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BRIEF REPORT

**Chris Adamopoulos**
**Matthew Tsagourias**
**Kostoula Arvaniti**
**Fotini Veroniki**
**Dimitrios Matamis**

# Weaning failure from mechanical ventilation due to hypertrophic obstructive cardiomyopathy

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C. Adamopoulos · M. Tsagourias ·
K. Arvaniti · F. Veroniki · D. Matamis (✉)
ICU,
Papageorgiou General Hospital,
Ring Road, Nea Efkarpia, 56403 Salonika,
Greece
e-mail: dmatamis@otenet.gr

**Abstract** *Objective:* To report the importance of hypertrophic obstructive cardiomyopathy as a potential substrate for difficult weaning from mechanical ventilation. *Design and setting:* Case report in an adult intensive care unit of a tertiary care hospital. *Patients:* Two patients who developed intractable pulmonary edema with subsequent difficult weaning from mechanical ventilation in the early postoperative period despite medical treatment for heart failure based on pulmonary artery catheter data. *Interventions:* Mechanical ventilation, medical treatment. *Measurements and results:* Pulmonary artery catheter data in these two elderly patients under mechanical ventilation were compatible with congestive heart failure, and the initial therapy was diuretics, vasodilators, and inotropes. Despite this reasonable medical treatment no hemodynamic improvement and thereby failure of weaning from mechanical ventilation was observed. Transesophageal echocardiography demonstrated hypertrophic obstructive cardiomyopathy in both cases, and therapy based on negative inotropes was instituted. This treatment change induced hemodynamic improvement and successful weaning from mechanical ventilation. *Conclusions:* In mechanically ventilated patients because of congestive heart failure who have difficult weaning, despite appropriate therapy, intensivists should always suspect causes other than the obvious systolic heart failure. In this clinical setting echocardiography is warranted and produces excellent diagnostic yield for clarifying complex cardiac disturbances.

**Keywords** Mechanical ventilator weaning · Hypertrophic cardiomyopathy · Echocardiography

## Introduction

Although hypertrophic cardiomyopathy (HCM) is not a rare clinical entity, its presence is often underestimated in critically ill patients. Lung congestion and low cardiac output syndrome are commonly encountered in mechanically ventilated patients. These disturbances if untreated may impede weaning from mechanical ventilation by increasing the work of breathing and decreasing blood flow to the respiratory muscles. Although congestive heart failure (CHF) is the obvious substrate for these abnormalities, the recognition of the exact pathophysiological mechanism is of capital importance. Pulmonary congestion in heart failure patients is due to elevated pulmonary venous pressure, and CHF may subsequently be present in patients with diastolic dysfunction who do not have impaired systolic function. HCM is a clinical condition that may present the same features as CHF. However, treatment usually administered for classical "systolic" CHF (vasodilators, diuretics, and positive inotropic agents) may be detrimental in this condition. On the other hand, drugs usually contraindicated in the case of CHF such as negative inotropes may dramatically improve clinical course in HCM. In this report we describe

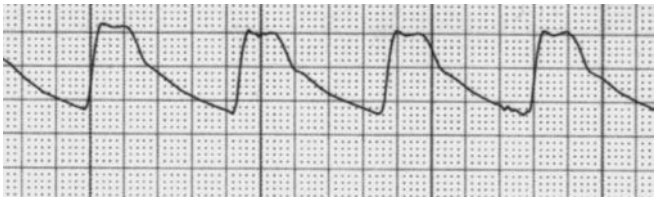

**Fig. 1** Pulsus bisferiens. Two systolic peaks separated by a midsystolic dip

the course of two patients who had HCM and developed intractable pulmonary edema and weaning failure despite reasonable medical treatment according to pulmonary artery catheter (PAC) data.

