# Successful percutaneous balloon valvuloplasty in a preterm infant weighing 1500 g with critical pulmonary valve stenosis

M.W. Freund, T. Schouten, P. Lemmers, C. Schroer, J. Strengers

We describe a successful cardiac intervention in an infant, born at 32 weeks of gestation, with a birth weight of 1040 g, who had a critical pulmonary valve stenosis with the right ventricular pressure twice the systemic pressure. Continuous prostaglandin E infusion kept the arterial duct open and at the age of four weeks and a weight of 1500 g a balloon valvuloplasty was performed, which reduced the systolic right ventricular pressure to below the systemic pressure. Two weeks later the procedure was repeated because of increasing right ventricular pressure. At two months of age the mean systolic transpulmonary Doppler gradient was 30 mmHg with an adequate right ventricular volume. The neurological status of the infant was normal and the femoral vein was patent. Timely interventional heart catheterisation is a successful method of treatment in critical pulmonary valve stenosis in infants with a low birth weight. (Neth Heart J 2008;16:264-6.)

Keywords: pulmonary valve stenosis (critical), low birth weight, intervention, complication

# M.W. Freund

# J. Strengers

Department of Paediatric Cardiology, Wilhelmina Children's Hospital, University Medical Center Utrecht, the Netherlands

# T. Schouten

Department of Anaesthesia, Wilhelmina Children's Hospital, University Medical Center Utrecht, the Netherlands

# P. Lemmers

Department of Neonatology, Wilhelmina Children's Hospital, University Medical Center, the Netherlands

# C. Schroe

Department of Paediatrics, Maxima Medical Centre, Veldhoven, the Netherlands

Correspondence to: M.W. Freund Department of Paediatric Cardiology, Wilhelmina Children's Hospital, University Medical Center Utrecht, PO Box 85090, 3508 AB Utrecht, the Netherlands E-mail: m.w.freund@umcutrecht.nl **C** ritical pulmonary valvular stenosis (PS) in infancy is a ductus-dependent congenital heart disease. The open ductus arteriosus warrants pulmonary circulation in critical PS. Balloon valvuloplasty (BVP) is now the first therapeutic option.<sup>1-3</sup> It was first described in 1983 to 1984.<sup>4,5</sup>

However, interventional heart catheterisation in low birth weight infants may be achievable, 6,7 but only a few cases of balloon valvuloplasty in infants weighing 1500 g or less are described in the literature<sup>6,8</sup> with an acceptable mortality but still moderate morbidity. Critical pulmonary valve stenosis with suprasystemic right ventricular pressure is an indication for therapeutic intervention to avoid severe right ventricular hypertrophy with involution of the right ventricular volume and continuous intravenous prostaglandin medication.9 Timely successful balloon valvuloplasty will therefore prevent extensive surgical therapy and long-lasting duct dependency of the pulmonary circulation. Beta-blocker therapy can treat the dynamic and reversible infundibular obstruction after pulmonary valve balloon dilatation.<sup>10</sup> Cardiac surgery in small preterm infants with a weight of less than 1500 g using a heart-lung machine is feasible but is accompanied with higher mortality and morbidity<sup>11,12</sup> than is described for interventional heart catheterisation.

We report on an infant, one of a triplet, born at 32 + 5 weeks of gestation with a birth weight of 1040 g and a length of 38.5 cm (BSA 0.11 m²). Due to a distinct systolic murmur, echocardiography was performed on day 2. Two-dimensional Doppler echocardiography revealed a critical valvular stenosis with a small pulmonary valve ring of 5.3 mm, moderate thickening of the pulmonary valve leaflets with severe doming (figure 1), a right ventricular systolic pressure of about 100 mmHg estimated by tricuspid valve regurgitation and an open arterial duct with left-toright shunting. Intravenous prostaglandin E was started to keep the ductus arteriosus (PDA) open and maintained a sufficient pulmonary circulation.



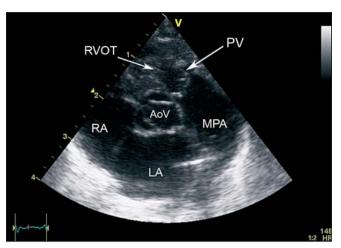


Figure 1. Parasternal short-axis view showing a hypertrophic right ventricular outflow tract, doming of the pulmonary valve (PV), poststenotic dilatation of the main pulmonary artery (MPA), an open aortic valve (AoV), and the left and right atrium (LA, RA).

