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Letter to the Editor

Adherence to Home Ventilation in Myotonic Dystrophy-1 may not only depend on Cognition, Mood and Apathy

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We were interested to read the article by Vosse *et al.* on a cross-sectional study on the influence of cognition, mood and apathy on adherence to home mechanical ventilation (HMV) in patients with myotonic dystrophy type 1 (MD1) using the Montreal Cognitive Assessment (MOCA), the Hospital Anxiety and Depression Scale (HADS) and the Apathy Evaluation Scale (AES)^[1]. Among the 60 MD1 patients included, abnormal test results were obtained in 40% (MOCA), 72-77% (AES) and 18% (HADS)^[1]. These values did not differ between patients with high adherence (>5h/night on HMV) and patients with low adherence (<5h/night on HMV)^[1]. The HADS score was higher in the low adherence group than in the high adherence group^[1]. Older age and higher BMI were associated with high adherence to HMV^[1]. Cognitive impairment and apathy were found to be common in MD1 patients but had no effect on adherence^[1]. The study is excellent, but some points should be discussed.

The first point is that it has not been reported whether the degree of myopathy, muscle weakness and involvement of the limbs and respiratory muscles affected the ability to handle HMV. Therefore, we should know to what extent motor involvement in MD1 affected the ability to handle HMV itself and how it affected outcomes. Was the MIRS associated with adherence to HMV? How many individuals required assistance in handling HMV due to motor impairment? In this context, we should also know to what extent CTG length influenced the study results. Did patients with a long CTG repeat length have more cognitive impairment, depression and apathy than patients with a short length?

The second point is that the study was conducted during the pandemic, which means that at least some of the included patients may have had SARS-CoV-2 infection (SC2I) or SARS-CoV-2 vaccination (SC2V). Since SC2I/SC2V can have a strong influence on the effect of HMV, sleep and mood and thus on the study results, we should know how many of them had SC2I/SC2V. In how many patients was the breathing disorder attributed to SC2I?

The third point is that the current medication was not specified. As the effect of HMV is highly dependent on current medication, it would have been important to know how many were regularly taking tranquilizers, hypnotics, antidepressants, neuroleptics or antiepileptics.

The fourth point is that MD1 can also be associated with epilepsy^[2]. Therefore, we should know how many of them had epilepsy or nocturnal seizures and how this affected the study results. How many were regularly taking anti-seizure medication?

The fifth point is that the logistic regression showed that a high BMI was associated with higher adherence. How can this result be explained? One would expect that a high BMI would tend to be associated with low adherence.

The sixth point is that at least some patients had mild cognitive impairment (MCI), which raises the question of how these patients were able to give informed consent. Were these patients legally allowed to give informed consent? Were they aware of the type of tests being carried out and the implications of the results?

In summary, it can be said that this interesting study has limitations that relativize the results and their interpretation. Addressing these limitations could strengthen the conclusions and support the study's message. Adherence to home ventilation in MD1 may depend not only on cognition, mood and apathy, but on a variety of other factors that should be considered when evaluating the influence of cognition and mood on HMV.

Declarations

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Author contribution: JF was responsible for the design and conception, discussed available data with coauthors, wrote the first draft, and gave final approval.

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