Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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CASE SUMMARY

The Authors presented a case of 61-year old male patient with hypertrophic cardiomyopathy with left ventricular outflow tract obstruction. In echocardiography following features were shown: concentric hypertrophy of left ventricle with maximal diastolic thickness of septum 21 mm, pressure gradient in LVOT 98 mmHg (max. 114 mmHg), without SAM, elongated leaflets of the mitral valve, moderate aortic insufficiency, aorta not widened, free pericardium. The main problem is the further patient management.

DISCUSSION

Initial approach to a patient with hypertrophic cardiomyopathy (HCM) involves the assessment of several clinical issues: risk stratification for sudden death, management of heart failure symptoms, screening/genetic testing and physical activity restrictions. In patients with limiting heart failure symptoms, the presence of LV outflow tract obstruction must be determined so that optimal management strategies can be formulated. For patients without flow obstruction at rest (or with provocation) and limiting heart failure symptoms, medical therapy should be initiated. Beta blockers are generally preferred and are dose-titrated to symptom benefit. Alternatively, calcium channel blockers can be tried, and, in some patients, disopyramide extended release can be added to a beta blocker, although this option is often limited by adverse effects. It is only recommended invasive septal reduction therapy (myectomy or alcohol septal ablation) in patients with obstruction with drug-refractory advanced heart failure
symptoms. If a patient has no evidence of outflow obstruction at rest or with provocation and heart failure symptoms, AV nodal-blocking agents can be offered to improve diastolic dysfunction. If patients continue to experience progressive heart

failure despite these drugs, then heart transplantation is the only long-term treatment option. If a patient is asymptomatic, there is no compelling role for drug therapy. (1)

Up to one-third of patients with HCM have mild AR, probably caused by sub-aortic obstruction and high-velocity flow in the LV outflow tract. Moderate-to-severe AR is much less common and is usually caused by primary disease of the aortic valve leaflets or aortic root and infective endocarditis when present in a patient with LVOTO, a non-SAM-related mechanism for obstruction, such as a sub-aortic membrane, should be excluded. Aortic regurgitation may also occur following septal myectomy, particularly in children and young adults. (2) The severity of AR should be assessed in accordance with ESC guidelines by evaluating the anatomy of the valve, the size of the aortic root and of the ascending aorta, and other qualitative, semi-quantitative and quantitative parameters. The size of the LV cavity is an unreliable marker of the severity of AR in HCM. (3)

EXPERT'S OPINION

] By consensus, patients with symptomatic LVOTO are treated initially with non-vasodilating ß-blockers titrated to maximum tolerated dose. If ß-blockers alone are ineffective, disopyramide (when available), titrated up to a maximum tolerated dose (usually 400–600 mg/day), may be added. Verapamil (starting dose 40 mg three times daily to maximum 480 mg daily) can be used when ß-blockers are contraindicated or ineffective.

Invasive treatment to reduce LVOTO should be considered in patients with an LVOTO gradient ≥50 mm Hg, moderate-to-severe symptoms (New York Heart Association (NYHA) functional Class III–IV) and/or recurrent exertional syncope in spite of maximally tolerated drug therapy. (3)

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