

Intraoperative Evaluation and Surgical Planning in the Hypertrophic Cardiomyopathy

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A male, 45 year old patient, with diagnosis of septum hypertrophic cardiomyopathy, having undergone a cardiofibrilator implant in the past. Despite the optimized clinical treatment, he evolved to a progressive clinical deterioration that led to invasive treatment. However, there was not an important gradient in the exit pathway of the left ventricle (EPLV) at the echocardiogram in rest and the challenging test with dobutamine was non conclusive because it did not reach the preconized cardiac frequency. The intraoperative evaluation with a transesophageal echocardiogram using isoproterenol was fundamental for the diagnosis of the EPLV obstruction. The septum myectomy was performed successfully and the patient presented good post-operative evolution.

Introduction

Result of a disarrangement of the myocardial fibers, the hypertrophic cardiomyopathy (HCM) has several degrees of clinical onset, depending on the anatomical and genetic characteristics.

The majority of the patients responds adequately to the clinical treatment, specially those without obstruction of the left ventricle exit pathway (EPLV). The presence of refractory symptoms to the clinical treatment, in the presence of major gradient (≥ 50 mmHg) in rest or with challenging test, identifies the group that needs the surgical treatment or alcoholic septum ablation^{1,2}.

The objective of this paper is to report the case of a patient with HCM. Refractory to the clinical treatment and without major obstruction of EPLV at the echocardiogram in rest and during the stress, However, when submitted to intraoperative evaluation, little conventional to measure the EPLV gradient, the use of the challenging test with isoproterenol proved that it is useful, as well as the measurement of the intraoperative transesophageal echocardiogram parameters for the surgical planning.

Keywords

Cardiomyopathy, hypertrophic; intraoperative care; thoracic surgery; advance care planning.

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Case report

Male, 45 year old patient, followed up in other service due to symmetric septum HCM, went to the institution with worsening of the cardiac insufficiency symptoms for functional class III of the New York Heart Association (NYHA) in the last six months, despite the optimization of the clinical treatment (atenolol 100 mg/day + verapamil 480 mg/day + amiodarone 200 mg/day), associated to successive events of pre-syncope at the minimum efforts. The diagnosis had been established 10 years before, due to cardiac insufficiency (CCI) and syncope at efforts. The echocardiogram of that time revealed symmetric septum hypertrophy (septum thickness of 20 millimeters and of the posterior wall of 9 millimeters), however, without EPLV gradient. The clinical treatment with betablockers and calcium channel blocker was effective for three years. He was, then, submitted to the cardiodefibrilator implant, as a primary prevention for sudden death due to family history and recurrent syncope.

The events of syncope, however, persisted, despite the non correlation with any cardiac arrhythmia at the implantable cardiodefibrilator.

When coming to our institution, he presented major functional limitation and significant worsening in the quality of life, in functional class III of NYHA and with successive events of pre-syncope triggered by the minimum efforts. The echocardiogram showed increase in the thickness of the septum in relation to the previous exam (23 millimeters), gradient in EPLV in rest of 8 mmHg, ejection fraction of 64% with diastolic dysfunction degree I and competent mitral valva.

The stress echocardiogram with dobutamine was considered non conclusive because it reached only 69% of the preconized heart rate. The maximum gradient in EPLV with 40 microgram/kg/minute of dobutamine was of 46mmHg. At that moment, he presented intensive dyspnea and classical signs of low cerebral debit.

The surgical indication was justified by the exuberance of the symptoms and increased septum thickness, even in the absence of important gradient documentation in EPLV. Soon after the anesthetic induction, it was performed the transesophageal echocardiogram with isoproterenol infusion at 20 micrograms/kg/min, keeping the average blood pressure over 90 mmHg. There was increment of the heart rate that went from 80 to 120 beats per minute. Due to these circumstances, it was possible to have the gradient documentation in EPLV of 100 mmHg and appearance of major mitral insufficiency by previous systolic movement. Measures were determined in order to plan the septum resection (fig. 1A): place with interventricular septum

