

My Approach to Echocardiography in Radiofrequency Ablation of Hypertrophic Obstructive Cardiomyopathy

Como Eu Faço Ecocardiograma na Ablação por Radiofrequência da Cardiomiopatia Hipertrófica Obstrutiva

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Introduction

Hypertrophic cardiomyopathy (HCM), an autosomal dominant genetic disease that affects one in 500 people,¹ is considered the most common isolated form of hereditary heart disease. HCM is characterized by varying degrees of ventricular hypertrophy, myocardial fiber disarray, interstitial fibrosis, and microvascular disease in the absence of cardiac or systemic conditions to justify these findings.²

Echocardiography, an invaluable tool for the diagnosis and follow-up of patients with HCM, is used to assess morphology, hemodynamic disorders, left ventricular (LV) function, and patient prognosis.³ It also has a fundamental role in cases of hypertrophic obstructive cardiomyopathy (HOCM) in the indication of invasive treatments. Radiofrequency ablation (RFA) is a possible treatment under study.

After briefly reviewing HCM, we will describe the role of echocardiography in patient selection, intraprocedural RFA, and follow-up.

Genetic basis

HCM is caused by mutations in genes encoding the sarcomere protein, Z disk, or intracellular calcium modulators.⁴ More than 1,500 mutations have been identified among more than 11 genes.⁴

Pathophysiological mechanisms

The combination of left ventricular outflow tract (LVOT) obstruction (LVOTO), mitral regurgitation (MR), diastolic dysfunction, and myocardial ischemia comprise the pathophysiological basis of HCM.⁵

The distinction between non-obstructive HCM and HOCM has great clinical importance, as the two conditions have different therapeutic strategies and prognosis.⁶

HOCM has a prevalence of one in 1,500 people.

Keywords

Echocardiography; Cardiac Hypertrophy; Ablation, Radio Frequency.

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About one-third of patients with HCM have the non-obstructive form, one-third have significant obstruction at rest (defined as an instantaneous peak pressure gradient ≥ 30 mmHg), and one-third have latent obstruction (peak pressure gradient ≥ 30 mmHg after provocative maneuvers).⁶ A pressure gradient ≥ 50 mmHg is considered hemodynamically important.

Obstruction is determined by an interaction between the septum, mitral valve, and subvalvular apparatus in addition to flow vectors generated in the ventricular cavity. The mechanisms responsible for LVOTO are: a) septal hypertrophy with LVOT narrowing, which increases the velocity and displaces the blood flow trajectory, which end up “dragging” the anterior mitral valve leaflets toward the interventricular septum⁷ (Figure 1, Videos 1A and B); and b) morphological changes in the mitral valve apparatus (papillary muscles and anterior leaflet implantation and/or dimension) that may contribute to LVOTO.⁸

Left midventricular obstruction occurs in approximately 10% of patients with HCM; these patients often present with heart failure symptoms and are at increased risk of sudden death (SD) (Figure 2, Videos 2A and B).¹⁰

Mitral valve systolic anterior motion (SAM) is greater on the anterior versus posterior leaflet, resulting in coaptation distortion and dynamic mid-systolic regurgitation that is often directed to the lateral and posterior wall of the left atrium. The degree of MR varies according to the degree of LVOTO and the size of the posterior leaflet.¹⁰

The presence of a central or anterior jet should raise the suspicion of a primary mitral valve anomaly; in these cases, transesophageal echocardiography (TEE) is indicated to better elucidate the mechanism.²

The origins of the diastolic dysfunction and increased left ventricular filling pressures are multifactorial.¹¹ The hypertrophy, ischemia, and replacement fibrosis present in the myocardium are factors that increase myocardial stiffness and decrease compliance. The assessment of multiple parameters is recommended to define the degree of diastolic dysfunction (Figure 3).¹²

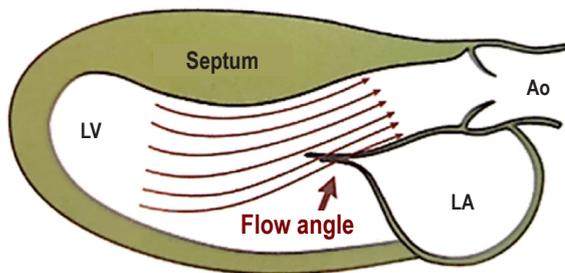
Diagnosis

The current diagnostic criteria for HCM are:

