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Letter To The Editor

Left Ventricular Ejection Fraction Cannot Be the Only Outcome Variable of Home Mechanical Ventilation in Sarcoglycanopathies

Claudia Stöllberger.[1], Josef Finsterer, MD, PhD.[2]

[1]Medical Dpt, Krankenanstalt Rudolfstiftung Vienna, Austria.
[2]Krankenanstalt Rudolfstiftung Messerli Institute, Veterinary University of Vienna Austria.

*Corresponding Author
Finsterer J, MD, PhD

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In a recent article, Faysoill *et al.*, (2018) reported about the effects of home mechanical ventilation (HMV) on left ventricular function in 10 patients with genetically proven sarcoglycanopathy (Faysoill *et al.*, 2018). After a median follow-up of 3 years, an increase of left ventricular ejection fraction (LVEF) was observed. We have the following questions and concerns:

Regarding the inclusion criteria it is stated that patients were included before introduction of HMV. Thus, we do not understand why patient #6 was included, who was already under invasive mechanical ventilation at recruitment.

Regarding the study design and follow-up duration, it remains unclear which endpoints were chosen.

Regarding pharmacotherapy, it remains unclear why patient #1 did not receive antihypertensive medication since her blood pressure at inclusion was 180/120 mmHg. We should also be informed how many patients started with pharmacotherapy during the follow-up period.

In the methods it is indicated that electrocardiograms were recorded but no results are presented. Among the echocardiographic results, only LVEF is reported. Were there other echocardiographic abnormalities? Were there any patients with myocardial thickening, dilation of atria or ventricles, or left ventricular hypertrabeculation / noncompaction (LVHT), a cardiac abnormality frequently associated with neuromuscular disorders and also with limb girdle muscular dystrophy (Finsterer, J. *et al.*, 2014)?

How did neuromuscular abnormalities change during follow-up? Did the time of mechanical ventilation increase during follow-up due to increasing weakness of the respiratory muscles?

Organization of HMV requires a high degree of familial and social support. Thus, it is interesting to know how HMV was managed by the patients and their families. Which measures for airway clearance techniques were applied (Chatwin, M. *et al.*, 2018)? Were there any patients who denied HMV and what was the course of their disease?

Forty percent of the patients suffered from sleep apnea which is associated with changes in the autonomic nervous system. Were any investigations of the positive effects of HMV carried out in the patients with sleep apnea? Were there any changes in heart rate observed before and after initiation of HMV?

In the concluding paragraph, the authors state that HMV in patients with sarcoglycanopathies is not harmful and may protect left ventricular function. However, HMV may also be associated with side effects. Thus, it would be interesting to know if HMV-treated patients suffered from any side effects like respiratory infections or pneumothorax.

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The main finding of the study was that LVEF increased during a three year period of HMV in patients with sarcoglycanopathies. Did the authors attribute this improvement only to HMV or did the patients also receive cardiac therapy that could explain the increase of the LVEF during follow-up? At least one patient received angiotensin converting enzyme inhibitors (ACEI) and beta-blockers and one patient ACEI, beta-blockers, and diuretics.

We conclude that when investigating the benefits of HMV in sarcoglycanopathies, not only the LVEF but also other cardiac and extra-cardiac aspects have to be considered. Consideration of medical and social aspects will make it easier to assess the benefits of HMV in patients with sarcoglycanopathies.

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