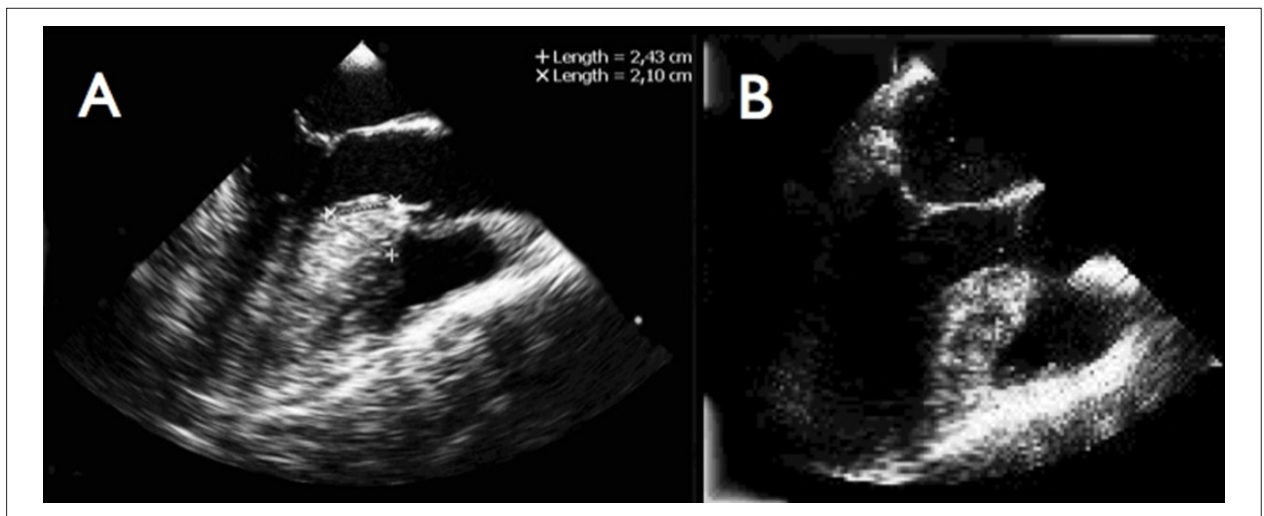


Figure 1 - Image of EPLV by transesophageal echocardiogram at the medium esophagus level with 120° angle of the transducer. Measurements of the septum thickness and distance of the greatest diameter to the aortic ring before (A) and after (B) the myectomy.

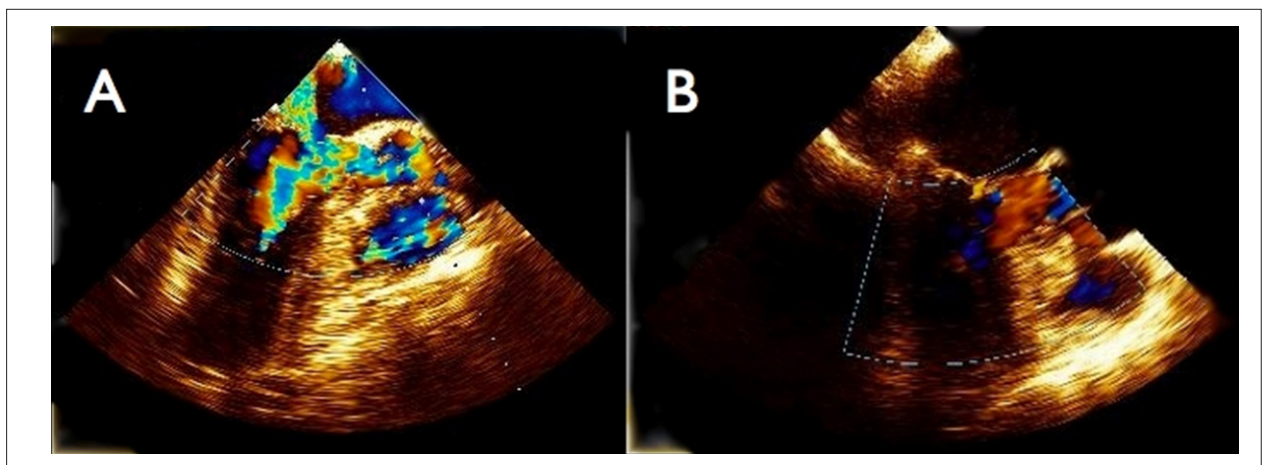


Figure 2 - Image of EPLV with color Doppler by transesophageal echocardiogram at the medium esophagus level with 120° angle and challenging test with isoproterenol. Presence of turbulent flow (high gradient) in EPLV and significant mitral insufficiency before the myectomy (A), with disappearance of the turbulent flow in EPLV and absence of mitral insufficiency after the resection (B).

thickness and its relation to the aortic ring, in addition to the septum thickness distance in direction to the left ventricle end.

