Guided Transcatheter Valvulotomy in Pulmonary Atresia With Intact Ventricular Septum

Juan Alcibar,[a] Alberto Cabrera,[b] Natividad Peña,[a] Christian Baraldi,[c] Josune Arriola,[a] and José Aramendi[c]

[a]Sección de Hemodinámica, Servicio de Cardiología, Hospital de Cruces, Baracaldo, Vizcaya,
[b]Sección de Cardiología Pediátrica, Hospital de Cruces, Baracaldo, Vizcaya,
[c]Servicio de Cirugía Cardíaca, Hospital de Cruces, Baracaldo, Vizcaya, Spain.

Transcatheter valvulotomy in pulmonary atresia with an intact ventricular septum can be used as a first step to create biventricular circulation and to stimulate further development of the hypoplastic right ventricle. We describe our experience in a case of a neonate with this congenital cardiac defect who underwent successful transcatheter perforation of the atretic pulmonary valve. This report highlights the utility of a special technique based on the use of a gooseneck snare positioned just above the atretic valve to guide the advance of a coronary guidewire. Other therapeutic alternatives are considered.

Key words: *Pulmonary atresia. Valvulotomy. Intact ventricular septum.*

Full English text available at: www.revespcardiol.org

Valvulotomía mecánica percutánea dirigida en la atresia pulmonar con septo íntegro

La valvulotomía percutánea con catéter en la atresia pulmonar con septo íntegro puede ser el primer paso para establecer una circulación en serie con el posterior desarrollo del ventrículo derecho. Presentamos nuestra experiencia en un neonato con esta cardiopatía, al que realizamos una apertura mecánica, haciendo especial referencia a la técnica empleada con guía especial para desobstrucción coronaria y dirigida con catéter-lazo abierto sobre la válvula pulmonar atrésica. Se exponen otras técnicas alternativas y se discute la evolución de esta paciente en función de su desarrollo anatómico.

Palabras clave: *Atresia pulmonar. Valvulotomía. Septo íntegro.*

INTRODUCTION

Pulmonary atresia with an intact ventricular septum (PAIVS) is a serious heart condition that accounts for 0.7% to 3.1% of all cases of congenital heart disease.[1] Except in patients with severe right ventricular hypoplasia or right ventricle-dependent coronary circulation, establishing right ventricle-pulmonary artery continuity may be the first step to achieving biventricular circulation, as indicated in the guidelines for clinical practice and invasive techniques in pediatric cardiology.[2] To this aim, several techniques have been described for perforation of the atretic membrane. Early efforts were mechanical, using a conventional guidewire.[3-6] Later techniques involved laser[7] and radiofrequency,[5,6,8-11] followed by conventional balloon valvuloplasty.

To limit the possibility of cardiac perforation, which has been reported with all of these techniques, we performed transcatheter perforation in a newborn with this severe heart defect, using a special coronary guidewire guided by an open gooseneck snare positioned just above the atretic valve, followed by valvuloplasty.

