Malnutrition complicating clinical presentation of obstructive hypertrophic cardiomyopathy

Anastasiia Karadzha, MD, and Hartzell V. Schaff, MD, Rochester, Minn

From the Department of Cardiovascular Surgery, Mayo Clinic, Rochester, Minn.

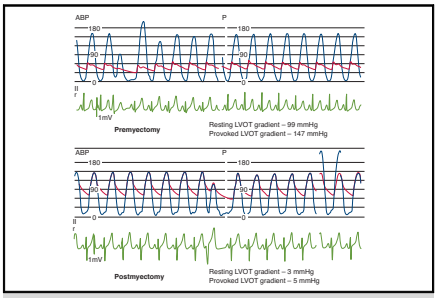
Received for publication June 30, 2023; revisions received July 31, 2023; accepted for publication Aug 14, 2023; available ahead of print Aug 28, 2023.

Address for reprints: Hartzell V. Schaff, MD, Department of Cardiovascular Surgery, Mayo Clinic, 1216 2nd St, SW, Rochester, MN 55902 (E-mail: schaff@mayo.edu).

JTCVS Techniques 2023;22:225-7

2666-2507

https://doi.org/10.1016/j.xjtc.2023.08.015



Direct measurement of left ventricular outflow tract gradient before and after myectomy.

CENTRAL MESSAGE

Postprandial symptom exacerbation is common in obstructive hypertrophic cardiomyopathy. Septal myectomy can relieve symptoms and improve diet and weight loss in severely symptomatic patients.

Postprandial symptom exacerbation (PPSE) is a clinical feature of several cardiac conditions, including obstructive hypertrophic cardiomyopathy (oHCM).[1] This phenomenon remains relatively unknown or underappreciated by many clinicians. Previous studies of patients with HCM have documented that meal consumption can trigger left ventricular outflow tract (LVOT) obstruction, and the presence of PPSE may be a marker of more severe disease.[2,3] Moreover, PPSE can contribute to poor nutrition which, in turn, compounds oHCM-related symptoms of fatigability and effort dyspnea.

In this case report, we present an unusual instance of extreme PPSE leading to malnutrition and weight loss in a patient with oHCM. Our aim is to increase awareness of the correlation between PPSE and cardiac pathology to improve patient management. Institutional review board approval is waived for single case reports at our institution. The patient provided informed written consent for the publication of her study data.

CASE PRESENTATION

A 49-year-old woman with oHCM was referred for further management of debilitating symptoms. Fourteen years before presentation, she underwent double lung transplant for cystic fibrosis. Her posttransplant course was complicated by chronic rejection with bronchiolitis, and pulmonary function was reduced as reflected by forced expiratory volume in 1 second that was approximately 40% of predicted normal. She described herself as markedly limited in activity. Additionally, she reported experiencing early satiety and chest fullness after meals that led to a progressive reduction in dietary intake and activity level. She reported a 13-kg weight loss to her weight at presentation of 45.5 kg (body mass index [BMI], 16).

Initially, her deterioration was attributed to pulmonary disease based on her medical history. However, upon further evaluation, a transthoracic echocardiogram revealed severe oHCM, with a resting LVOT gradient of 104 mm Hg. Medical treatment was unsuccessful, and consultation with a transplant pulmonologist led to a recommendation of feeding tube placement to support nutrition and achieve a BMI of 18.5 before undergoing septal reduction. But due to the persistence of symptoms and uncertainty of the benefit of tube feedings, operation for septal myectomy was ultimately recommended.

Transaortic septal myectomy was performed through a secondary sternotomy, with a bypass time of 23 minutes and an aortic crossclamp time of 19 minutes. The LVOT gradient was 147 mm Hg before and 5 mm Hg after myectomy (Figures 1 and 2). Postoperative recovery was uncomplicated, and she was dismissed on the fourth postoperative day.

Three years postoperatively, the patient's condition had significantly improved. She no longer experienced cardiac symptoms following meals or during physical exertion, and her weight had increased to 54.7 kg (BMI, 19.6).