At a weight of 1500 g and the age of four weeks we performed a balloon valvuloplasty under general anaesthesia. Via the right femoral vein a 3 French Braun (Braun Melsungen AG, Melsungen, Germany) sheath was percutaneously introduced via the Seldinger technique and exchanged with a 4 French Terumo (Tokyo, Japan) sheath. A 4 French right Judkins catheter (Cordis, Miami, USA) was advanced to the right ventricle. Systolic right ventricular pressure measured 105 mmHg and systemic systolic pressure 50 mmHg. A hand injection of 2 ml of contrast medium (Omnipaque 300) showed a tripartite right ventricle with a small right ventricular outflow tract. A very small jet via the severe PS into the dilated pulmonary artery trunk was visible (figure 2).

The severe pulmonary valve stenosis could not be crossed with a 0.021 inch wire (Cordis, Miami, USA); instead perforation of the right ventricular outflow tract occurred without further sequelae. With a 14 inch balanced middle weight wire (Guidant, Santa Clara,

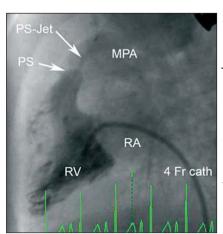


Figure 2. Right ventricular angiogram in the lateral projection. A 4 French Gensini catheter is inserted via the right atrium (RA) into the right ventricle (RV). In systole the pulmonary valve stenosis (PS) is shown with only a small jet of contrast medium tracing the dilated main pulmonary artery (MPA).

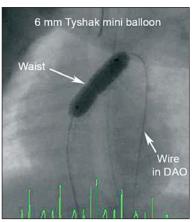


Figure 3. Dilatation of the pulmonary valve stenosis with a 6 mm Tyshak mini catheter. In the frontal projection, the waist of the balloon at the level of the pulmonary valve ring is seen. The wire is inserted via the pulmonary artery and the arterial duct in the descending aorta (DAO).

USA) the PS was crossed and positioned via the PDA in the descending aorta. The PS was dilated with a 5 and a 6 mm NuMed Tyshak mini balloon catheter, respectively, with 4 atmospheres (NuMed Cornwall, Canada) (figure 3).

After the procedure a decline in the systolic RV pressure to 50 mmHg and a rise of the systemic arterial pressure to 70 mmHg were noted. Prostaglandin E was continued because the right ventricle showed a dynamic infundibular pulmonary stenosis. Follow-up echocardiography demonstrated an increase in RV pressure. For that reason, a second BVP was performed with a 7 mm Tyshak mini catheter (132% of the pulmonary valve ring) via the same route with success and without complications two weeks later. The right femoral venous vein remains patent. Metoprolol was started to treat secondary infundibular obstruction (figure 4). Prostaglandin was discontinued three days later and the PDA closed spontaneously. Two months after the procedure the child's weight was 3200 g, neurological investigation and sonography of the brain were normal. Echocardiography revealed a mean transpulmonary systolic gradient of 30 mmHg, trivial pulmonary valve regurgitation, and in the short axis the ventricular septum was convex, confirming infrasystemic pressure in the right ventricle.

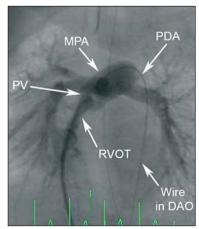


Figure 4. Angiography in the right ventricular outflow tract (RVOT) in the frontal projection. The RVOT shows a dynamic infundibular obstruction and a good antegrade flow via the pulmonary valve (PV) into the main (MPA) and left and right pulmonary artery. The wire is inserted from the MPA via the persistent ductus arteriosus (PDA) in the descending aorta (DAO).

## Conclusion

Interventional catheterisations in preterm infants with a weight of 1500 grams or less are feasible and can be performed safely in selected cases. Critical pulmonary valve stenosis is assumed to be the classical indication for balloon valvuloplasty in this age group and can be performed safely with success, without serious complications and seems superior to surgical intervention. Performing BVP early in critical pulmonary stenosis is justified to prevent severe right ventricular hypertrophy followed by loss of right ventricular compliance and right ventricular volume and to stop ductus dependency and to discontinue prostaglandin E. An essential condition for success is working in a team with paediatric cardiac interventionalists, paediatric cardio-anaesthetists and neonatologists.

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