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Pulmonary Arterial Hypertension in adults with Isolated Atrial Septal Defects

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Background: Natural course of atrial septal defect (ASD) is complicated by pulmonary arterial hypertension (PAH). Its optimal treatment strategy is unclear.

Aims: To identify clinical/genetic risk factors for development of PAH in patients with ASD and to define predictors of PAH resolution and functional status after defect closure.

Methods and Results: 1203 patients with isolated ASD, originating from the Dutch CONCOR registry, a nationwide registry of adult patients with congenital heart defects, were included in this retrospective study. 71 patients with PAH (6%) were identified, either by echocardiography or heart catheterization. Sinus Venosus type ASD and duration of unrepaired ASD, but not gender, were predictive for the presence of PAH.
In a subsequent case-matched study of 68 ASD-PAH patients and 184 ASD-non-PAH patients, matched for ASD-type, age at defect closure and gender, no additional risk factors for PAH could be identified. 56 ASD-PAH were tested for BMPR2-mutations, no mutations were found.
A larger diameter of the ASD, older age at ASD-repair and the presence of PAH were all associated with worse NYHA functional class.
In 43 of the 68 patients with ASD-PAH, ASD closure was performed and in 27 (63%) PAH resolved after repair. No pre-operative factors (gender, NYHA class, ASD-diameter, age at repair or hemodynamics) were predictive for post-operative PAH-resolution. However, mean NYHA class improved significantly after ASD-closure, irrespective of resolution of PAH. However, mortality was higher in patients with PAH compared to those without PAH (4/68 vs. 2/186; p=0.05)

Conclusions: Sinus Venosus type defect and later age at ASD-repair, but not gender or BMPR2-mutations were predictive for the development of PAH in ASD-patients. Early defect closure will reduce the prevalence of PAH in ASD-patients. ASD-repair in patients with associated PAH resulted in resolution of PAH in 63% of the patients and, independent from PAH-resolution, improved functional class. However long-term resolution of PAH remains a matter of concern.