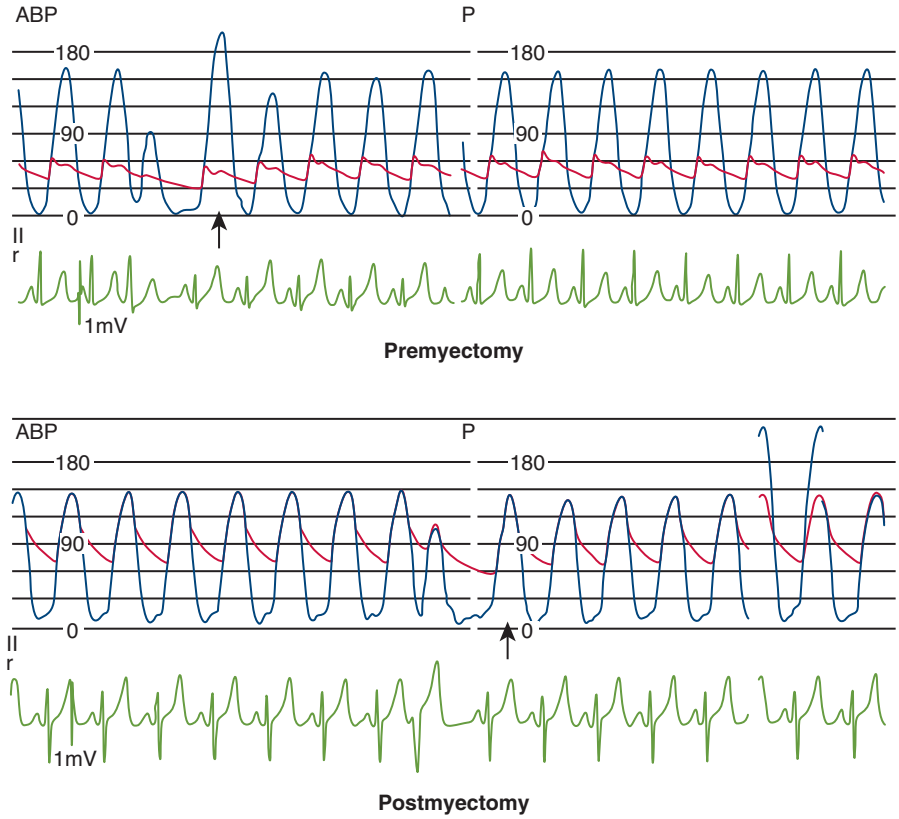


FIGURE 1. Intraoperative direct measurements of left ventricular and aortic pressures premyectomy (*upper panel*) and postmyectomy (*lower panel*). The arrows indicate the augmented beat after a premature ventricular contraction (PVC). Before myectomy the resting left ventricular outflow tract (*LVOT*) gradient was 99 mm Hg, and the post-PVC gradient was 147 mm Hg. Following septal myectomy the resting LVOT gradient was 3 mm Hg, and the post-PVC gradient was 5 mm Hg.

COMMENT

The incidence of PPSE among HCM patients ranges from 33% to 37%.[1,3] Symptoms that can occur and worsen after meals vary widely. The most frequent symptoms include chest pain, dyspnea, palpitation, presyncope, and syncope. In addition to the classic HCM symptoms, postprandial symptoms can be less specific, such as a warm sensation or feelings of heaviness or sleepiness.[1-3] PPSE typically manifests around 15 minutes after a meal, reaches its peak at 30 to 60 minutes, and can persist for several hours thereafter.[1-3]

Gilligan and colleagues'[1] pioneering study in 1991 demonstrated that food intake in HCM patients leads to a decrease in peripheral vascular resistance and an increase in heart rate. But due to limited LV diastolic filling, the stroke volume fails to increase, causing a worsening of LVOT obstruction. Adams and colleagues[3] found a strong correlation between the presence of PPSE in patients with HCM and higher LVOT gradient, obstruction at rest, New York Heart Association functional class III or IV dyspnea, the presence of presyncope, and a diminished quality of life. These findings support the hypothesis that PPSE is associated with the severity of the disease. Provocative testing using food intake during stress echocardiography can reveal latent LVOT obstruction. In a study by La Canna and colleagues,[4] different methods of gradient provocation were compared among patients considered to have non-oHCM. The investigators found that 23% of patients were reclassified as oHCM after the fasting Valsalva maneuver, 33% after treadmill exercise testing, and 74% after the postprandial test.

Some clinicians have recommended alterations in dietary intake as an adjunct to medical management in patients with HCM with PPSE. A study by Kansal and colleagues[5] demonstrated that specific dietary restrictions, such as consuming small frequent meals and avoiding caffeinated fluids, can significantly alleviate symptoms, particularly in patients resistant to medical treatment.

