

SHORT CASES IN CARDIOLOGY

Late onset pulmonary valvar stenosis after arterial switch operation for transposition of the great arteries

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The arterial switch operation (Jatene procedure) is the treatment of choice for transposition of the great arteries with intact ventricular septum (TGA-IVS) in neonatal age, because of the low mortality and morbidity during long term follow up.¹ Right ventricular outflow obstruction is the most frequent postoperative complication,²⁻⁴ occurring with sufficient severity to require reintervention in up to 10% of cases.¹⁻⁵ Usually, the obstruction is either at the level of the pulmonary trunk or pulmonary bifurcation and, more uncommonly, in the right ventricular infundibulum. Conversely, right ventricular outflow obstruction resulting from late onset pulmonary valve stenosis (previously aortic valve) is exceedingly rare.⁶

We report two neonates who had the arterial switch procedure, in whom right ventricular outflow tract obstruction caused by neopulmonary valve stenosis developed during medium term follow up. As with simple pulmonary valvar stenosis, balloon valvuloplasty proved to be effective in treating this rare late onset surgical complication.

Case reports

Two neonates, 1 and 5 days old respectively, were referred because of TGA-IVS. Significant ventriculo-arterial pressure gradient or dyspla-

sia of the ventriculo-arterial valves was not found during echocardiography or cardiac catheterisation. After successful arterial switch, the neonates underwent clinical follow up during which a progressive right ventricular outflow tract obstruction caused by stenosis of the neopulmonary valve (fig 1) and, to a lesser extent, of the pulmonary trunk was diagnosed. Balloon valvuloplasty was performed at 16 and 30 months respectively, because of right ventricular hypertension (RV:LV ratio 0.6). After the procedure, the RV:LV ratio dropped to 0.3, and the transvalvar peak pressure gradient decreased to 15 and 20 mmHg, respectively. There was no significant restenosis during follow up of 84 and 10 months, respectively.

Discussion

TGA-IVS is a fairly common congenital heart disorder in which dysplasia and/or stenosis of the aortic valve, which will become the pulmonary valve after an arterial switch procedure, is quite uncommon.⁷ The arterial switch operation (Jatene procedure) in neonatal age has become the most widely performed procedure for TGA-IVS, owing to theoretical advantages over the physiological repair of Mustard or Senning and an acceptable mortality and morbidity.¹⁻³ Progressive right ventricular outflow obstruction is a common complication after surgery. The obstruction is usually located at the pulmonary trunk level, resulting from scarring of the anastomotic site or, more frequently, at the bifurcation of the pulmonary trunk owing to the traction on the pulmonary arteries after the Lecompte manoeuvre.¹⁻⁴ Conversely, late onset stenosis of the neopulmonary valve (previously aortic valve) has been seldom reported.⁶ In our patients, the preoperative examination did not show native aortic valve dysplasia or stenosis, and direct inspection during surgery showed a trileaflet aortic valve. No significant right ventricular outflow obstruction early after surgery was recorded. Thus, the cause of the late onset neopulmonary valve stenosis is not well understood, although it could be supposed that, over time, a negligible, mild valvar dysplasia evolved into frank stenosis. It could also be speculated that the increased pulmonary trunk pressure secondary



Figure 1 Right oblique view angiogramy showing the postprocedure pulmonary valve "doming" before the pulmonary balloon valvuloplasty.

to the supra-valvar stenosis may have caused turbulence and shear stress of the valve leaflets, resulting in progressive valvar stenosis. This form of right ventricular outflow tract obstruction was successfully treated by percutaneous balloon dilatation, with long lasting relief of the stenosis during medium term follow up, as previously reported for valvuloplasty performed in the first year of life.⁸ This favourable result of balloon angioplasty is at variance with previously reported experience.⁶

In conclusion, the pulmonary valve can be the site of right ventricular outflow tract obstruction after arterial switch operation and, unlike other types of right ventricular outflow obstruction secondary to arterial switch operation,⁶ it seems amenable to successful dilatation by percutaneous balloon valvuloplasty, thus avoiding further surgical procedures.

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