## Case report

Two women, 74 and 73 years old without any previous medication, smoking history, or other known comorbidity factors except moderate obesity, were admitted to our hospital for surgical repair of hip fracture. Patients are not presented separately because they share quite similar clinical, hemodynamic, and respiratory features. Preoperative assessment in both patients revealed normal lung auscultation, moderate cardiac enlargement on radiography, and exertional dyspnea attributed to their obesity and reduced physical reserve. Electrocardiography showed atypical ST segment and T wave abnormalities and tall QRS complexes in the first patient and the presence of Q waves in the inferior leads in the second. Both patients developed pulmonary edema and acute respiratory failure in the immediate postoperative period while recovering from regional anesthesia. They were intubated and mechanically ventilated, and a PAC was inserted for hemodynamic management. PAC revealed low cardiac output and high filling pressures and initial treatment consisted of diuretics (because of a positive intraoperative fluid balance of 1.5 l), vasodilators (nitroglycerin), and inotropes (dobutamine), considering that these aged patients were suffering from acute decompensation of preexisting and unrecognized congestive heart failure. Despite the above treatment and additional diuretic therapy, leading to a negative fluid balance of 2.5 l, both patients maintained low cardiac output and high filling pressures. The first 24 h they remained in the recovery room sedated with remifentanyl and propofol, and no weaning trial was attempted. On the second postoperative day they were transferred to the ICU for further management and weaning from mechanical ventilation. In the ICU lung auscultation and radiography demonstrated fulminant pulmonary edema, heart auscultation revealed a harsh systolic murmur best heard in apex in both patients, and a pulsus bisferiens was recorded on arteriography (Fig. 1). Transesophageal echocardiography (TEE) was performed to assess cardiac function because of the poor acoustic window in the transthoracic approach. In both patients a typical echocardiographic pattern of HCM was observed with septal hypertrophy of 2.1 and 2.2 cm, respectively, systolic anterior motion of the mitral valve, severe mitral regurgitation and dynamic left ventricular outflow tract (LVOT) obstruction gradient of 92 and 108 mmHg, respectively (Figs. 2, 3, Table 1). Based on the TEE data initial therapy with inotropes and vasodilators was interrupted, and hemodynamic improvement was observed while the patients remained sedated and mechanically ventilated. As soon as sedation was interrupted, and the weaning process was initiated according to our standard protocol, consisting of gradual reduction (every 2–4 h) of pressure support ventilation from 20 to 10 $cmH_2O$ followed by a 30 min T-piece trial [1, 2,3], the patients rapidly deteriorated clinically and hemodynamically. They developed signs of distress, diaphoresis, agitation, tachypnea, tachycardia, low cardiac output, and high filling pressures. A negative inotropic therapy with $\beta$-blockers (atenolol 200 mg/day) was started with moderate hemodynamic improvement, and therefore we cautiously added calcium antagonists (diltiazem 300 mg/day). Under this double-negative inotropic treatment dynamic LVOT obstruction and mitral regurgitation markedly decreased, pulmonary edema resolved, and cardiac index and tissue peripheral oxygenation improved (Table 1). The first patient was successfully weaned from mechanical ventilation on the 6th and the second on the 4th postoperative day. Data from PAC and echocardiography on admission after initial treatment and sedation interruption, and after treatment for HCM are presented in Table 1.

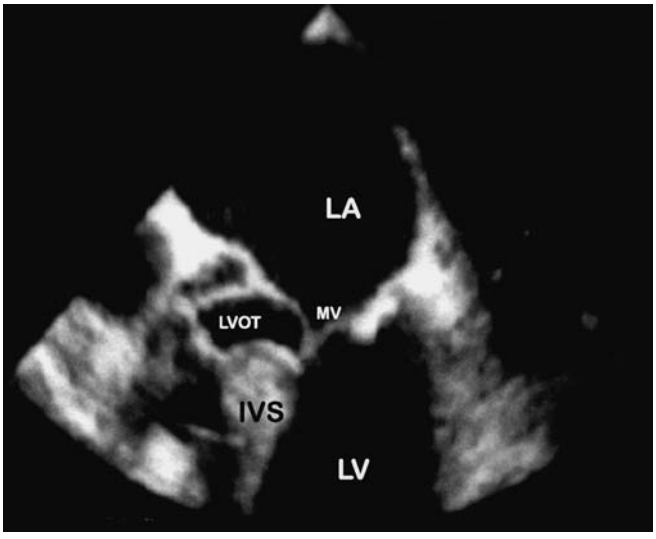


**Fig. 2** TEE four-chamber view. Systolic anterior motion of the mitral valve and marked septal hypertrophy resulting in LVOT obstruction

