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Isolated Supravalvular Aortic Stenosis – Is a Coexistent Pulmonary Stenosis a Predictor of Unfavourable Outcome?

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Objective: Certain factors have been implicated as indicators for increased risk of reoperations in children with supravalvular aortic stenosis (SVAS) including younger age at first operation, diffuse form of disease, coexistent valvular aortic stenosis and pulmonary stenosis (PS). According to the literature, a relatively high percentage of SVAS patients need a surgical or interventional relief of PS. Bilateral outflow tract obstruction is also known to increase the mortality risk in Williams Beuren Syndrome (WBS) patients. We sought to evaluate whether the presence of PS influences the rate of reoperations and mortality in patients with SVAS.

Methods: We identified the patients with isolated SVAS from our surgical database. The patients with a multi-level aortic stenosis as well as the patients with concomitant procedures were excluded from this study. Follow-up was conducted between 2008 and 2010 and is 100% complete.

Results: Twenty-six patients underwent surgical correction of SVAS between 1974 and 2006. The patients were operated at the age between 6 months and 26 years (median 8.8 years). Seventeen patients (65%) were diagnosed with WBS. Six patients (17%) had a diffuse form of SVAS and 10 (39%) had a diagnosed PS. No patient had a surgical or interventional procedure for PS during follow-up. There was no statistically significant correlation between PS and WBS ($p=0.302$) or diffuse form of SVAS ($p=0.128$). Patients with PS were operated at younger age (4.4 ± 2.7 compared to 12.2 ± 7.1 years; $p=0.028$). Median follow-up time was 14.6 years. Overall mortality was 11.5%. One patient with preoperatively severely reduced LV-function died 27 days postoperatively. Two late deaths occurred 7 and 10 years after the initial operation. Reoperations were required in 4 patients (15%), 4-19 years after the original operation, due to stenosis of the aortic arch, supravalvular restenosis or poststenotic aortic dilatation. PS was found to be a risk factor for reoperation ($p=0.005$) as well as for the combined end point reoperation/death ($p=0.003$).

Conclusions: PS in patients with SVAS is a risk factor for reoperations in the aortic region and might be considered an indicator of the severity of the arterial disease and a predictor of an unfavourable outcome.