Mechanical or neurohumoral left ventricle ballooning: Challenging to differentiate between and to manage. Authors' reply

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We appreciate the interest of the authors of the letter [1] and their insightful comments on our article. In cases of left ventricle (LV) ballooning with left ventricular outflow track obstruction (LVOTO), distinguishing between mechanical and neurohumoral causes can be challenging not only due to similar demographics (studies indicate that both phenotypes occur most often in postmenopausal women), but also because mechanical apical ballooning usually occurs in patients with mild hypertrophy. Significant hypertrophy appears to have a protective effect in cases of a sudden increase in afterload. Moreover, both the incidence of a sudden LVOTO in hypertrophic cardiomyopathy (HCM) and the incidence of takotsubo syndrome can be provoked by stress. As the authors of the letter note, severe hemodynamic instability (cardiogenic shock, refractory heart failure) occurs more often in mechanical than in neurohumoral left ventricle apical ballooning, while our patient's condition remained stable. However, out of 13 cases of mechanical apical ballooning in HCM described by Sherrid et al. [2], only half presented symptoms of hypotension or cardiogenic shock. It is worth emphasizing that cardiogenic shock caused by mechanical apical ballooning is treatable and that a prompt diagnosis is of particular importance because in cases of ineffectiveness of typical pharmacological treatment, such as beta blockers, intravenous fluids, and pure alpha-adrenergic agonist therapy, urgent surgical relief of LVOTO should be considered.

After the acute phase, it is necessary to closely monitor the patient's clinical symptoms (mainly exercise tolerance), LV systolic function, and the gradient through the left ventricular outflow track. The resolution of LV regional contractility disorders and the normalization of the flow gradient through the LVOT may indicate takotsubo syndrome as the cause of a left ventricular ballooning incident, while its mechanical nature may be proven by a latent gradient in LVOT at rest or provoked by a Valsalva maneuver or on stress echocardiography. In such a case, further steps are required. In HCM this involves risk stratification in the patient and screening tests for his or her relatives, but also the implementation of appropriate therapeutic steps in cases of symptomatic, significant LVOTO i.e., pharmacological and, if that proves ineffective, invasive. It is worth noting that the LV hypertrophy in our patient was mild i.e., up to 15 mm. In the past, myectomy was considered contraindicated in patients with HCM and hypertrophy of less than 18 mm, mostly due to the increased risk of iatrogenic ventricular septal defect [3]. However, in light of the significant improvements in myectomy techniques and a significant decrease in mortality over the years, myectomy is now considered one of the safest open-heart procedures when performed in experienced centers [4]. Accordingly, in a recently published study by Rowin et al. [5], as many as 40% out of 113 patients with mild LV hypertrophy (≤15 mm) but

significant LVOTO developed symptoms sufficient to become candidates for surgical intervention, and in these patients shallow myectomy combined with interventions on intraventricular structures and mitral valve apparatus was not only effective in reducing symptoms, but also safe (no iatrogenic ventricular septal defect or perioperative deaths).

The question remains about the preventive measures for a sudden LVOTO recurrence in our patient, who during the 3-month follow-up has no latent gradient through the LVOT (at rest or after the Valsalva maneuver), and does not present clinical symptoms, although the residual regional contractility disorders partially persist. After their resolution, we may consider performing a stress echocardiography, mainly to confirm the mechanical nature of the apical ballooning incident. This would seem particularly important due to the fact that in the study by Rowin et al. [5], in 73% of symptomatic patients with HCM and mild hypertrophy, a significant gradient was generated only during exercise. However, it is difficult to determine the clinical significance of an exercise-provoked LVOT gradient in an asymptomatic patient shortly after a mechanical apical ballooning incident. The mainstay of LVOTO recurrence prevention in our patient is surely pharmacotherapy as well as non-pharmacological measures such as avoiding dehydration. Mavacamten, however promising, has a strongly negative inotropic effect and may further depress the left ventricular function in cases of a recurrent, sudden, breakthrough LVOTO. Invasive procedures such as myectomy or alcohol ablation are indicated only in symptomatic patients with a persistent gradient of >50 mm Hg refractory to medical therapy [4].

In summary, mechanical left ventricle ballooning syndrome is an extremely rare entity, without a clearly defined management path to date. Further research is needed, and considering the casuistic nature of this clinical presentation of hypertrophic cardiomyopathy, sharing experiences in this field is of the utmost importance.

Article information

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