EDITORIAL

Comprehensive Treatment of Obstructive Hypertrophic Cardiomyopathy

Tratamiento integral en la miocardiopatía hipertrófica obstructiva

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Obstructive hypertrophic cardiomyopathy is a cardiac condition characterized by ventricular hypertrophy and a progressive worsening of symptoms associated to increased subaortic gradient. Pharmacological treatment with negative inotropic agents usually relieves symptomatology and functional capacity. When symptoms do not subside, which occurs in 5% of the patients, an invasive behavior must be established, either with surgical septal myectomy, initiated by Morrov in 1968, (1) or alcohol septal ablation. This last method was introduced by Sigwart in 1995 in three patients to provoke hypokinesia (controlled infarction) in the area with consequent gradient reduction. (2, 3)

Obstructive hypertrophic cardiomyopathy occurs in one out of 500 people. It presents with morphological, functional and clinical heterogeneity, and represents 10% to 20% of all forms of left ventricular outflow tract obstruction. (4) Several theories have been raised concerning its origin: the genetic theory takes into account a special predisposition for cellular proliferation, with gene mutations of the contractile apparatus (myosin heavy chain, troponin T and alpha-tropomyosin), resulting in sarcomere damage. It may run in the family, but usually occurs at puberty. The mechanical theory suggests the generation of turbulence (mechanical stress), with endothelial damage and development of hypertrophy. This theory is based on the fact that the disease may appear in patients without previous obstruction, its presence is not usual in the first years of life and recurrence is possible after septal myectomy. It has also been suggested that obstructive hypertrophic cardiomyopathy may develop as a result of poor alignment of the interventricular septum or to a small outflow tract. In the geometric theory, it is argued that an increase in the mitro-aortic distance produces changes in blood flow dynamics during fetal life leading to septal hypertrophy. (5) In conclusion, genetic predisposition and flow turbulence factors are evidently associated to the etiopathogenesis of this disease, leading to the proliferation of cardiac muscle tissue.

The evolution in the majority of patients with moderate symptoms is usually slow. This is the result of asymmetric hypertrophy, decreased distance between the left ventricular posterior wall and the septum and the systolic anterior motion of the mitral valve, producing left ventricular outflow tract gradient.

The therapeutic choice must include the analysis of other structural anomalies that may accompany the condition. Although both septal myectomy and alcohol ablation are considered in some studies with similar indications, (6) we must analyze this point more thoroughly. Precisely, the main benefit of extended septal myectomy is to treat the mitral and septal components concomitantly, which represents an advantage over percutaneous alcohol treatment. Young patients who usually suffer from obstructive hypertrophic cardiomyopathy along with significant septal hypertrophy also tend to respond better to surgical treatment. In addition, we must consider that patients with accelerated progression of the disease improve after septal myectomy. Both in this group and in young patients presenting obstruction, the prognosis worsens with the progression of the cardiomyopathy.

Patients with valvular involvement due to regurgitation or systolic anterior motion should have surgical indication, as well as those who are young and generally have greater septal hypertrophy. The results achieved in these patients have been optimal.

Valve involvement is added to obstructive hypertrophic cardiomyopathy as a result of mitral regurgitation and anterior systolic motion. The strategy of achieving mitral valve correction has excellent results in the series analyzed. (7)

The surgery to treat this condition has evolved with the knowledge of its pathophysiology and the results obtained. Therefore, simple subaortic ventricular myotomy, whose outcome showed persistent high gradients, had to be replaced. Towards 1968, Morrow introduced septal resection (myectomy). This procedure allowed adequate results by decreasing subaortic gradients and flow velocity and eliminating the attraction force of the anterior mitral valve leaflet due to a Venturi effect, which caused anterior systolic motion. However, nowadays the Venturi effect has been resolved, and septal hypertrophy is the cause not only of the gradient, but also of anterior systolic motion.

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Therefore, the technique currently supported, as proposed by the work of Vrancic et al. (8) involves not only extended septal myectomy, but its association with concomitant mitral surgery. This possibility is a clear advantage over alcohol ablation in this type of patients with subaortic hypertrophy who have complementary and associated pathologies.

Regarding the strict surgical technique, septal resection should be analytically performed. If it is small, it will not succeed in lowering the gradient, and in case of exceeding the resected tissue, there is risk of complications (ventricular septal defect, cardiac blocks). Some authors propose to perform myectomy with the help of a cardioscope (video-assisted myectomy), to allow resection at the base of papillary muscle implantation, thus avoiding the risk of damaging the mitral subvalvular area. (7) A technical subtlety is represented in Figure 1 where the use of the broken scalpel handle allows acting without obstructing the view.

Mitral pathology includes an increase in leaflet size with anterior position of the papillary muscles; therefore, leaflet plication becomes important to prevent an isolated myectomy from leaving mitral regurgitation. This technique should be performed especially in patients with anterior systolic motion, mitral regurgitation and increased leaflet size. Valve plication can be performed longitudinally, but others also perform it transversely. (9)

Gradient analysis in the left ventricular outflow tract, mitral regurgitation and systolic anterior motion should be considered both in the therapeutic choice: septal myectomy or alcohol ablation, as in the evaluation of long-term results.

The work presented by Vrancic et al. (8) shows a very good experience both in the surgical decisions adopted as in the results obtained. The gradients achieved are within international standards. The small series makes the percentage of complete atrioventricular block (14.2%) seem relevant. In this aspect, care must be taken with the 5 mm of septum situated below the aortic ring and the commissure between the non-coronary and right coronary cusps. Finally, we agree with the authors regarding the concept that in centers of experience the disease can be fully treated with low morbidity and mortality. (10)

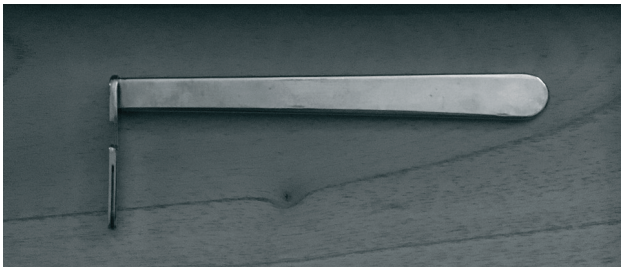

Fig. 1. Broken scalpel. Courtesy of Dr. Jorge Trainini (Buenos Aires, Argentina).

CONFLICTS OF INTEREST

None declared.

(See authors' conflicts of interest forms on the website/Supplementary material).

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