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Work-Up for Diaphragmatic Dysfunction in Neuromuscular Disorders Requires Standardized Diagnostic Criteria

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Commentary:

In a recent article, Türk et al. reported about a retrospective study of 30 patients with respiratory insufficiency due to diaphragmatic dysfunction having undergone nerve conduction studies of the phrenic nerve or needle electromyography (EMG) of the diaphragm [1]. We have the following comments/concerns.

Inclusion criteria are contradictory. Since the study had a retrospective design, the authors knew the diagnosis of all patients, except those with unknown cause, (n=8) in advance. According to the inclusion criteria, those with a known cause of phrenic nerve lesion (n=22) should have been excluded.

It is not comprehensible why the reference limit for distal latency of the phrenic nerve was 8.5ms for all patients, regardless of age, sex, and height [2]. It is easily comprehensible that tall probands will have a longer distal latency than short probands. Since men are usually taller than females, it is also comprehensible that the distal latency of the phrenic nerve will be longer in males as compared to females. Furthermore, it is comprehensible that the phrenic nerve distal latency will increase and that nerve conduction will decrease with age. Inspiration/expiration and affection of the upper motoneuron may additionally influence phrenic nerve conduction [3].

According to table 1, four patients had undergone needle-EMG of the diaphragm, but only in one patient the results of this investigation were provided [1]. Which were the needle-EMG findings in the other three patients? Since 28/30 patients had neuropathy of the phrenic nerve, it is quite likely that these three patients had a neurogenic pattern with enlarged motor unit action potentials and a thinned interference pattern.

Pompe disease may not only manifest with myopathy but also with neuropathy [4]. In the one patient with Pompe disease and a myogenic EMG, were nerve conduction studies of the phrenic nerve or other nerves normal?

Isolated neuropathy of the phrenic nerve is rather rare [5]. Did the 28 patients with neuropathy of the phrenic nerve also present with neuropathy of the peripheral nerves supplying the limb muscles or the facial muscles?

Five patients were reported to have experienced complete recovery of diaphragmatic dysfunction [1]. Two of these patients had a neuropathy of the phrenic nerve of unknown cause [1]. How do the authors explain that two patients recovered without receiving any specific diagnosis or treatment for diaphragmatic dysfunction? The three others that fully recovered responded favorably to treatment for myasthenia, borelliosis, or CIDP [1].

Concerning those with idiopathic phrenic nerve neuropathy, was the family history taken and positive for

neuropathy and did relatives of these patients undergo clinical exam, nerve conduction studies, and if indicated, needle EMG?

In conclusion, this retrospective study could be more meaningful if inclusion / exclusion criteria would be more strict, if age-, sex-, and height dependent reference limits would have been applied, if EMG results of all patients would have been reported, if nerve conduction studies of nerves other than the phrenic nerve would have been provided, and if full recovery of idiopathic phrenic nerve neuropathy would have been discussed.

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