CLINICAL CASE

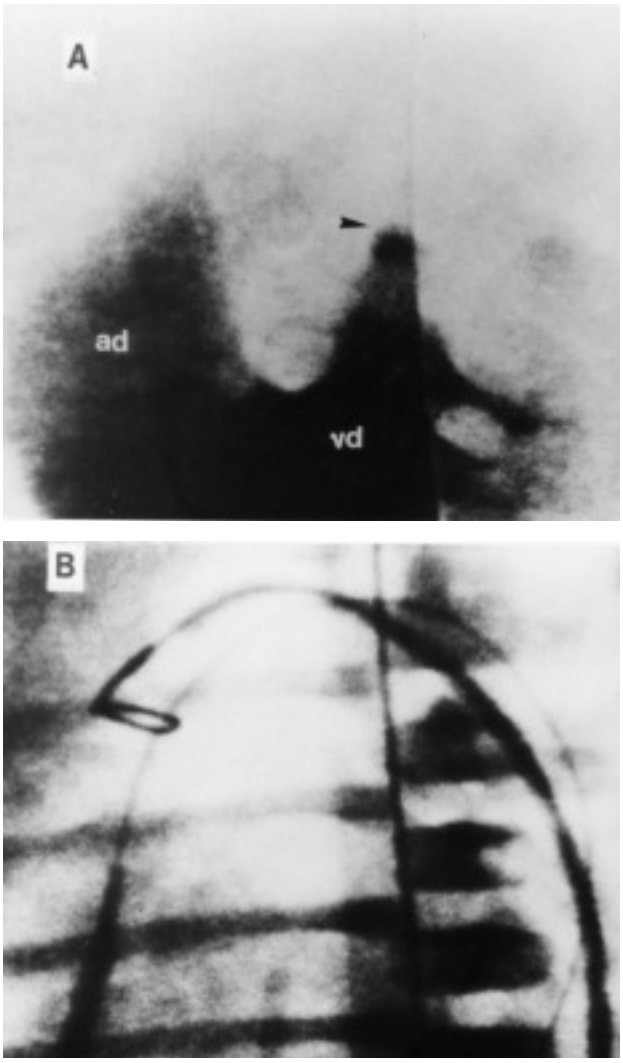
A term neonate born to a primiparous mother was admitted to our hospital for cyanosis 18 hours after birth. The examination disclosed a weight of 3580 mg, generalized cyanosis and 50% saturation at 100% $Fi0_2$, normal pulses, blood pressure 60/37 mm Hg, and heart rate 130 bpm. On auscultation, a mild functional murmur was detected at the left sternal border, with a single second sound. The ECG disclosed sinus rhythm with QRS axis +100º and right atrial growth. Heart disease was diagnosed by Doppler ultrasound, which showed reduced tricuspid mobility, an 8.5 mm annulus (Z=–3.5), anterograde flow and severe regurgitation, with a gradient of 130 mm Hg. The atretic pulmonary valve presented a favorable membranous morphology

Correspondence: Dr. J. Alcibar Villa.
Avda. de Algorta, 60, 2.º D. 48990 Algorta. Vizcaya. España.

Received 14 May 2002.
Accepted for publication 26 March 2003.

ABBREVIATIONS

PAIVS: pulmonary atresia with intact ventricular septum.
PA: pulmonary artery.
RV: right ventricle.



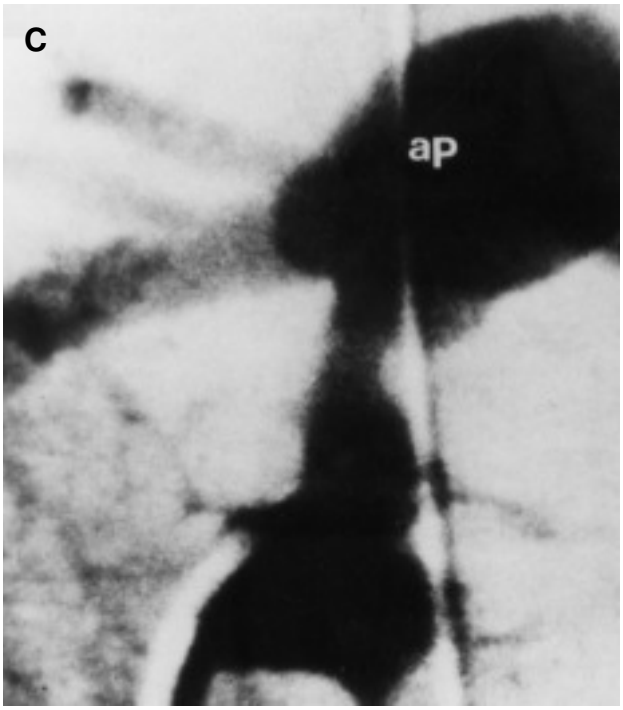


Fig. 1. A. Right ventriculography, anteroposterior view. Markedly underdeveloped right ventricle and pulmonary atresia; very severe tricuspid regurgitation. B. Guidewire passage: perforation of the valve, through the loop and ductus to the descending aorta. C. Valve opening. Contrast medium in the pulmonary artery appears clearly. Tricuspid regurgitation disappears.
PA indicates pulmonary artery; RA, right atrium; RV, right ventricle.

and a diameter of 8 mm; the pulmonary trunk and branches were well developed. The ventricular septum was intact, although the atrial septum showed a small, 4 mm right-to-left defect. Following the diagnosis of PAIVS, the patient was stabilized with prostaglandin E1 at a dose of 0.5 µg/kg/min and dopamine at 8 µg/kg/min, achieving 94% saturation.

