Fixed subaortic stenosis

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Abstract

Fixed subaortic stenosis (FSS) is an obstruction of the left ventricular outflow tract below the aortic valve. The discrete fixed membranous subaortic stenosis (DMSS), the discrete fibromuscular subaortic stenosis (DFSS) and the tunnel subaortic stenosis (TSS), which both are frequently more severe than the membranous form, represent the anatomic spectrum of the cardiac lesion. Eight percent to 20% of patients with left ventricular outflow obstruction are affected by FSS. Subaortic stenosis occurs in association with other cardiac lesions and aortic arch obstruction due to interruption of the arch or coarctation; these lesions include ventricular septal defect and malalignment of the outlet septum with displacement into the left ventricular outflow tract, double outlet right ventricle (Taussig-Bing anomaly), and atrioventricular septal defect with abnormal mitral valve attachment. Subvalvular stenosis is one of the lesions of the Shone complex (coarctation of the aorta, parachute mitral valve, supravalvular stenosing ring above the mitral valve); in more than 20% of patients, the aortic valve is abnormal (valvular stenosis, small aortic annulus, bicuspid valve); the subvalvular membrane is occasionally adherent to one of the valve cusps of the aortic and mitral valve and this may interfere with valve function, resulting in mild degrees of insufficiency. Morphologic abnormalities within the left ventricular outflow tract result in altered shear stress, which reveals a genetic predisposition leading to proliferation of fibrous cells in the outflow tract (high recurrence rate). The best method for diagnosing subaortic stenosis is transoesophageal echocardiography showing the left-ventricular outflow tract in 110° cranio-caudal long axis projection. Current operative methods are considered generally as safe and effective, although doubts have been raised for the long-term results in cases associated with aortic valve abnormalities. Resection of the fibromuscular ridge, Konno, modified Konno and Ross-Konno operations are techniques used worldwide.

Keywords
Cardiac lesion, left ventricular outflow obstruction, transoesophageal echocardiograph, konno opération
Disease name and synonyms

- Fixed subaortic stenosis (FSS)
- Discrete fixed membranous subaortic stenosis (DMSS)
- Discrete fibromuscular subaortic stenosis (DTSS)
- Tunnel subaortic stenosis (TSS)

European Paediatric Cardiac Code
Reference of "Fixed subaortic stenosis" is 07.09.03.

Excluded diseases
Hypertrophic obstructive cardiomyopathy (HCM)

Diagnosis criteria / definition
A crescent or a complete ring (which is less common) of fibroelastic tissue lying 0.5 – 1.0 cm beneath the aortic valve is found within the left ventricular outflow tract. It is sometimes adherent to the aortic cusps or to the anterior leaflet causing aortic insufficiency; the tunnel type is associated with fibromuscular hypertrophy of varying length, sometimes extending to the anterior leaflet of the mitral valve resulting in mitral insufficiency (8, 9).

Differential diagnosis
- Hypertrophic obstructive cardiomyopathy (HCM)
- Left ventricular outflow tract obstruction by an aberrant papillary muscle
- Left ventricular outflow tract obstruction by outgrowth of aberrant mitral cusp tissue
- Herniation of tricuspid valve tissue through a ventricular septal defect
- “Goose-neck” deformity of the left ventricular outflow tract in atrioventricular septal defect

Frequency
Eight percent to 20% of patients with left ventricular outflow obstruction are affected by FSS.

Clinical description
Patients present with carotid thrill, powerful left ventricular apex and a long aortic ejection systolic murmur at the left sternal border; no systolic ejection click like in valvular aortic stenosis is found; associated systolic or diastolic murmurs are found in case of aortic- or mitral insufficiency. Pulses with a sharp upstroke depending on the presence of a dynamic muscular component of the obstruction are also found. On the electrocardiogram varying degrees of left ventricular hypertrophy without Q-waves in leads V5/V6 are observed; the presence of ST depression and T inversion in leads V5/V6 suggests subaortic obstruction. Left anterior hemiblock is found in older patients and reveals severe myocardial disorder associated with excessive dysplastic muscle in younger patients; right bundle branch block occurs in about 3% of the patients.

Generally, chest X-ray is not particularly relevant; sometimes poststenotic dilatation of the ascending aorta is visible, slight enlargement of the left atrium may be apparent in older patients. The anatomic features can be shown by transthoracal (parasternal long-axis view, apical four-chamber view) and more detailed by transoesophageal echocardiography (110° crano-caudal); colour flow- and cw-doppler are helpful in the assessment of the severity of the obstruction; flutter of the aortic cusps in m-mode imaging is characteristic for subaortic stenosis.

Management
Conservative management is recommended: no sports even at mild degrees of left ventricular outflow tract obstruction. Prophylaxis for endocarditis is mandatory (16).

Interventional therapy consists of balloon dilatation in the membranous form of subaortic stenosis (17).

Surgical therapy can take several forms: resection of the fibromuscular ridge (13), Konno (14), modified Konno (15) and Ross Konno operations (13).

Etiology
The rarity of fixed subaortic stenosis in newborns and its absence in the developing heart, its progressive and probably acquired nature have been the subject of several studies (18). Shear stress has been shown to trigger significant cellular proliferation; even minimal stenosis of the left ventricular outflow tract can result in altered shear stress, which reveals a genetic predisposition, leading to cell proliferation and deposition of fibrous material in the left ventricular outflow tract.

Diagnostic methods
Clinical findings help establishing the diagnosis. Echocardiography, especially transoesophageal echocardiography (110°-120°) needs to be carried out.

In case of associated anomalies, cardiac catheterization is mandatory.

Antenatal diagnosis
The obstruction is not usually manifest in the fetus and develops after birth.
References

17. Ascuito RJ, Ross-Ascuito NT, Pickoff AS, Fox LS. Percutaneous balloon dilatation of discrete subaortic stenosis as a palliative procedure to promote recovery of left ventricular contractile function. Pediatr Cardiol 1993;14:122-123