Intraoperative Transesophageal Echocardiography in Septal Hypertrophic Cardiomyopathy

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We report the case of a patient with septal hypertrophic cardiomyopathy undergoing surgical correction in which the use of intraoperative transesophageal echocardiography permitted the planning of the surgical approach and an immediate knowledge of the surgical outcome.

Introduction

Surgical treatment of hypertrophic cardiomyopathy (HCM) is indicated for symptomatic patients who are refractory to optimized drug therapy and who present a left ventricular outflow tract (LVOT) gradient higher than 50 mmHg.

The use of Intraoperative Transesophageal Echocardiography (IOTEE) has been proposed to guide surgeons in septal myectomy planning, in real-time anatomical or functional aspects, and in *in situ* detection of residual defects or complications subject to correction before termination of extracorporeal circulation[1].

For these reasons, IOTEE is a Class I indication in the Guidelines of the Brazilian Society of Cardiology (BSC) and in the American consensus of several entities[2,3].

Case report

A 45-year-old female Caucasian patient presented with history of progressive dyspnea for approximately four years, when she was diagnosed with hypertrophic cardiomyopathy (HCM). At that time, she underwent transthoracic echocardiography (TTE) which showed left ventricular diastolic diameter (LVDD) of 46 mm; left ventricular systolic diameter (LVSD) of 23 mm; left ventricular ejection fraction (LVEF) of 81%; interventricular septal (IVS) thickness of 18 mm; and LV posterior wall (PW) thickness of 9.5 mm. Study of the intracardiac flows showed obstruction of the left ventricular outflow tract (LVOT), with peak gradient of 33 mmHg and mean gradient of 15 mmHg. She was started on metoprolol tartrate and furosemide with escalating doses, reaching a mean heart rate of 56 bpm.

After three years of clinical follow-up and observation of worsening of symptoms, she underwent new TTE which showed worsened echocardiographic parameters, with IVS of 22mm; PW of 15 mm; LVOT peak gradient of 130 mmHg; LVOT mean gradient of 71 mmHg, with moderate mitral regurgitation and anterior systolic motion (ASM) of the anterior cusp of the mitral valve; LVDD of 40 mm; LVSD of 25 mm; and LVEF of 68%. These findings were confirmed by magnetic resonance imaging (MRI) (Figure 1).

Surgical treatment for HCM was chosen. Surgery was performed with the diagnostic support of IOTEE (Figure 2A). In the intraoperative transesophageal study prior to surgical correction, counterclockwise rotation of the papillary muscles (anatomical distortion caused by cardiomyocyte hypertrophy) was observed. Septal myectomy was performed via the transaortic approach with incision on the IVS, with removal of some fragments. IOTEE study after termination of extracorporeal circulation and optimization of blood volume showed IVS: 15 mm; PW: 15 mm; peak intraventricular systolic gradient: 38 mmHg; LVEF: 68%; and improved counterclockwise rotation of the papillary muscles. The pathological study of the fragments removed showed endocardial fibrous thickening with hypertrophy and cardiomyocyte disarrangement.

The patient had a good postoperative outcome and was asymptomatic in the clinical follow-up after surgery.

Discussion

HCM is a condition of unknown cause, anatomically and histologically characterized by myofibrillar disarrangement and hypertrophy, and functionally by difficult ventricular filling. Usually, ventricular systolic function is preserved, whereas diastolic function is impaired. Its prevalence is of approximately 0.2% in the general population, and it is more frequently identified in adults between 30 and 40 years of age.

Among the tests for diagnostic investigation, electrocardiography may show ST-segment and T wave alterations, LV overload, and prominent T waves in inferior, septal and anterior walls, depending on the wall predominantly affected by hypertrophy.

Currently, echocardiography is the most frequently used technique for the diagnostic investigation of HCM. It has been used both for the investigation of suspected cases and for the screening of relatives of HCM patients. Echocardiography permits the assessment of morphologic, functional and hemodynamic aspects of HCM. The most characteristic

Key Words

Cardiomyopathy, hypertrophic; echocardiography, transesophageal; cardiac surgical procedures.