On the third day of life, catheterization was performed by right femoral arterial/venous puncture and (with the systemic saturation levels indicated above) manometry data were as follows: right atrium: a, 14 mm Hg; mean, 10 mm Hg; right ventricle, 110/0-15 mm Hg; left atrium: mean, 9 mm Hg; left ventricle: 64/0-9 mm Hg. Left anteroposterior ventriculography disclosed a morphologically normal ventricle and a left aortic arch, clearly visible anterograde opacification of the coronary arteries and, by the ductus, well-developed pulmonary atresia. Right ventriculography demonstrated a markedly underdeveloped tripartite right ventricle with valvular dimensions indicated above, pulmonary atresia and severe tricuspid regurgitation (Figure 1A). The sinuses were not visualized and the coronary arteries showed no contrast enhancement by dependent circulation. Valvotomy was decided on the basis of these findings. Using the femoral artery approach, the catheter was advanced to the arch by retrograde direction and a 4 Fr Judkins right catheter was placed in the ductus. A 5 mm (microvein) Amplatz Goose Neck snare loop was then advanced through the catheter, the open loop was placed over the atretic valve and the coronary catheter was withdrawn to the descending aorta. Another 5 Fr Judkins right catheter was then introduced through the femoral vein, but it could not be properly positioned in the infundibulum. It was exchanged for a 5 Fr balloon wedge pressure catheter (Arrow International), which was coaxially placed in the infundibulum with its distal

orifice close to the valve. Then, guided by the open snare loop as a «target» and with rotating movements over the valve, a 0.014 inch PT Graphix Standard coronary guidewire (Boston-Scimed) was advanced; once past the barrier, it was inserted inside the loop until it reached the descending aorta through the ductus (Figure 1B). In order to gain support and stability, the guidewire was caught and withdrawn in the femoral artery, and with this thrust the valve was dilated with a 3×20 Maverick balloon catheter (Boston-Scimed) to 8 atmospheres, and subsequently with a Balt 8×20 (Montmorency) to 6 atmospheres, observing significant indentation with both maneuvers. After opening the valve, right ventricular systolic and diastolic blood pressure decreased (50/0-7 mm Hg) and the pulmonary valve gradient was 10 mm Hg. Right ventriculography confirmed free passage of contrast to the pulmonary artery and an important reduction in tricuspid regurgitation (Figure 1C).

Mechanical ventilation was maintained during the following days, prostaglandins were reduced, and there was an expected decrease in saturation. Twenty days after the valvotomy, a trunk-to-trunk systemic/pulmonary shunt and ligation of the ductus were performed. The infant remained stable with saturation levels of 90% and was discharged six days later. Doppler ultrasound prior to discharge disclosed good pulmonary flow, with a gradient of 32 mm Hg, slight pulmonary regurgitation, absence of tricuspid regurgitation and good functioning of the shunt.

DISCUSSION

Transcatheter valvotomy in PAIVS requires favorable anatomical conditions: a tripartite right ventricle with a moderate degree of hypoplasia allowing subsequent development, a patent infundibulum, and a membranous-type valvular atresia with anatomical continuity. As in our patient, experience has shown that transcatheter valvotomy is a definitive treatment in only 50% of cases; systemic-pulmonary shunt and/or infundibulum resection is required.[4-6] In this regard, we were aware of the significantly underdeveloped ventricle in our patient, and valvular perforation was attempted as a palliative measure before creating a shunt. Based on the extensive experience of Jou-kou Wang et al,[5] transcatheter valvotomy was the definitive treatment in PAIVS, with a tricuspid valve Z value ≥ –0.1, pulmonary valve Z value ≥-4.1 and right-to-left ventricular area ratio ≥0.65. All patients with tricuspid valve Z value ≤–0.8 and pulmonary valve Z value ≤–4.2 with a right-to-left ventricular area ratio ≤0.54 required shunting and/or enlargement of the ventricular outflow tract. Another fundamental anatomic condition is the exclusion of right-ventricle dependent coronary circulation.[4-10] The presence of anatomical coronary obstructions and supplementary sinusoid circulation would trigger catastrophic ischemia with valve opening and decompression of the right ventricle. For this reason, the coronary arteries should be studied carefully by aortography or left ventriculography.

