

Percutaneous Balloon Pulmonary Valvuloplasty in a 40-Year Old Woman with Cyanosis

JULIO C. ECHARTE MARTÍNEZ¹, FRANCISCO L. MORENO-MARTÍNEZ², JUAN VALIENTE MUSTELIER³

Received: 01/17/2011

Accepted: 02/21/2011

Address for reprints:

Dr. Francisco L. Moreno-Martínez
Hemodynamics and Interventional
Cardiology
Cardiocentro Ernesto Che Guevara
Gaveta Postal 350, CP 50100
Santa Clara, Villa Clara, Cuba
e-mail: flmorenom@yahoo.com
Personal Phone: +53 5 2944651

SUMMARY

Isolated pulmonary valve stenosis is uncommon. As the diagnosis and treatment of this condition is made during childhood, it is a rare finding in adult patients. We describe the case of a 40-year old female patient with dyspnea during slight exertion, central cyanosis and nail clubbing with a critical pulmonary valve stenosis and right-to-left shunt through a patent foramen ovale. The pulmonary valve gradient was 150mm Hg. Percutaneous pulmonary valvuloplasty was performed using a Nucleus balloon catheter (Numed Cardiac Diagnostics). Immediately after the procedure, the pulmonary gradient measured by cardiac catheterization and echocardiography decreased to 23mm Hg. Six months later the cyanosis had disappeared and the gradient was 19mm Hg. The patient lives a normal life with no limitations in her quality of life and lifestyle.

REV ARGENT CARDIOL 2012;80:xxx-xxx.

Key words >

Pulmonary Valve Stenosis - Percutaneous Pulmonary Valvuloplasty

Abbreviations >

PVS	Pulmonary Valve Stenosis	RV	Right Ventricle
PAT	Pulmonary Artery Trunk	PPV	Percutaneous Pulmonary Valvuloplasty

BACKGROUND

Isolated pulmonary valve stenosis (PVS) occurs in 7-12% of all congenital heart defects and in 80-90% of all outflow tract obstruction of the right ventricle (RV).⁽¹⁾ Its legacy ranges between 1.7 % and 3.6% .⁽¹⁾

The need for mechanical relief is often resolved in childhood, due to the scientific and technological advances and early diagnosis, so that it is becoming increasingly difficult to find this type of disease in adulthood. Years ago, surgical treatment was the choice, however, with the development of interventional cardiology, nowadays very few patients need it. ^(1,2)

Patients with severe stenosis may have dyspnea and decreased exercise capacity, therefore have a worse prognosis if they are not treated promptly. ⁽¹⁾

In this presentation is described the case of a woman with PVS diagnosed at birth, due to the natural evolution of the disease, RV hypertrophy appeared and pressures increased retrogradly in the atrium of the same side, which achieved to break up the foramen ovale and produced a right-to-left shunt, making an acyanotic disease in a cyanotic one.

CASE REPORT

A Caucasian, 40 years old patient with a previous diagnosis of congenital PVS that at income presented

dyspnea before little effort, central cyanosis and nail clubbing. She had 4/6 holosystolic heart murmur, with epicenter on the high left sternal border and the electrocardiogram showed signs of growth in the right cavities with QRS> 120 msec. The echocardiogram evidenced severe ventricular hypertrophy of the RV (17mm in the anterior wall), with diastolic diameter of 29mm, thickening of the pulmonary semilunar, without calcium, and high transvalvular gradients (peak of 150 mmHg and mean of 79 mmHg); there was also severe tricuspid regurgitation and right-to-left shunt through the foramen ovale, which was permeable. The pulmonary artery trunk (PAT) was normal.

The hemoglobin was 22.6g/dl and she had required previous multiple blood samples. The remaining humoral determinations showed glucose 5.1 mmol/L, creatinine 101 mmol/L, glutamic pyruvic transaminase 26.4 IU, cholesterol 3.22 mmol/L and triglycerides 1.25 mmol/L, the partial pressures of oxygen, measured during the procedure at different levels were greatly diminished (Table 1).

It was decided to carry out a percutaneous pulmonary valvuloplasty (PPV). It was carried out a puncture of the artery and right femoral vein. In the artery was left a 5 Fr introducer for taking blood samples for oximetries in the aorta before and after

¹ Interventional Cardiologist. Instituto de Cardiología y Cirugía Cardiovascular. La Habana, Cuba

² Interventional Cardiologist. Cardiocentro Ernesto Che Guevara. Villa Clara, Cuba

³ Echocardiographer. Instituto de Cardiología y Cirugía Cardiovascular. La Habana, Cuba

the procedure, like for monitoring systemic blood pressure. In the femoral vein was placed a 8 Fr introducer to specify the oximetries, as well as the record of pressures on all lower circuit, and to measure pulmonary transvalvular gradient. In angiocardioqram in the RV, in lateral view, which was carried out using a 6 Fr pigtail catheter, Cordis, radiological signs of PVS (Figure 1A) were confirmed. The pulmonary annulus (19mm) was measured in lateral view. Afterwards, with a Judkins' catheter of right coronary (Cordis, Johnson & Johnson), and using a straight guide, it was able to access the left branch of the pulmonary artery. The pulmonary transvalvular gradient was 150 mmHg, which coincided with the value found on the echocardiogram, the right chambers pressures were increased, and in PAT the systolic, diastolic and mean pressures were 20/10/12 mm Hg (table 1).

