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Case report

# Fixed subaortic stenosis: Case report and literature review☆

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ABSTRACT

The authors present a case of a 40-year-old female patient with fixed subaortic stenosis (SAS). This defect is classified into a group of diseases characterized by the presence of obstruction in the left ventricular outflow tract, which collectively includes stenotic lesions located in the region extending from the anatomical outflow tract of the left ventricle distally to the descending aorta. Obstructions are named according to their relationship to the aortic valve (subvalvular, valvular, and supravalvular) and their common denominator is an increase in the left ventricular afterload. Fixed subaortic stenosis may be either a focal stenotic lesion formed by a fibrous membrane, or a narrow muscular or fibromuscular tunnel diffusely tapering the outflow tract of the left ventricle. Individuals with SAS usually present with other congenital heart diseases such as a bicuspid aortic valve or perimembranous ventricular septal defect. Surgical resection of SAS is already indicated in asymptomatic individuals with severe SAS (peak gradient >50 mmHg). It is also advisable to consider surgery in some cases of less severe obstruction such as in those with coexistent moderate aortic regurgitation, ventricular septal defect, or in women planning pregnancy. In many cases, however, recurrence of SAS requires secondary surgical treatment. In the focal type of SAS, it is possible to indicate percutaneous balloon dilatation in some cases.



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## Case report

The reported patient was referred to our department in 2009, when she was 37 years old, for investigation of a systolic murmur over the aortic valve. Her past medical history was unremarkable except for two uncomplicated vaginal deliveries at age 28 and 32 years, respectively, after the second of which she received implantation of an intrauterine device IUD. Transthoracic echocardiography revealed significant subaortic membranous stenosis with maximum and mean gradients of 100/40 mmHg. The aortic valve showed no signs of morphological and functional impairment. The left ventricle was normal sized and not hypertrophic, with normal global systolic function assessed by ejection fraction of 70%. In the absence of symptoms, the patient agreed to regular clinical and echocardiographic monitoring in lieu of surgical treatment. In the following 6 years of follow-up, she remained asymptomatic, while echocardiography demonstrated only very slow progression of the subaortic stenosis with gradients 127/87 mmHg (Fig. 1). In autumn of 2015, mild concentric left ventricular hypertrophy was evident during echocardiographic examination, accompanied by the development of grade 1 diastolic dysfunction. Despite the lack of symptoms, surgical treatment was strongly recommended to the patient due to the progression of left ventricular findings, and this solution was accepted. Preoperative transesophageal echocardiographic examination was performed in order to further characterize the lesion. This examination confirmed the presence of a circular subaortic fibrous membrane extending to the anterior leaflet of the mitral valve and obstructing the left ventricular outflow tract to 1 $cm^2$ as assessed by 3D planimetry (Figs. 2 and 3). Cardiac surgery was performed through a transverse aortotomy above the coronary orifices. First, the presence of subaortic stenosis was verified; it was formed by a rigid fibrous membrane that almost circularly extended to the anterior leaflet of the mitral valve (Fig. 4). Subsequently, subaortic membrane resection was performed including ablation of membranous material from the anterior leaflet of the mitral valve. The postoperative course was uncomplicated and the patient was subsequently discharged in good condition.

## Discussion

Fixed subaortic stenosis (SAS) is a relatively rare heart disease whose prevalence in the population of adults with congenital heart disease is estimated at around 6.5%, with a male to female ratio of 2:1 [1]. It is a unit comprising discrete focal stenotic lesions formed by a fibrous membrane as well as diffuse types of obstruction of the left ventricular outflow tract formed by a narrow muscular or fibromuscular tunnel. An exceptional cause of SAS is an abnormal accessory mitral valve chordae anchored at the base of the interventricular septum.

While the discrete membranous form of SAS represents the vast majority of obstructions (90%), muscular tunnel lesions are usually associated with a more severe degree of stenosis [2]. Since it is rarely present in newborns or in early childhood, SAS is considered by many authors to be rather an acquired heart defect than a congenital one [3]. The pathogenesis of SAS has not been clearly elucidated yet. An intriguing hypothesis has been published by Rosenquist et al., who found a consistently increased distance between the mitral and aortic valves in patients with SAS, which may result in a change of the blood flow direction near the distal interventricular septum and thus form a basis for the differentiation of germinal cells into fibroblasts [4]. This hypothesis is also supported by the fact that the development of SAS may occur after some surgical operations in the left ventricular outflow tract, where secondary proliferation of connective tissue due to the turbulent blood flow in a given area is considered to be the cause of obstruction [5].

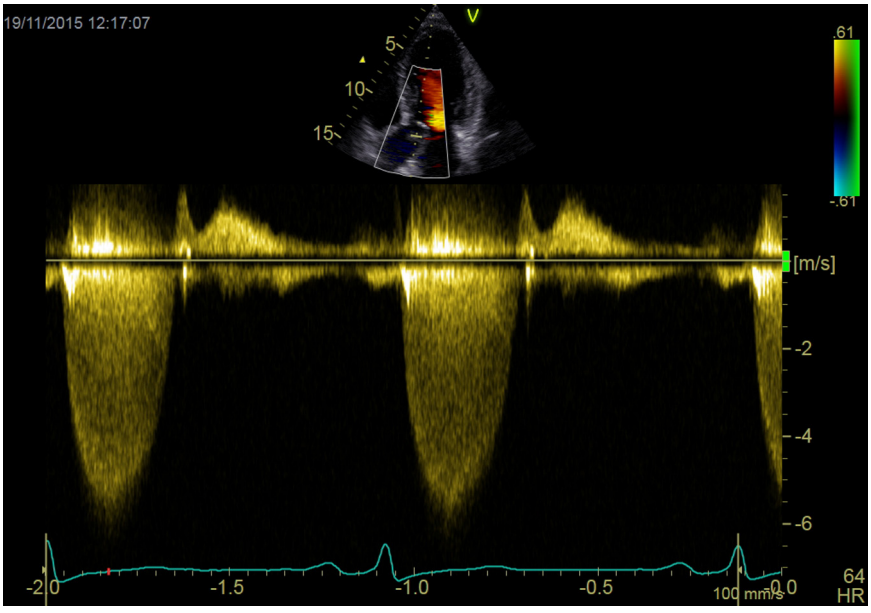


**Fig. 1 – Transthoracic echocardiography. Continuous-wave Doppler recording showing hemodynamically significant fixed subaortic stenosis with peak and mean pressure gradients of 127 mmHg and 87 mmHg, respectively.**

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