Since the pioneer work by Latson[3] in the early 1990s, pulmonary valvotomy has been performed mechanically,[3-6] with laser[7] or radiofrequency methods,[5,6,8,10,11] using a conventional catheter or a special 2 Fr Coe catheter[9] and through anterograde[8] or transductal retrogrades approaches.[11]

Although the success rate with the mechanical technique is lower,[5] we believe outcome can be improved by using a special guidewire for treating obstructive coronary conditions and directing the perforation with a gooseneck snare. We also consider this technique to be simpler than the use of repeated contrast in the infundibulum or echocardiographic monitoring.[5,6] In our setting, we are accustomed to treating chronic obstructive coronary disease. Here, we have attempted to adapt the material for use in PAIVS and in other severe obstructive heart diseases. The standard PT Graphix guidewire is used in patients with complex anatomy requiring maximum thrust to cross over the lesion, as in cases of total occlusion and coronary revascularization. The guidewire is slightly rigid at its distal end, and when rotated in a clockwise direction, perforates the valve. Its special ICE hydrophilic coating buffers vascular friction and allows it to glide easily through the ductus to the descending aorta. It can then be retrieved and withdrawn, as has been previously shown.[5,6,8,11]

Based on the experience described, and contemplating the application of this technique in less complex patients, we suggest that mechanical valvotomy using a special guidewire for obstructive coronary conditions and targeted by a transductal gooseneck snare may be an effective procedure to establish right ventricle-pulmonary artery continuity in anatomically indicated cases of PAIVS.

IN MEMORIAM

To Pablo Martínez Corrales, cardiac surgeon and friend, provider of invaluable help and encouragement in interventional pediatrics

REFERENCES

1. Freedom RM. Etiology and Incidence. En: Freedom RM, editor. Pulmonary atresia with intact ventricular septum. Mount Kisco: Futura, 1989; p. 1-8.
2. Alcibar Villa J, García Fernández E, Gutiérrez-Larraya Aguado F, Moreno Granado F, Pan Álvarez-Osorio M, Santos de Soto J. Guías de actuación clínica de la Sociedad Española de Cardiología. Requerimientos y equipamiento de las técnicas inva-

sivas en cardiología pediátrica: aplicación clínica. Rev Esp Cardiol 1999; 52:688-707.
3. Latson LA. Nonsurgical treatment of a neonate with pulmonary atresia and intact ventricular septum by transcatheter puncture and balloon dilatation of the atretic valve membrane. Am J Cardiol 1991;68:277-9.
4. Shimpo H, Hayakawa H, Miyake Y, Takabayashi S, Yada I. Strategy for pulmonary atresia and intact ventricular septum. Ann Thorac Surg 2000;70:287-9.
5. Wang JK, Wu MH, Chang CI, Chen YS, Lue HC. Outcomes of transcatheter valvotomy in patients with pulmonary atresia and intact ventricular septum. Am J Cardiol 1999;84:1055-60.
6. Justo RN, Nykaneu DG, William WG, Freedom RM, Benson LN. Transcatheter perforation of the right ventricular outflow tract as initial therapy for pulmonary valve atresia and intact ventricular septum in the newborn. Cathet Cardiovasc Diagn 1997;40:408-13.
7. Qureshi SA, Rosenthal E, Tynan M, Anjos R, Baker EJ. Transcatheter laser-assisted balloon pulmonary valve dilation in pulmonic valve atresia. Am J Cardiol 1991;67:428-31.
8. Cheatham JP, Coe JY, Kugler JD, Fletcher SE, Tower AJ. Successful transcatheter perforation of the atretic pulmonary valve membrane in a newborn using the new Coe radiofrecuency end hole catheter. Cathet Cardiovasc Diagn 1998;45:162-6.
9. Coe JY, Timinsky J, Villnave D, Trevenen C. Transductal approach to establish pulmonary connection in pulmonary atresia using a new radiofrecuency catheter in lambs [abstract]. Circulation 1997;96:373.
10. Camino M, Brugada J, Mortera C, Thio M, Rovirosa M, Bartrons J. Valvulotomía pulmonar percutánea mediante radiofrecuencia en la atresia pulmonar con septo interventricular íntegro. Rev Esp Cardiol 2001;54:243-6.
11. Hijazi ZM, Patel H, Cao QL, Warner K. Transcatheter retrograde radio-frecuency perforation of the pulmonic valve in pulmonary atresia with intact ventricular septum, using a 2 French catheter. Cathet Cardiovasc Diagn 1998;45:151-4.