxious to the right ventricle, with a negative impact both on systolic and diastolic functions, mainly if accompanied by residual obstruction of the outflow tract [20,21,24]. Therefore, strict follow-up of right ventricular function is crucial in these patients.

After initial percutaneous valvar perforation, followed or not by a systemic-pulmonary shunt, the patient with pul-

monary atresia and an intact ventricular septum should undergo serial echocardiographies and a new catheterization at the approximate age of 12 months [15,25]. On that occasion, the degree of growth and improvement in the diastolic function of the right ventricle should be evaluated, and the cardiac output after temporary occlusion of the oval foramen or of the atrial septal defect should be calculated, as should the Blalock-Taussig shunt, if present. If cardiac output is maintained after test occlusion of the defects, these may be occluded in the same procedure by the percutaneous route with different intravascular devices [15,25], advancing the perspective that some patients with such a complex anomaly may be entirely treated without surgery [15]. If the degree of development of the right ventricle is not satisfactory with a reduction in cardiac output, hypotension (>20% of the base line), and a significant increase in right atrial pressure (>15mmHg) after test occlusion [15,25], the case should be individualized and the patient should be treated according to an algorithm for univentricular correction or that for a 1.5 ventricle [6,9].

In summary, we report the cases of 3 patients with pulmonary atresia and intact septum, in which the pulmonary valve was opened in the neonatal period through a percutaneous route with new techniques that proved effective. The coaxial system of perforation using radiofrequency designed by Nykanen is easy to handle, adding simplification and increased safety to a procedure formerly considered high risk.

Acknowledgments

Addendum

After writing this article, we learned of another newborn infant who had pulmonary atresia and intact ventricular septum with clinical, morphological, and functional characteristics similar to those of the above reported patients and who underwent an attempt at valvar perforation with the hard tip of the steerable coronary guidewire, because the radiofrequency system was not yet definitively available in our service on that occasion. In this latter case, the infundibulum had mild to moderate hypoplasia and the pulmonary valve was thicker and probably fibrotic, which made mechanical perforation unfeasible. Unintentional perforation of the right ventricular outflow tract occurred twice with minimum extravasation of the contrast medium to the pericardium, not causing tamponade or hemodynamic impairment. The newborn infant was maintained on prostaglandin infusion with oximetric, metabolic, and hemodynamic stability, and was referred for surgical pulmonary valvotomy and a right Blalock-Taussig shunt 2 days after catheterization. The patient HAO low cardiac output that did not respond to volume and usual vasoactive drugs, and died on the 7th postoperative day. This outcome confirms the previous comments made in this article.

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