Maximum myocardial thickness measured in diastole > 15 mm of unexplained cause (in any LV segment); and

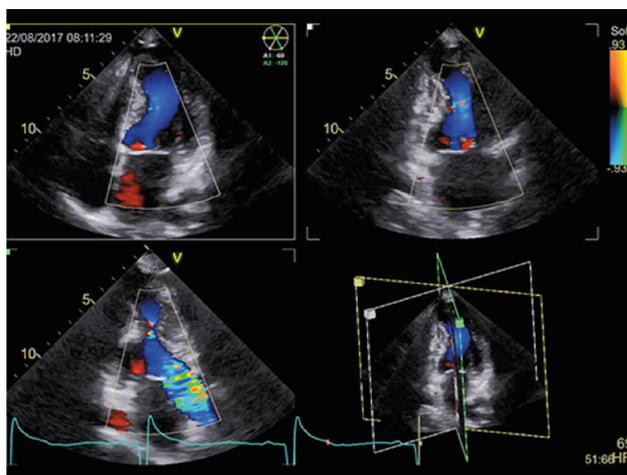
Septal and inferolateral wall thickness ratio > 1.3 in normotensive subjects or > 1.5 in hypertensive patients.⁸

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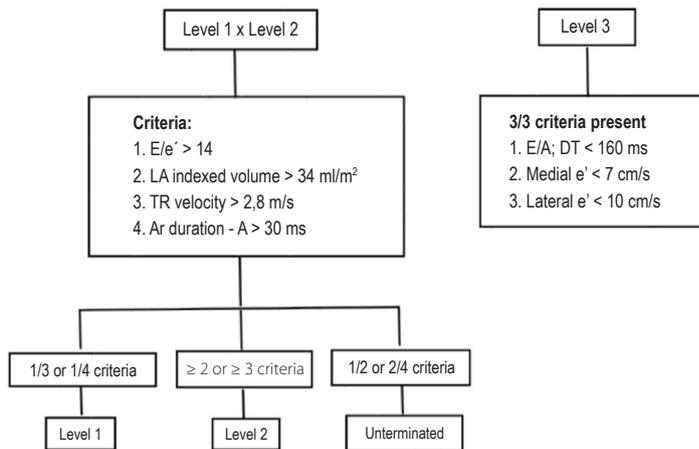
Source: Otto et al.⁹
LV: left ventricle; LA: left atrium; Ao: aorta.

Figure 1 – Flow deviation caused by septal hypertrophy, generating an ejection flow angled in relation to the edges of the mitral valve that is ideal for “dragging” the leaflets toward the septum.



Source: Echocardiography Service of Instituto Dante Pazzanese de Cardiologia.

Figure 2 – Hypertrophic cardiomyopathy with midventricular obstruction. Flow acceleration is visualized in three-chamber view (arrow).



Ar-A, duration of the pulmonary reversal A wave – mitral A wave; DT, deceleration time; LA, left atrium; TR, tricuspid regurgitation. Source: Elliott PM et al.²

Figure 3 – Algorithm used to determine the degree of diastolic dysfunction and filling pressures. It can be used regardless of the presence of LVOTO.

A myocardial thickness ≥ 13 mm may be considered in the diagnosis of HCM in patients with a family history of HCM (especially if the electrocardiogram findings are abnormal). The distribution pattern of hypertrophy varies, with the asymmetric septal form being more common (70% of cases).¹³

Treatment

The pharmacological treatment of HCM is indicated in symptomatic patients. Beta-blockers are the first choice, disopyramide the second option, and verapamil is indicated for patients who are contraindicated to beta-blockers.¹⁴

In cases of significant obstruction (gradient > 50 mmHg at rest or after a provocative maneuver) with symptoms refractory to drug treatment, invasive measures may be necessary to alleviate symptoms and reduce the intraventricular gradient.¹⁵

Two invasive treatment methods are traditionally used to alleviate LVOT: surgical myectomy (SM) and alcohol septal ablation (ASA). SM is considered the gold standard and consists of partial resection of the hypertrophied septum, reducing the dynamic outflow tract obstruction.² ASA induces necrosis of the hypertrophied myocardium through the injection of alcohol into the septal perforating artery. Its use requires adequate anatomy, with the performance of contrast echocardiography being mandatory. If the contrast is not exclusively visualized in the basal septum, the procedure should be canceled.²