Postprandial symptom exacerbation is a common and troubling phenomenon in patients with oHCM. Many patients will alter their dietary habits and daily routines to minimize symptoms. In the patient presented here, chronic PPSE led to marked reduction in caloric intake and malnutrition. Relief of LVOT obstruction eliminated PPSE,

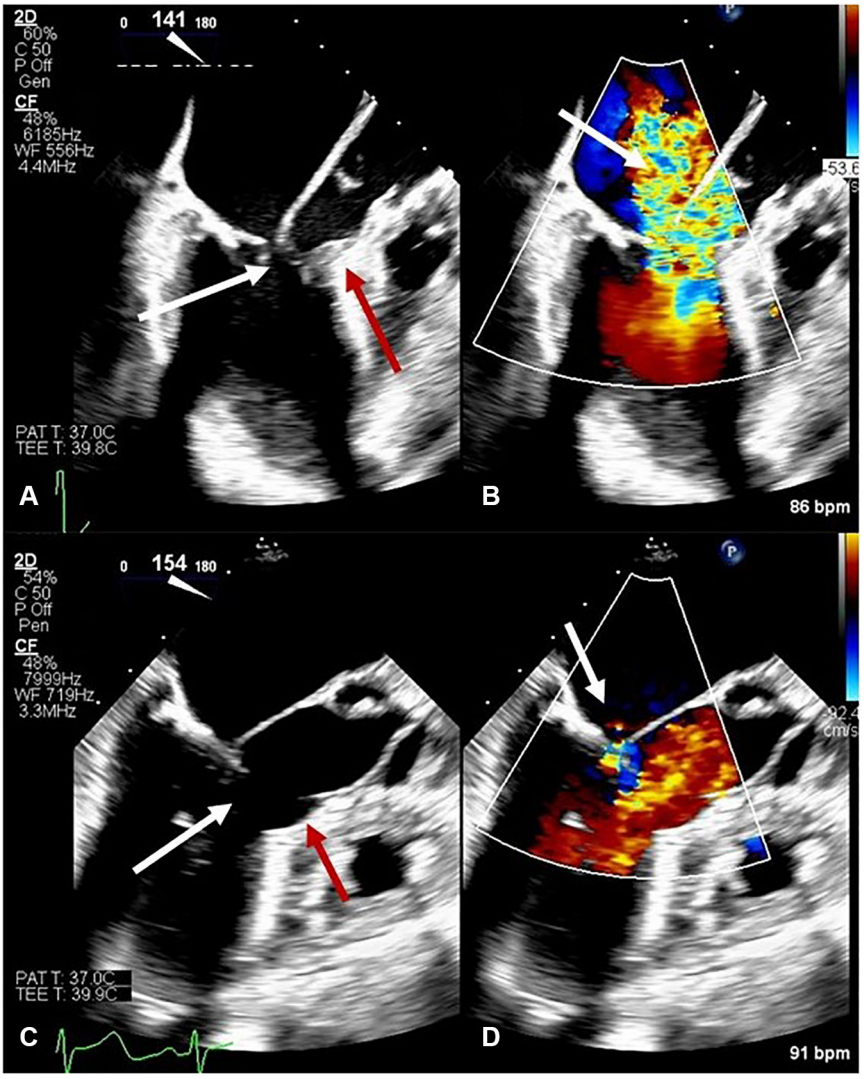


FIGURE 2. Intraoperative transesophageal echocardiography before and after septal myectomy. A, Systolic anterior motion of the mitral leaflets (*white arrow*) and subaortic septal hypertrophy (*red arrow*). B, The white arrow points to the Doppler signal of severe mitral valve regurgitation. C, Following septal myectomy the left ventricular outflow tract is widely patent (*white arrow*), and the myectomy site is easily visible (*red arrow*). D, Only trivial residual mitral regurgitation (*white arrow*).

which improved diet and weight gain. This case report emphasizes the importance of recognizing and understanding the relationship between postprandial symptoms and HCM.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

1. Gilligan DM, Nihoyannopoulos P, Fletcher A, Sbarouni E, Dritsas A, Oakley CM. Symptoms of hypertrophic cardiomyopathy, with special emphasis on syncope and postprandial exacerbation of symptoms. *Clin Cardiol*. 1996;19:371-8.
2. Gilligan DM, Chan WL, Ang EL, Oakley CM. Effects of a meal on hemodynamic function at rest and during exercise in patients with hypertrophic cardiomyopathy. *J Am Coll Cardiol*. 1991;18:429-36.
3. Adams JC, Ommen SR, Klarich KW, Tajik AJ, Nishimura RA. Significance of postprandial symptom exacerbation in hypertrophic cardiomyopathy. *Am J Cardiol*. 2010;105:990-2.
4. La Canna G, Scarfò I, Arendar I, Alati E, Caso I, Alfieri O. Phenotyping left ventricular obstruction with postprandial Re-test echocardiography in hypertrophic cardiomyopathy. *Am J Cardiol*. 2020;125:1688-93.
5. Kansal MM, Mookadam F, Tajik AJ. Drink more, and eat less: advice in obstructive hypertrophic cardiomyopathy. *Am J Cardiol*. 2010;106:1313-6.