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Long-term outcome of discrete subaortic stenosis in adults: a multicenter study

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Introduction
Discrete subaortic stenosis (DSS) is often diagnosed early in life and notable for its rapid haemodynamic progression during childhood. However, little is known about the evolution of DSS in adulthood. Therefore, our goal was to evaluate the long-term outcome of DSS, both the natural course as well as after surgical treatment in a large cohort of adults.
Methods
All adult patients with a pre-existing diagnosis of congenital fibromuscular DSS seen between 1980 and 2011 were included in this retrospective multicenter cohort study. Clinical and surgical data were obtained from chart abstraction. Patients were classified into 2 (overlapping) groups: patients who had no surgery over time (natural course group) and patients who underwent at least 1 DSS operation (surgical group).
Results
A total of 427 patients (51.3% male) were included in this study: 149 patients in the natural course group, and 313 patients in the surgical group (412 operations). Additional congenital lesions were found in 48% of patients and 63% had aortic regurgitation. Median age at baseline was 19.3 (IQR 14.5-29.0) years. Follow-up duration was 6.1 (IQR 3.0-12.4) years in the natural course group and 12.9 (IQR 6.2-20.1) years in the surgical group.
Peak left ventricular outflow tract (LVOT) gradient at baseline was 32±17 mmHg in the conservative group, rising to 47±29 mmHg after follow-up ($p<0.001$). Progression of LVOT obstruction was predicted by: higher baseline LVOT gradient ($p<0.001$), greater left ventricular mass ($p=0.001$), presence of aortic regurgitation ($p=0.007$), younger age at baseline ($p=0.001$) and younger age at diagnosis ($p=0.004$).
In the surgical group, the peak LVOT gradient decreased from 76±28 mmHg pre-operatively to 15±14 mmHg post-operatively, but increased to 31±22 mmHg after follow-up ($p<0.001$). Re-operation was predicted by higher residual LVOT gradient (HR 1.04(1.03-1.06)) and smaller post-operative aorto-septal angle (HR 0.94 (0.91-0.97)).
Conclusions
DSS in adulthood progresses slowly in adulthood. In particular younger patients with a higher baseline peak LVOT gradient, aortic regurgitation and left ventricular hypertrophy are at risk for faster disease progression and should be monitored cautiously. Post-operatively the risk of re-operation is higher in patients with a high residual LVOT gradient and smaller aorto-septal angle.