Recent articles on RFA reported favorable results.¹⁶ RFA is an option in situations in which there are limitations to ASA and/or in midventricular obstructions. RFA is performed by electrophysiologists in a hemodynamics room and guided by a trained echocardiographer. Anatomical (place of greater thickness) and functional (place of greater velocity) information is analyzed on echocardiography, while electroanatomical mapping provides the other necessary information.¹⁶ The patient is examined under general anesthesia in the supine position. The right femoral artery is punctured for retroaortic access to the septal region of the left ventricle, while the right femoral vein is punctured for the positioning of a quadripolar catheter in the right septum (to identify the bundle of His) and in the tip of the right ventricle. The therapeutic catheter is impacted at the point of greatest obstruction, where the radiofrequency is applied for 120 seconds (80 W and 60°C).

Role of echocardiography

In planning (preprocedural)

The purpose of the pre-RFA assessment is to determine the site of greatest myocardial thickness, assess the point of greatest flow obstruction (greater aliasing), record the maximum peak gradient, and identify changes in the mitral valve apparatus. The identification of the major primary or mixed MR makes SM more appropriate due to the possible need for mitral valve replacement. On the other hand, the identification of a significant midventricular gradient makes SM less indicated.

The morphofunctional analysis of the mitral cusps, subvalvular apparatus, and papillary muscles can elucidate the pathological mechanism mostly by TTE. TEE can be used in cases of limited acoustic window or uncertain mechanism of MR by TTE.

The LVOT flow assessment should consider the shape of the curve recorded by continuous Doppler, which typically resembles a dagger. This flow with peak mid-systolic velocity often varies with the Valsalva maneuver or physical exercise (Figures 4 and 5). Take care not to mismeasure MR as an intraventricular gradient.

During the procedure (intraprocedural)

The initial assessment is performed by TEE and aims to confirm the previously described findings: maximum peak gradient, greater aliasing point, and MR degree. Data on systolic function, left ventricular segmental contractility, and pericardial effusion are extremely important as they are compared with post-RFA findings (Figure 6).

Zero degree angles (in middle esophagus for five-chamber visualization), angles between 120° and 150°, and deep transgastric angles are the most commonly used to assess the site of greatest turbulence and measure maximum peak gradient. The gradient analysis should always consider the patient's blood pressure since, in cases of hypotension (very common during anesthesia), the value of the aorta-ventricle gradient may be higher than the one recorded in a previous examination.

The access route is usually retroaortic, but in cases in which passage of the catheter through the aortic valve is impossible (stenosis and significant calcification), access can occur via the transeptal route. The myocardium will be more echogenic (thermal injury) and slightly hypokinetic after the ablation

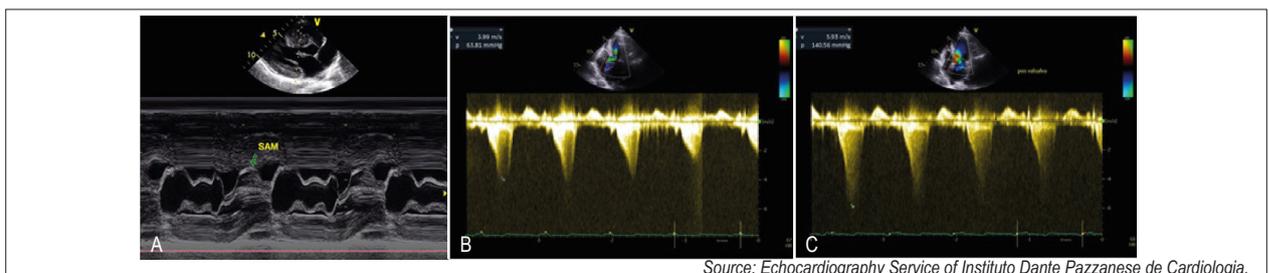


Figure 4 – Preprocedural transthoracic echocardiogram: (A) M mode showing systolic anterior motion; (B) and (C) left ventricular outflow tract flows before and after the Valsalva maneuver.

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(Figure 7). A decreased SAM, and consequently a decreased degree of valve insufficiency, is an indicator of a good response to therapy (Figure 8, Table 1, Videos 3A and B).

