

Diagnosis and outcomes of patients with supra-avalvular pulmonary stenosis treated with percutaneous balloon dilation

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Introduction: Congenital supra-avalvular pulmonary stenosis (PS) is a rare subtype of PS. Outcomes following percutaneous dilation of the pulmonary artery (PA) and pulmonary valve (PV) seem to be dependent on the presence of a supra-avalvular component. The aim of this study was to determine the reliability of echocardiography in identifying the origin of the PS and to evaluate the efficacy of percutaneous dilation of supra-avalvular stenosis.

Methods: Retrospective study of 109 patients having undergone percutaneous dilation of a native PS between 2006 and 2017 in a tertiary pediatric hospital. Patients were classified as having supra-avalvular PS or non-supra-avalvular PS based on angiography. Echocardiographic measurements (main PA and PV diameters) and interventional results (hemodynamic and echocardiographic right ventricular (RV) to PA gradients) were compared between groups.

Results: Mean age at intervention was 1.93 ± 4.22 years old. Angiography identified a supra-avalvular component in 26 patients (24%). Anatomical description of the PV and PA by echocardiography had a sensitivity and a specificity of 58% and 84% respectively. Independently of the coexistence of another type of PS, patients with a supra-avalvular component had a significantly smaller PA:PV ratio measured by echocardiography: 0.84 ± 0.30 vs 1.47 ± 0.54 ($p < 0.001$). Based on the PA:PV observation, ROC analysis determined an area under the curve of 0.851 (95%CI 0.739-0.963) and a discriminative cut-off ratio of 1.048 according to Youden's index; increasing both the sensitivity and specificity to 86%. After dilation, hemodynamic gradients were reduced from 37.35 ± 20.51 mmHg to 26.04 ± 12.84 mmHg in patients with a supra-avalvular component compared to a decrease from 39.80 ± 20.47 mmHg to 11.75 ± 8.67 mmHg in patients without ($p < 0.001$). The gradient remained significantly higher among patients with a supra-avalvular PS at the 6-12 month follow-up: 48.94 ± 28.75 mmHg compared to 22.54 ± 21.28 mmHg ($p < 0.001$). Of patients with supra-avalvular stenosis, 19% required a second intervention (percutaneous or surgical) compared to 6% of those without a supra-avalvular component ($p = 0.04$).

Conclusions: Systematic calculation for the PA:PV ratio significantly improves the accurate determination of the origin of the pulmonary stenosis compared to anatomical description alone. Percutaneous dilation of a supra-avalvular PS is less effective and patients are more likely to require a second intervention than patients with pulmonary stenosis without a supra-avalvular component.