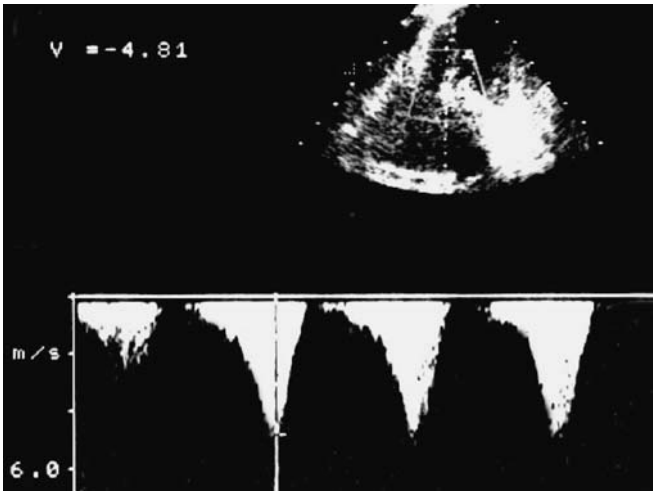


**Fig. 3** Continuous-wave Doppler shows the characteristic late-peaking high-velocity curve (Vmax=4.81 m/s) due to dynamic left ventricular outflow obstruction. GRADmax=92 mmHg

**Table 1** Patients' clinical data [*(a-v)DO$_2$* arteriovenous oxygen content difference, *ABP* arterial blood pressure, *CI* cardiac index, *CVP* central venous pressure, *HR* heart rate, *IVS* interventricular septum, *LVOT* left ventricular outflow tract, *PaO$_2$/FIO$_2$* ratio of arterial oxygen pressure to fractional inspiratory oxygen, *PAOP* pulmonary artery occluded pressure, *PAP* mean pulmonary arterial pressure, *PVRI* pulmonary vascular resistance index, *SVI* stroke volume index, *SVRI* systemic vascular resistance index]

| | Patient no. I | | | Patient no. II | | |
|---|---|---|---|---|---|---|
| | Initial treatment | Initial treatment interruption | Negative inotropic treatment | Initial treatment | Initial treatment interruption | Negative inotropic treatment |
| Hemodynamic data | | | | | | |
| HR (beats min$^{-1}$) | 100 | 120 | 70 | 125 | 120 | 85 |
| ABP (mmHg) | 150/78–106 | 165/85–112 | 128/70–82 | 125/80–88 | 135/85–90 | 120/70–82 |
| PAP (mmHg) | 103/46–68 | 105/45–68 | 90/40–60 | 55/30–42 | 60/35–45 | 40/18–30 |
| PAOP (mmHg) | 30 | 35 | 20 | 28 | 30 | 16 |
| CVP (mmHg) | 16 | 16 | 14 | 12 | 13 | 10 |
| CI (l/min$^{-1}$ m$^{-2}$) | 2.3 | 2.2 | 3.1 | 1.9 | 1.8 | 2.7 |
| SVI (ml/beat$^{-1}$ m$^{-2}$) | 23 | 18 | 44 | 15 | 15 | 32 |
| SVRI (dyne s$^{-1}$ m$^{-12}$ cm$-^{5}$) | 3,130 | 3,491 | 1,755 | 3,200 | 3,422 | 2,133 |
| PVRI (dyne s$^{-1}$ m$^{-2}$ cm$^{-5}$) | 1,322 | 1,200 | 1,032 | 589 | 667 | 415 |
| (a-v)DO$_2$ (ml O$_2$ dl$^{-1}$) | 5.8 | 5.8 | 4.6 | 6.6 | 6.4 | 5.8 |
| PaO$_2$/FIO$_2$ | 158 | 140 | 284 | 163 | 154 | 263 |
| Echocardiographic data | | | | | | |
| LVOT gradient (mmHg) | 92 | 90 | 23 | 108 | 105 | 41 |
| Mitral regurgitation | +3/4 | +3/4 | Trivial | +3/4 | +3/4 | Trivial |
| IVS thickness (mm) | 21 | | | 22 | | |
| Treatment | Dobutamine (5 μg/kg per minute), nitroglycerin (1 μcg/kg per minute), sedation | No treatment, no sedation | Atenolol (200 mg/ day per os), diltiazem (300 mg/day i.v.), no Sedation | Dobutamine (8 μg/kg per minute), nitroglycerin (0.5 μg/kg per minute), sedation | No treatment, No sedation | Atenolol (200 mg/ day per os), diltiazem (300 mg/day i.v.), no sedation |