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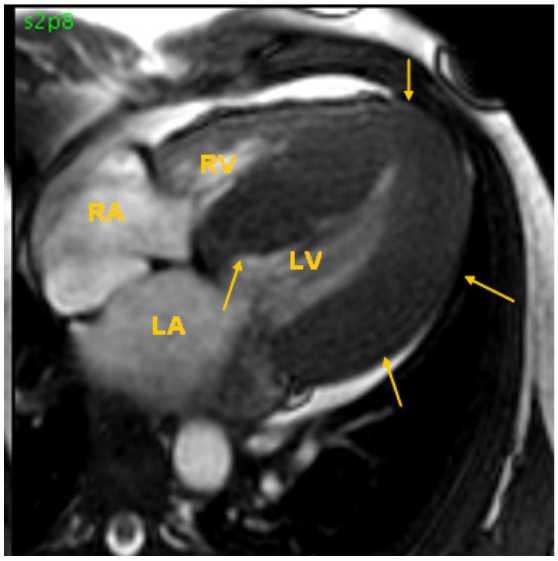

Figure 1 - MRI (long axis) of a patient with HCM; Arrows show significant LV hypertrophy; RA – right atrium; RV – right ventricle; LA – left atrium; LV – left ventricle.

echocardiographic finding in HCM is the presence of LV hypertrophy, although with a large variability in its degree and presentation. Most of the patients (90%) present the asymmetrical form with predominance in the IVS. In the form in which the septum is more severely affected, the IVS is usually ≥ 15 mm, with a > 1.3 IVS/PW ratio. Approximately 25% of the patients present dynamic LVOT obstruction. This occurs due to the hypertrophic IVS juxtaposion to the LVOT and also to the apposition of the anterior cusp of the mitral valve during cardiac systole. The mitral valve cusps are abnormally elongated, thus affecting the LVOT geometry, and leading to an intraventricular pressure gradient. In this situation, abnormal ASM of the anterior cusp is observed. HCM frequently leads to mitral regurgitation (40% to 75%), both in patients with the obstructive form and in those without LVOT obstruction. The LV cavity may be normal or proportionally smaller, measuring < 45 mm in most of the patients.

MRI and computed tomography are useful diagnostic methods when echocardiography fails to identify anatomic changes[4].

The clinical course of HCM is variable, with around 3% annual mortality rate among adults. The risk of sudden death is higher among children, approximately 6% per year. Clinical deterioration is usually slow and gradual, and progression to dilated forms is seen in 10% to 15% of the patients.

Clinical management is targeted at the relief and prevention of symptoms and their recurrence, as well as at the reduction of the risk of sudden death, thus improving the quality of life and, consequently, increasing life expectancy[5].

The great majority of patients require clinical treatment with betablockers or verapamil, and diuretics in advanced cases of heart failure. Invasive interventions are necessary in approximately 5% to 10% of the symptomatic patients who are refractory to medical treatment and persist with a high LVOT gradient[1].

Biventricular pacemaker implantation may be useful in patients with LVOT gradient and severe symptoms, providing reductions of up to 25% in the gradient values.

The objective of surgical treatment is to reduce the LVOT gradient, and is indicated for symptomatic patients

