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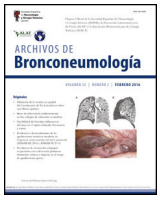

Letter to the Director

Whether Inspiratory Muscle Training Really Makes Sense in Myotonic Dystrophy Requires Appropriately Designed Trials

To the Director,

We read with interest Seijger et al.'s article about a 54 year-old male with myotonic dystrophy type-1 (MD1), which clinically manifested with progressive fatigue and dyspnea.[1] Because of respiratory dysfunction due to weakness of the respiratory muscles, the patient underwent inspiratory muscle training (IMT) using the electronic POWERbreathe KHP2 device and wearing a nose clip.[1] He conducted two sessions per day, five days a week for 12 weeks.[1] After a period of 12 weeks PImax improved by 176% to 47.9 cmH_2O and inspiratory endurance capacity increased from 187s on a resistance of 7 cmH_2O to 305 s on 19 cmH_2O.[1] It was concluded that MD1 responds positively to IMT, especially with regard to hypercapnia. The study is impressive, but some points should be discussed.

The major limitation of the study is that general conclusions were drawn from a single case report. Before concluding that IMT has a beneficial effect on respiratory functions in MD1, studies using an international, multicentre, double-blind placebo-controlled design are needed to assess the question whether IMT is truly beneficial for MD1 patients. When designing such a study, it is imperative that the studied cohorts are homogeneous not only in terms of demographic parameters but also in terms of MD1 genetic background, disease duration, comorbidities, and comedication. Only if these requirements are met can a possible positive effect of IMT be assessed. Otherwise, general conclusions about the effect of IMT remain uncertain.

A second limitation is that the number of CTG-repeats for the index patient was not reported. Knowledge of abnormal expansion is crucial because the phenotype is highly dependent on repeat length. In general, the longer the repeat-size, the more severe the phenotype.

A third limitation is that respiratory muscle weakness has not been documented. There was neither phrenic nerve stimulation nor electromyography of the diaphragm. Only an indirect measurement was carried out using the forced vital capacity and the maximum respiratory muscle strength.

A fourth limitation is that it was not reported whether the patient had thoracic spine deformities (scoliosis, hyperkyphosis, stiff spine), obstructive airway disease, or restrictive lung dysfunction. It was also not stated, whether the patient was a smoker or not.

Another limitation is that SARS-CoV-2 infection has not been ruled out. Since the patient was reported during the pandemic, it is imperative to know whether respiratory dysfunction was due to the viral infection or other causes.

Since respiratory function depends not only on the strength of the respiratory muscles but also strongly on cerebral and cardiac function, it is important to know whether the index patient had cerebral or cardiac involvement in MD1. The effect of IMT can also depend heavily on whether organs other than the respiratory muscles were affected or not.

In summary, the excellent study has limitations that should be addressed before drawing final conclusions. Clarifying the weaknesses would strengthen the conclusions and could improve the study. Whether IMT actually improves respiratory muscle strength in MD1 requires the use of appropriate study designs.

Authors' contributions

JF was responsible for the design and conception, discussed available data with coauthors, wrote the first draft, and gave final approval. SM: contributed to literature search, discussion, correction, and final approval.

Ethical approval

Not applicable.

Consent to participation

Not applicable.

Consent for publication

Not applicable.

Availability of data and material

All data are available from the corresponding author.

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Conflict of interests

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Reference

1. Seijger CGW, Nieuwenhuis J, van Engelen BGM, Wijkstra PJ. Benefits of inspiratory muscle training in myotonic dystrophy: a case report. Arch. Bronconeumol. 2023, http://dx.doi.org/10.1016/j.arbres.2023.11.008. S0300-2896(23)00376-9. English, Spanish.

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