An atrial septal defect in a young patient – not always a straightforward case

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Introduction: Pulmonary arterial hypertension (PAH) in atrial septal defects (ASDs) develops variably but atrioventricular (AV) canal abnormalities confer a higher and earlier risk[1]. Current guidelines recommend ASD closure when pulmonary vascular resistance (PVR) falls ≤ 5 Wood units (WU) after pulmonary vasodilator therapy, with still significant left-to-right shunt (Qp/Qs > 1.5)[2]. Individual treatment approach should be applied and these patients need assessment in a tertiary center with experience in adult congenital heart disease and PAH.

Case report: 26-year-old man with a history of cardiac murmur and severely reduced functional capacity (5.4 METs) on exercise testing was referred for further evaluation. Echocardiography and cardiac MRI demonstrated a large primum-type ASD with Qp/Qs 4.5:1 and severe left AV valve regurgitation with pulmonary hypertension. Ventricular septal defect (VSD) was mostly closed by right AV valve tissue and there was suspicion of residual restrictive inlet VSD. Right heart catheterization confirmed severe PAH (mean pulmonary artery pressure (mPAP) of 58 mmHg and PVR of 5.1 WU), precluding surgical septal defect closure. Targeted therapy with sildenafil and bosentan was initiated. Over the next 12 months, serial catheterizations showed a marked reduction in mPAP (45 mmHg) and PVR (3.3 WU), allowing reconsideration for surgery. Intraoperatively, a transitional AV canal defect was confirmed with description of small indirect Gerbode type VSD. Complete repair was successfully performed, including fenestrated primum ASD closure, VSD closure and both AV valves cleft repair. Recovery was uneventful, and follow-up echocardiography showed smaller RV with mildly reduced systolic function and residual mPAP of 38mmHg which persisted over the follow-up period of 3 years. Dual PAH therapy was continued postoperatively. Last excercise stress test showed markedly improved physical capacity (11 METs) and NTproBNP was normal.

Conclusion: Targeted pulmonary vasodilator therapy can reduce PVR and convert initially inoperable ASD (or transitional atrioventricular septal defect) into surgically correctable lesion. Optimal outcomes of "treat and repair strategies" require advanced imaging, expert interpretation and multidisciplinary management with repeated hemodynamic reassessment.

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LITERATURE

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