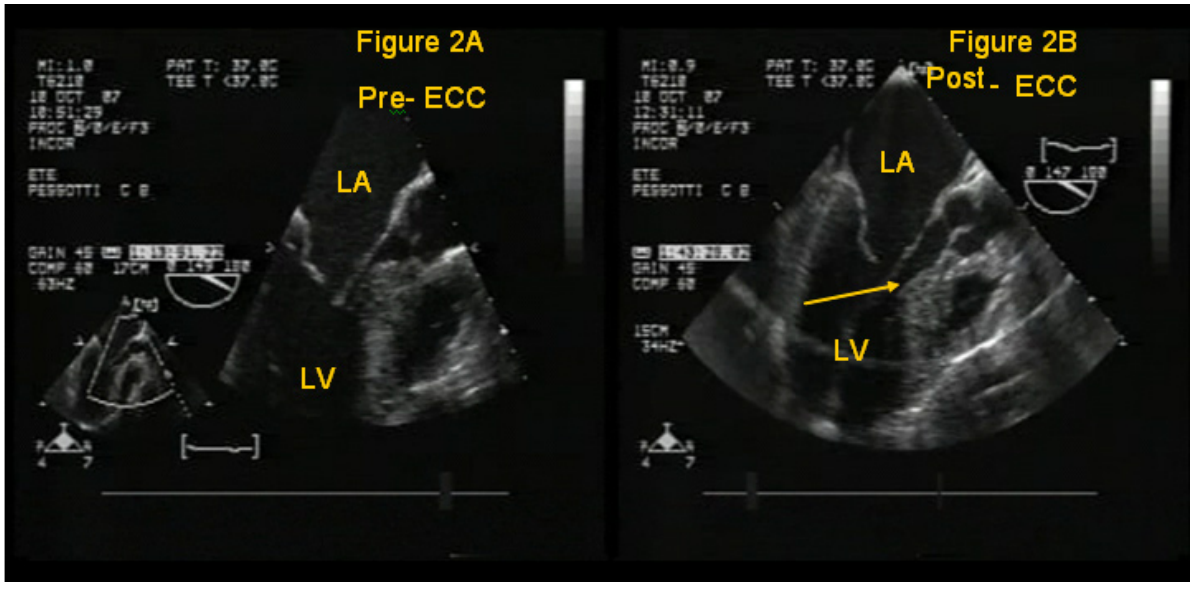

Figure 2 - Multiplane IOTEE of a patient with HCM; Figure 2A (prior to surgical correction), Figure 2B (after surgical correction). Arrow shows the surgical result after septal myectomy (anterior cusp of the mitral valve separated from the IVS); LA – Left Atrium; LV – Left Ventricle; ECC – Extracorporeal Circulation.

refractory to medical treatment and with LVOT gradient > 50 mmHg. The most frequently used surgical technique is septal myectomy, which consists of the excision of part of the hypertrophic septum via the transaortic approach.

There are limitations to the two-dimensional echocardiographic assessment regarding the anatomic analysis of patients with HCM. This results from geometric assumptions made because of the limited number of observation planes. In this context, three-dimensional echocardiography adds anatomic information, especially because of the position of the anterior cusp of the mitral valve in relation to the basal IVS segment (LVOT)[6]. 3-D echo permits the exact observation of the mitral valve cusp positioning during the septal myectomy procedure[7], as well as LVOT enlargement after alcoholization and myectomy[8].

Surgical treatment has become easier with the use of IOTEE, meant for planning the extent of surgical resection, assessing immediate results and ruling out possible complications.

Current operative mortality is lower than 5% in centers highly experienced in the surgical treatment of HCM, which is considered a safe procedure, with excellent long-term clinical and echocardiographic results.

In the case reported, the use of IOTEE permitted surgical planning and immediate knowledge of the operative success, making reintervention unnecessary.

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.

References

1. Spirito P, Seidman CE, Mc Kenna WJ, Maron BJ. The management of hypertrofic cardiomyopathy. N Engl J Med. 1997; 336: 775-85.
2. Sociedade Brasileira de Cardiologia. Diretriz para indicações e utilização da Ecocardiografia na prática clínica Arq Bras Cardiol. 2004; 82 (supl. 2); 11-34.
3. TTE/TEE Appropriateness Criteria Writing Group. Douglas PS, Khanderia B, Stainback RF, Weissman NJ, et al. ACCF/ASE/ACEP/ASNC/SCAI/SCCT/SCMR 2007 appropriateness criteria for transthoracic and transesophageal echocardiography: a report of the American College of Cardiology Foundation Quality Strategic Directions Committee Appropriateness Criteria Working Group, American Society of Echocardiography, American College of Emergency Physicians, American Society of Nuclear Cardiology, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, and the Society for Cardiovascular Magnetic Resonance. Endorsed by the American College of Chest Physicians and the Society of Critical Care Medicine. J Am Soc Echocardiogr. 2007; 20 (7): 787-805.
4. Hoss AJ, Petterson CH, Scherer L. Hypertrophic cardiomyopathy: role of Doppler echocardiography in the diagnosis and therapeutic approach. Arq Bras Cardiol. 1998; 70 (4): 301-6.
5. Silva MAD. Tratamento clínico, elétrico e cirúrgico da cardiomiopatia hipertrófica. Arq Bras Cardiol. 1996; 66 (2): 97-102.
6. De Gregorio C, Recupero A, Grimaldi P, Coglitore S. Can transtoracic live 3-D echocardiography improve the recognition of midventricular obliteration in hypertrophic obstructive cardiomyopathy? J Am Soc Echocardiogr. 2006; 19 (9): 1190.e1-4
7. Qin JX, Shiota T, Asher CR, Smedira NG, Shin JH, Agler DA, et al. Usefulness of real-time three-dimensional echocardiography for evaluation of myectomy in patients with hypertrophic cardiomyopathy. Am J Cardiol. 2004; 94 (7): 964-6.
8. Sitges M, Qin JX, Lever HM, Bauer F, Drinko JK, Agler DA, et al. Evaluation of ventricular outflow tract area after septal reduction in obstructive hypertrophic cardiomyopathy: a real-time 3-dimensional echocardiographic study. Am Heart J. 2005; 150 (4): 852-8.