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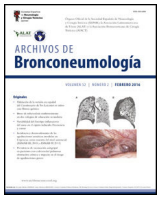

Letter to the Director

Effective IMT Starts With a Feasible Training Protocol and Identification of Clinical Relevant Outcome Parameters

To the Director,

We kindly appreciate the reply of colleague Finsterer.[1] The considerations of Finsterer are legitimate and appropriate in DM1, which is probably the most complex and clinically variable progressive multi system disorder. Therefore, a lot of factors (like CTG repeat, age, vital capacity, cardiovascular risks and need of walking aids) could influence survival and therefore need correction in outcomes.[2]

However, we disagree with the first point made by Finsterer. The clinical case we presented showed promising effects on hypercapnia after 12 weeks of IMT and as low evidence is inherent on clinical cases we concluded in the final part of the report that further investigations are needed.

The phenotype of the patient is described as a classical onset and he uses a walker outside. There were no other reasons for nocturnal hypercapnia like thoracic cage deformities or COPD in this non-smoking patient. Clinically, there have been no signs of SARS-CoV-2 infection before he presented with worsening of the clinical course. Besides atrial fibrillation in the past there were at this moment no cardiac comorbidities (he preventively visits the cardiologist once a year).

The power of a case report is to present (surprising) novel clinical findings to a larger public. In this way it can serve as a podium for ideas, which can be further explored in larger studies. We agree with Finsterer that interfering factors needs to be addressed before a valid general conclusion of IMT effects can be made. However, before an IMT multi-centre randomized controlled trial in DM1 can be designed, a lot of other research questions needs to be answered: First, it is not known which training schedule is optimal, regarding the known training parameters of frequency, intensity, time and type (FITT principle). As the most common reported complaint in DM1 patients is fatigue, it is essential to have an efficient training schedule, requiring a short time effort of the patient. Second, training adherence is an important factor which influences outcomes. For example, an earlier DM1 study reported a high dropout of 47% during a 12 week aerobic training on a cycle ergometer.[3] Cognitive and behavioural aspects like apathy and motivation deficits are potential serious counterworking factors which easily reduces adherence and therefore negatively influence the outcomes. Probably, a (digital) buddy can be helpful. Third, the optimal timing when to start IMT in DM1 patients is not known. An early start of IMT can probably delay respiratory muscle weakness, but when patients do not feel improvement by training the motivation could decrease and dropout rates will increase. Finally, in concordance with the suggestions of Finsterer, we think that the parameters to measure outcome effects needs to be further assessed, together with their minimal clinically important differences of these values. Besides physiological outcomes like spirometry, PImax and transcutaneous gas-exchange monitoring, also diaphragm specific values can be used like diaphragm ultrasound, phrenic nerve stimulation or electromyography. In addition, effects on symptoms and quality of life need specific attention as well.

In conclusion, we agree with Finsterer that that this study is not providing evidence that IMT generally leads to better outcomes in patients with DM1. Precisely characterized patients in methodological well designed studies are needed to draw conclusions on IMT in DM1 patients.

Conflict of interest

None.

References

1. Finsterer J. Whether inspiratory muscle training really makes sense in myotonic dystrophy requires appropriately designed trials. Arch Bronconeumol. 2024, http://dx.doi.org/10.1016/j.arbres.2023.12.018.
2. Wahbi K, Porcher R, Laforet P, Fayssoil A, Becane HM, Lazarus A, et al. Development and validation of a new scoring system to predict survival in patients with myotonic dystrophy type 1. JAMA Neurol. 2018;75:573–81.
3. Orngreen MC, Olsen DB, Vissing J. Aerobic training in patients with myotonic dystrophy type 1. Ann Neurol. 2005;57:754–7.

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