With an Amplatzer guide, it was achieved an excellent support for the placement of a Nucleus balloon catheter of Numed Cardiac Diagnostics, 23 × 60mm. Four inflated were needed (Figure 1, lower panel), but the result was optimal (Figure 1 B). The gradient measured by catheterization and echocardiogram (ALOKA Prosound Alpha 10), immediately after the procedure, was 23 mmHg. There were not complications, just a syncope after the first inflated due to an infundibular spasm, which was relieved with the administration of 2 mg of intravenous propranolol.

The patient was transferred the same day of the procedure to her hospital of origin, where she evolved favorably and was discharged six days later with 60 mg/day of propranolol and antiplatelet agents.

Today, after 6 months of the PPV, cyanosis has disappeared (Figure 2) and the echocardiogram shows a pulmonary transvalvular gradient of 19 mmHg, absence of an interatrial shunt and disappearance of

tricuspid regurgitation. The patient leads a normal life without limitations in her quality of life.

DISCUSSION

Although this type of adult patients do not appear frequently in clinical practice, they are not a rarity because they respond to the lack of early attention of the disease. In this particular patient surgery was proposed during childhood, because at that time in Cuba there was no current development of interventional techniques, but her parents refused. Clearly, the patient experienced the natural evolution of a severe PVS, which was not treated opportunely.

The interest of this case, besides the success of the procedure is that it is uncommon for a PVS, severe since childhood, reaches adulthood without need of treatment (percutaneous or surgical); plus, the right-to-left shunt was resolved spontaneously once right atrial pressures decreased and tricuspid regurgitation disappeared.

In the bibliography there is a similar case to ours: it is about a 65 years old woman, in whom, besides the pulmonary valvuloplasty, it was necessary to close the foramen ovale in a second percutaneous intervention. (3)

According to Rao, (2) since Kan described for first time PPV in 1982, this procedure has been recommended and used widely for the treatment of PVS. For years it has been considered as an excellent therapeutic option (4) and is the treatment of choice for patients with this disease (1,2,5) even in the presence of infundibular stenosis or severe tricuspid regurgitation. (6)

Rao himself, (2) in 2007, said that it was recommended for patients with a maximum gradient greater than 50 mmHg, however, in the most recent guidelines of the European Society of Cardiology (1) is proposed that the obstruction RV outflow tract, at any level, should be repaired regardless of symptoms when the maximum Doppler gradient is greater than 64 mmHg (maximum speed > 4 m/sec), whenever RV function is normal and valve replacement is not needed. And they also point out that intervention in patients with a gradient less than 64 mmHg should be considered only in the presence of: a) secondary symptoms to pulmonary stenosis, b) decreased RV function, c) RV with double chamber, d) important arrhythmias or e) right-to-left shunt by atrial or ventricular septal defect. (1)

Fawzy et. al (6) argue that the evolution and prognosis of these type of patients after valvuloplasty depend on the location (valvular or infundibular) of residual gradient and the magnitude of it, tricuspid regurgitation disappears and the long-term results are excellent.

In our patient, the pulmonary transvalvular gradient continued decreasing gradually after the procedure and, at six months, it was 18 mmHg. As Gupta et. al affirm, (7) after a successful valvuloplasty

Table 1. Right pressure and oximetric determinations before and after valvuloplasty.

Variable	Before	After
Pressure (mmHg)		
Right atrium	15	10
Right ventricle	170/15	42/10
PAT (systolic/diastolic/mean)	20/10/12	20/10/13
Transvalvular gradient	150	23
Oxygen in blood (mmHg)		
Right femoral vein	30.5	34.1
Superior vena cava	30.4	35.5
Mean right atrium	29.9	35.7
Right ventricle	30.0	35.7
PAT	25.3	35.4
Femoral artery	40.2	51.8
Aortic root	41.1	52.1

PAT: pulmonary artery trunk.

Fig. 1. A. RV angiography before valvuloplasty where is observed the passage of contrast for the minimum valve orifice (arrow).

B. Immediately after the procedure (the arrows show the apparent increase of the flow). 1, 2 and 3. Indicate the sequence of the first inflated balloon, which had the effect 'in watermelon seed'. 4. Totally relaxed balloon at valve plane level.