## Discussion

This report demonstrates that failure in the diagnosis of HCM can seriously jeopardize the weaning process from mechanical ventilation in critically ill patients. It also emphasizes the role of TEE as a diagnostic tool in cases of difficult weaning due to complex cardiac pathophysiological mechanisms. HCM is not a rare entity. Epidemiological investigations have shown its prevalence in the adult general population at 0.2% [4]. However, clinicians and particularly intensivists have limited exposure to this disease, and this accounts for the uncertainty that prevails its diagnosis and management. The clinical course of HCM is unique among cardiovascular diseases by virtue of its potential for clinical presentation at any phase of life. Elderly patients (aged over 75 years) represented 25% of an HCM cohort [5].

The importance of prompt and accurate diagnosis of HCM in the ICU is particularly stressed in this report. Past medical history is often unavailable or unhelpful. Patients may be asymptomatic until exacerbation or have symptoms such as exertional dyspnea and angina that can be attributed to CHF. Electrocardiographic patterns, although abnormal in most cases, are not specific for the disease [6]. PAC, which is very popular and extensively used in critical care, fails to make the diagnosis between heart diseases that may lead to pulmonary edema. Echocardiography remains the unique tool for the diagnosis of HCM. Echocardiographic findings (Figs. 2, 3) include septal hypertrophy (2.1 and 2.2 cm in our patients) with small LV cavity, systolic anterior motion of the mitral valve resulting in LVOT obstruction (92 and 108 mmHg in our patients), mitral regurgitation, and diastolic dysfunction [7]. Although HCM and dilated CHF may share common clinical and PAC findings, factors that may decompensate a patient with HCM are entirely different. A decrease in preload or afterload and/or an increase in contractility may seriously exacerbate or unmask HCM. In our two patients a decreased preload as a result of diuretics and nitrates, enhanced contractility induced by positive inotropic therapy in the recovery room, and/or release of endogenous catecholamines (e.g., postoperative pain, anxiety, and discomfort) during the weaning period in the ICUmay have caused HCM exacerbation leading to refractory pulmonary edema responsible for weaning failure.

In cases of HCM erroneously treated as CHF positive inotropic agents, diuretics, and vasodilators should be discontinued, and volume replacement must be undertaken according to volume status assessed by echocardiography. If no improvement is observed, negative inotropic drugs, particularly $\beta$-adrenoreceptor blockers, are the first choice medical therapy [6]. $\beta$-Blockade decreases the dynamic LVOT obstruction induced by endogenous catecholamines and improves left ventricular compliance and therefore diastolic function. Calcium antagonists may be efficacious in patients responding inadequately to $\beta$-blockers. Although verapamil has been the most widely used calcium antagonist in this condition, diltiazem has also shown beneficial effects in HCM [8]. Double-negative inotropic treatment, although exceptional in the literature, induced a dramatic reduction in mitral regurgitation (from +3/4 to trivial in both), dynamic LVOT obstruction (from 92 to 23 and 108 to 41 mmHg, respectively) and cardiac filling pressures in both patients. Consequently pulmonary edema resolved and weaning

proceeded successfully. Weaning from mechanical ventilation was more prolonged in the first patient (6 days) because, as seen in Table 1, she was also suffering from severe pulmonary hypertension.

In summary, we present two patients with weaning failure due to intractable pulmonary edema, attributed to CHF according to PAC data. TEE disclosed HCM in both cases, and the patients were successfully weaned from mechanical ventilation following appropriate treatment. We conclude that in cases of difficult weaning due to persistent cardiogenic pulmonary edema despite appropriate therapy intensivists should always suspect more complex pathophysiological mechanisms of heart failure than the obvious CHF. In these cases echocardiography may be of great help [9].

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