Prevalence of and performance of echocardiography in the detection of asymptomatic pulmonary vein obstruction in children after pulmonary vein repair

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Objectives: Pulmonary vein (PV) repair is burdened by a significant risk of (re-)stenosis. A proportion of patients with PV stenosis is asymptomatic. Echocardiography is routinely performed to screen for PV stenosis while cardiac magnetic resonance (CMR) is the gold standard for the detection of PV obstruction.

The aims of this study were a) to determine the prevalence of postoperative PV stenosis in a cohort of patients without clinical suspicion of obstruction, and b) to evaluate the ability of echocardiography to identify PV stenosis in these patients.

Methods: Echocardiography and CMR in asymptomatic pediatric patients after PV repair were retrospectively compared.

Results: Thirty-five patients were included: 30 had had total anomalous pulmonary venous connection (APVC), 3 had had partial APVC, and 2 had been operated for primary PV stenosis. Repair techniques included conventional (n=13) and sutureless (n=22). For 4 out of 22 sutureless patients, the indication was recurrent PV stenosis. The time between the surgery and CMR was a median of 83 months (6 days to 17 years).

CMR revealed significant stenosis in at least one pulmonary vein in a total of 11 children (31%), two following conventional repair and 9 after sutureless repair. The time between echocardiography and CMR was a median of 1.6 months (0 – 18). Echocardiography was unable to visualize all PVs in 16 patients (46%). In 8 patients (23%) the results regarding PV patency by echocardiography were incorrect, as compared to CMR. In 26% (9/35) a significant stenosis of at least one PV was missed by echocardiography. Compared to CMR echocardiography diagnosed PV stenosis correctly with a sensitivity 0.18, a specificity of 0.88, a positive predictive value of 0.40, and a negative predictive value of 0.70. The Kappa analysis between echocardiography and MRI showed poor agreement (0.11).

Conclusions: A third of this, albeit selected, cohort of patients who are asymptomatic after PV repair had significant obstruction of at least one PV. Echocardiography frequently missed obstruction in these patients.

CMR should be considered as an additional screening test in children after PV repair, irrespective of symptoms or echocardiographic findings.