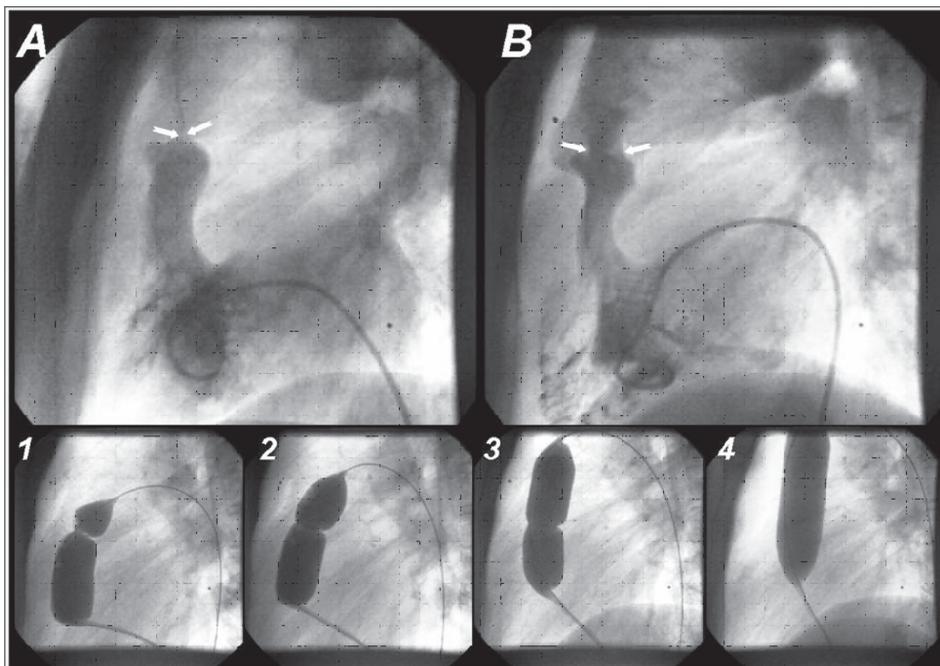


Fig. 1. Photographs with permission of the patient. **A.** Before valvuloplasty, with frank cyanosis. **B.** Without cyanosis, six months later.



the gradient may continue decreasing, even in those patients with high infundibular gradients.

The PPV is the treatment of choice in patients with severe PAT.

RESUMEN

Tratamiento percutáneo de estenosis valvular pulmonar crítica en una mujer de 40 años con cianosis

La estenosis valvular pulmonar aislada es poco frecuente. Se diagnostica y se trata casi siempre durante la infancia, razón por la cual es poco común encontrarla en la edad adulta. En esta presentación se describe el caso de una paciente de 40 años con disnea ante pequeños esfuerzos, cianosis central y uñas en vidrio de reloj, que presentaba una estenosis valvular pulmonar crítica con cortocircuito de derecha a izquierda a través del foramen oval. El gradiente transvalvular pulmonar era de 150 mmHg. Se realizó una valvulotomía pulmonar percutánea con un catéter balón Nucleus de Numed Cardiac Diagnostics. El gradiente medido por cateterismo y ecocardiograma inmediatamente después del procedimiento fue de 23 mmHg. Seis meses después había desaparecido la cianosis y el gradiente transvalvular pulmonar fue de 19 mmHg. La paciente lleva una vida normal, sin limitaciones en su calidad y estilo de vida.

Palabras clave > Estenosis de la válvula pulmonar - Valvulotomía pulmonar percutánea

BIBLIOGRAPHY

- Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, et al. Guía de práctica clínica de la ESC para el manejo de cardiopatías congénitas en el adulto (nueva versión 2010). *Rev Esp Cardiol* 2010;63:1484.e1-e59.
- Rao PS. Percutaneous balloon pulmonary valvuloplasty: State of the art. *Catheter Cardiovasc Interv* 2007;69:747-63.
- Combes S, Dauphin C, Motreff P, Joly H, Rodrigues R, Jean F, et al. Sudden transformation of pulmonary stenosis to trilogia of Fallot in a 65 year old patient. *Arch Mal Coeur Vaiss* 2005;98:586-9.
- Sharieff S, Shah-e-Zaman K, Faruqi AM. Short- and intermediate-term follow-up results of percutaneous transluminal balloon valvuloplasty in adolescents and young adults with congenital pulmonary valve stenosis. *J Invasive Cardiol* 2003;15:484-7.
- Fawzy ME, Awad M, Galal O, Shoukri M, Hegazy H, Dunn B, et al. Long-term results of pulmonary balloon valvulotomy in adult patients. *J Heart Valve Dis* 2001;10:812-8.
- Fawzy ME, Hassan W, Fadel BM, Sergani H, El Shaer F, El Widaa H, et al. Long-term results (up to 17 years) of pulmonary balloon valvuloplasty in adults and its effects on concomitant severe infundibular stenosis and tricuspid regurgitation. *Am Heart J* 2007;153:433-8.
- Gupta D, Saxena A, Kothari SS, Juneja R. Factors influencing late course of residual valvular and infundibular gradients following pulmonary valve balloon dilatation. *Int J Cardiol* 2001; 79:143-9.