Discrete subaortic stenosis in elderly women

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Discrete subaortic stenosis (DSS) is likely an acquired cardiac disorder which requires anatomic precursors and a genetic background. DSS occurs usually within the first decade, provoking rapidly progressive left ventricular outflow tract obstruction and secondary aortic regurgitation. DSS has been considered for a long time exclusively a disease of infancy and childhood and few reports and small series have described DSS in adulthood and only two cases are reported in elderly. Our case describes a discrete subaortic membranous ridge in an elderly woman with recent onset of dyspnea.

KEYWORDS
Discrete subaortic stenosis; Elderly

Case report
The patient, a Caucasian 77-year-old woman, has been admitted to our hospital because of recent onset of exertional dyspnoea and fatigue. Her previous clinical history was totally unremarkable. At the presentation she was moderately dyspnoeic with crackles at both pulmonary bases. An ejective systolic murmur (grade 4/6) was heard at the base, not modifying its intensity with Valsava manoeuvre or with squatting. Transthoracic echocardiography (TTE) showed normal left ventricle (LV) dimensions and moderate wall hypertrophy which was more prominent at interventricular septum (diastolic thickness = 18 mm) (Figure 1). An ejection fraction of 60% was calculated according to Simpson’s method. On apical five chamber view a mean LV-aortic gradient of 58 mmHg was recorded at continuous (CW)-Doppler analysis. Colour-Doppler analysis detected moderate eccentric aortic regurgitation (Figure 2).

Systolic anterior motion (SAM) of mitral leaflets was not present and pulsed-wave (PW) Doppler mapping of LVOT showed only a modest acceleration of blood flow. Because parasternal views did not permit a satisfactory evaluation of aortic valve, transoesophageal echocardiography (TEE) was performed. This exam showed a moderate degenerative process involving the three aortic cuspids with mild commissural fusion. A slightly reduced anatomic aortic valve area (=1.8 cm²) was measured (Figure 3). LVOT evaluation showed a circumferential membrane at LVOT level located very proximal to the aortic valve (Figure 4). Considering the clinical presentation and the echocardiographic findings the patient was referred to a cardiac surgeon but she refused the operation.

Discussion
Subaortic stenosis is likely an acquired cardiac disorder which requires anatomic precursors at level of LVOT and a genetic background. Subaortic stenosis may manifest with different aspects and grades of obstruction almost exclusively within the first decade of life. Tunnel subaortic stenosis, where the LVOT is hypoplastic with fibrous thickened endocardium, represents the most extreme type. The other spectrum of subaortic stenosis is the discrete variety (DSS), where LVOT is apparently normal and the stenosis is caused by fibromuscular ridge just proximal to the aortic valve. Rarely the two forms may coexist.

In children DSS is often associated with subtle morphologic and functional alterations of LVOT, such as small dimensions and steep aortoseptal angle. These abnormalities have been reported to alter shear stress stimulating cellular proliferation in LVOT. Under genetic predisposition, the altered septal shear stress may promote fibromuscular ridge growth and may be responsible for the rapid progression of subaortic stenosis. Therefore, DSS in children has been regarded as an adversely progressive disease and some investigators have proposed surgical repair irrespective of the transstenosis gradient.

On the other hand very little is known about DSS in adulthood and only a few anecdotal cases have been described in elderly. Oliver et al. published a single centre retrospective analysis on DSS in adults. According to their data, DSS was present in 6.5% of 2057 consecutive adults with congenital heart disease (mean age, 31 ± 17 years) and DSS was the only congenital abnormality in 66% of cases. At the opposite of what was described for children, in adults DSS seems to remain stable or progress slowly.

Transthoracic and transoesophageal Doppler echocardiography have improved diagnostic accuracy of DSS even for...
small membrane not causing hemodynamically significant pressure gradient. Nevertheless making a differential diagnosis of DSS versus obstructive subaortic hypertrophic cardiomyopathy (HOCM) can be particularly complicated. In fact patients with DSS may develop severe LV wall hypertrophy prevalently located at septal level which is sometimes associated with SAM of mitral apparatus. In the literature, it has been reported that some adult patients who received the diagnosis of HOCM were instead affected by DSS which was overlooked at first echocardiographic evaluation. In our clinical case, although a predominant hypertrophy of interventricular septum was detected, the diagnosis of HOCM was unlikely because of the non-dynamic nature of LV-aortic gradient and the negative familial history for hypertrophic cardiomyopathy.

In conclusion, we reported a rare case of DSS in an elderly woman who started to complain of symptoms of LV dysfunction. Our suggestion for early cardiac surgery was mainly dictated by the severity of LVOT obstruction and patient’s clinical features. Indeed, according to the literature a watchful approach to DSS in asymptomatic adults and elderly without severe LVOT obstruction seems to be plausible and wise.

References