In the follow-up period (postprocedural)

TTE should be performed 24–48 hours after the procedure to record the maximum peak gradient. During this period,

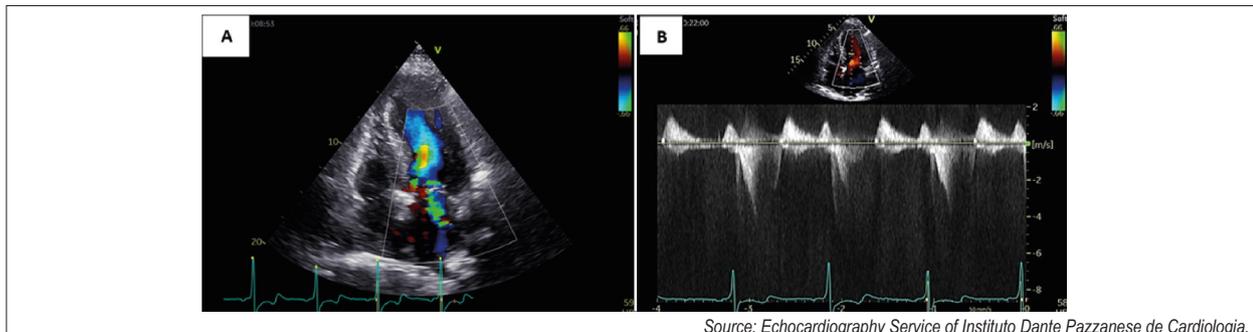


Figure 5 – Preprocedural transthoracic echocardiogram: (A) four-chamber apical view with color flow mapping showing the site of greatest systolic flow turbulence. (B) left ventricular outflow tract mid-systolic flow was also recorded in the spectral recording of mitral regurgitation.

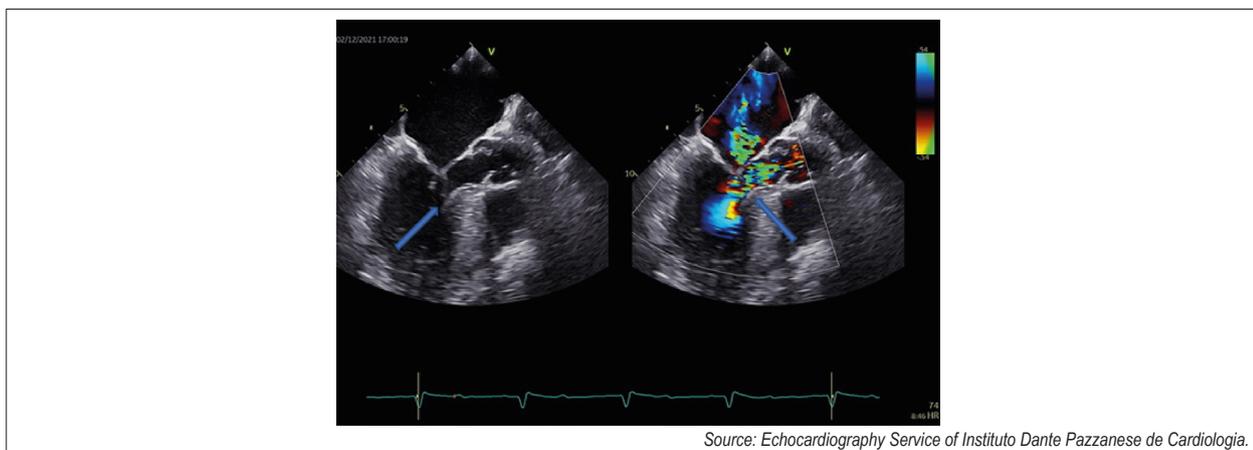


Figure 6 – Intraprocedural transesophageal echocardiography with versus without color (at 150°): mitral valve assessment and systolic anterior motion identification and location. At this angle, we could more easily identify the site of greatest flow acceleration and the degree of mitral regurgitation.