Expanded septum myectomy was performed with broad resection in wedge, between the fibrous trigonous of the heart, respecting the previously established measurements by the echocardiogram in relation to the depth of the septum resection, distance from the aortic ring and extension inwards of the ventricular cavity. After the extracorporeal circulation exit, it was performed a new echocardiographic (fig. 1B), which showed reduction of the interventricular septum thickness to 16 millimeters. Without gradient in rest and with gradient of 21 mmHg after the isoproterenol infusion, without mitral insufficiency (Fig. 2). The post-surgery recovery took place without problems receiving hospital discharge in 10 days.

In the 18 month follow-up, he was in the functional class I of NYHA without syncope or arrhythmia, currently

using the angiotensin II inhibitor, calcium channel blocker and amiodarone.

Discussion

The present case illustrates the following peculiarity: patient with diagnosis of symmetric septum HCM, exuberant clinical condition of ICC and repeat syncope, refractory to clinical treatment, but without evident demonstration of EPLV obstruction.

The HCM can present quite variable clinical course. However, some patients can present symptoms of ICC, embolic events and sudden death, other stay stable or symptomatic over the years, with more than 25% of the affected people presenting longevity equal to the one of the normal population. It is possible that its course changes according to the genetic substrate¹⁻³.

Case Report

Despite initially described as a predominantly non obstructive disease, among the patients that look for specialized treatment, the obstructive form was demonstrated as the most prevalent, in which symptoms of the ICC are justified by obstruction to the flow induced by effort⁴. In patients very symptomatic, as is this case, even in the absence of the significant gradient in rest, it is mandatory to research the dynamic obstruction that justifies such symptoms, Aggressive maneuvers, such as Valsalva, physical exercise or dobutamine infusion can be used. In selected cases, if doubts persist, the usefulness of isoproterenol in elucidating latent gradients in patients with HCM was described⁴⁻⁶. In a recent series, 14 of the 25 patients with HCM and symptoms of ICC functional class III/IV not initially considered for invasive treatment, due to the absence of significant gradient, presented gradient ≥ 50 mmHg during cardiac catheterization after the use of isoproterenol. The average increase of the gradient with the drug infusion was of 50 ± 41 mmHg in relation to the basal value. When submitted to septum myectomy or septum ablation by alcohol, 93% of these holders of inducible gradient experienced important relieve of the ICC symptoms⁶. This resource was used in this case, as despite the exuberant symptoms, the gradient of EPLV presented in the limit (46 mmHg) after the stress echocardiogram with dobutamin. The isoproterenol acts at the beta-adrenergic receptor levels causing increase of the heart rate, positive inotropism and vasodilation. Increase in the flow speed caused by the drug, in the presence of septum hypertrophy, can lead to anterior displacement of the mitral valvar apparatus due to the Venturi effect. This phenomenon is responsible not only for the subaortic obstruction, but also for the concurrent appearance of mitral insufficiency, due to the incomplete apposition of the cusps^{2,4}. The challenging test with isoproterenol did not present complication in greater casuistries⁶.

Septum myectomy was indicated as it is considered reference standard in patients with symptoms refractory to medication therapy and obstructive form of HCM, being

this suspicion confirmed after a test with isoproterenol. The surgical result was excellent, because it abolished the gradient in EPLV during the rest and kept the interventricular septum in 16 millimeters, corroborating the complete septum resection. The follow-up of 16 months shows the symptomatic patient, with expressive improvement of quality of life. This finding is based on the broad experience of the technique in reference centers^{7,8}.

In this disease, the addition of the surgical planning by the intraoperative echocardiogram played a crucial role^{9,10}. In addition to evaluate the EPLV gradient after the challenging test and the mechanism of the mitral insufficiency, helped in the estimate of the degree and of the extension of the septum resection. It was possible also to evaluate the relief degree of the obstruction to the EPLV flow after the intervention and analyze the eventual need of additional resection, because the recurrence of symptoms in the post-surgery period usually is a reflex of the incomplete resection.

HCM is a disease with complex physiopathology and is not totally elucidated. We illustrate a case with exuberant symptoms of ICC, where the challenging maneuver with isoproterenol could identify the patient with dynamic obstruction, who benefited from the surgical treatment of septum reduction.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

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