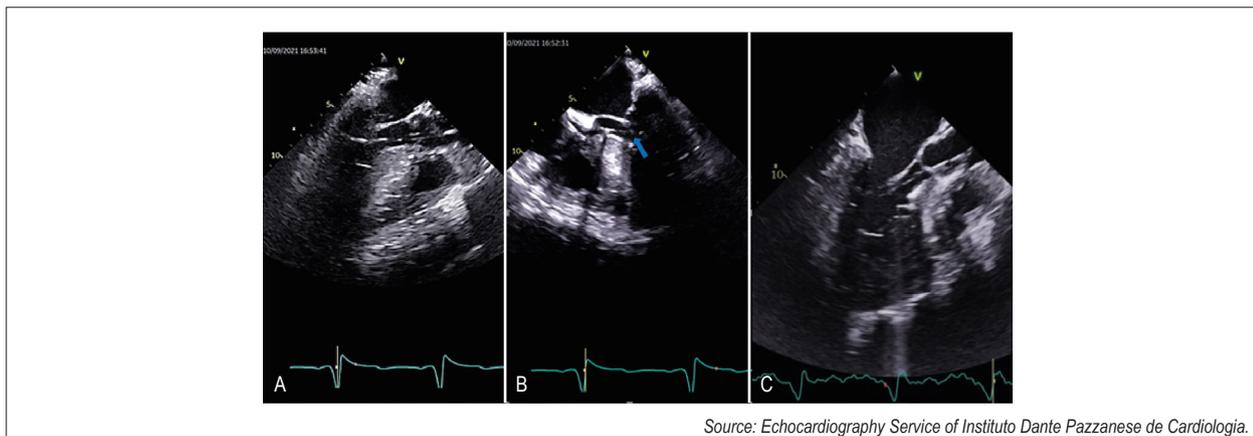


Figure 7 – Intraprocedural transesophageal echocardiography at 120° (A) and 0° (B) showing the passage of the retroaortic catheter and its repositioning in the site of greatest flow obstruction. Catheter at 120° (C) during radiofrequency application the moment the myocardium becomes more echogenic and hypokinetic due to the thermal injury.

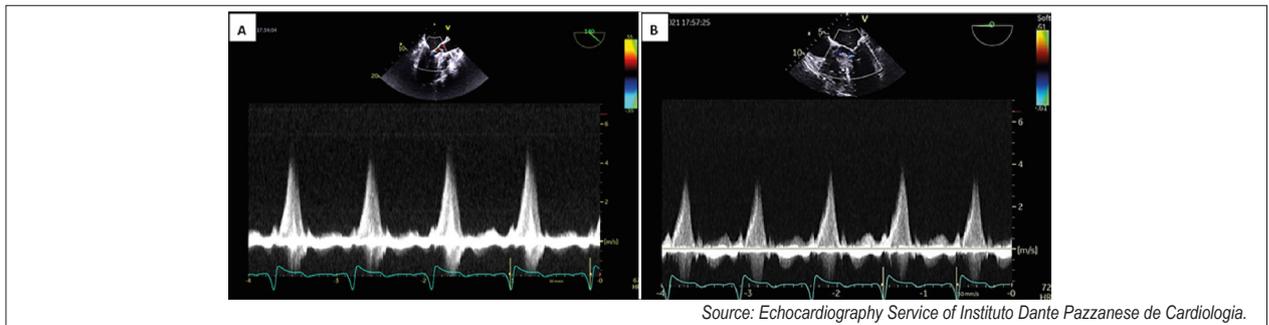


Figure 8 – Intraprocedural transesophageal echocardiography with recording of maximum left ventricular outflow tract gradients before (A) and after (B) radiofrequency ablation with estimated values of 96 mmHg and 58 mmHg, respectively.

Table 1 – Step-by-step echocardiography performed during the procedure.

Catheter access route and positioning
Step 1: Analyze gradients, valves, cavities, and pericardial space.
Step 2: Find where flow acceleration (SAM) occurs and what structures are involved.
Step 3: Guide the proper positioning of the ablation catheter (site of greatest acceleration).
During application
Step 1: Confirm that the catheter remains in the desired site.
Step 2: Confirm that the myocardium becomes hyperechoic, suggesting the therapy is effective.
Step 3: Confirm that the acceleration site changes with the application so that the catheter can be redirected, if necessary.
Step 4: Check the degree of MR, one indicator of therapeutic efficacy.
Step 5: Analyze the mitral and aortic valve apparatus to check if there was any damage during the procedure.

there are records of paradoxical gradient increase due to post-ablation wall edema.¹⁵ The use of periprocedural dexamethasone seems to reduce the edema; however, further studies are required to assess its efficacy.

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The literature reports progressively decreased gradients in up to a 1-year follow-up. TTE should be repeated after 3 months and annually thereafter if the clinical status shows no changes.¹⁶

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Authors' contribution

Research creation and design: Paladino AT, VILELA, AA; Data acquisition: Paladino AT, Valdigem B; Manuscript writing: Paladino AT, VILELA, AA; Critical revision of the manuscript for important intellectual content: Assef JE.

Conflict of interest

The authors have declared that they have no conflict of interest.

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