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Fatigue is part of the burden of neuromuscular diseases

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Patients with neuromuscular diseases ask relatively simple questions. They want a timely diagnosis, and information about prognosis and effects of disease on their personal functional status. With respect to treatment, patients long for a cure but most understand that they will have to reconcile to rehabilitation or even palliative treatment.

With respect to diagnosis, neurologists are trained to detect impairments, neurological signs and symptoms such as motor weakness using the MRC scale, or a Babinski sign. Also, though usually not in a structured way neurologists try to evaluate disability, such as not being able to go up the stairs or cycling. Functional status may be measured with the sickness impact profile (SIP) or the modified Rankin scale. Finally effects of disease on participation, for example not being able to work or taking part in sports, and on perceived health will be estimated. Measures of disability, participation and perceived health are increasingly being used as out-

come measures in clinical trials [1].

Fatigue may be another underestimated symptom of neuromuscular disease [2]. That can be explained by ignorance as most patients with the exception of patients with myasthenia gravis or myotonic dystrophy do not spontaneously complain of fatigue. Many severely handicapped neuromuscular patients keep trying to participate in social life and work. In contrast, patients with the chronic fatigue syndrome have no or little objective signs and maximal decrease of participation and perceived health [2]. Yet, if fatigue would be a common additional feature of neuromuscular diseases, neurologists must be aware in order to better comprehend their patients and to develop new treatment strategies.

This issue of the *Journal of Neurology* contains two papers on fatigue in hereditary neuromuscular diseases. Dr. Hagemans did a cross-sectional study using a questionnaire with the Fatigue Severity Scale in 225 patients with late-onset Pompe disease [3]. About 67% had severe fatigue. Possible confounding factors were wheelchair dependency and respiratory support in about half of the patients, but 59% of patients without these aids also had severe fatigue. Previously this group investigated quality of life in late-onset Pompe patients using the SF-36 [4]. Interestingly, they found that these patients score only slightly lower than the general population on the mental health domains, making depression or decreased vitality as explanations for fatigue unlikely [3, 4]. The second paper shows that fatigue and pain can be relevant symptoms in facioscapulohumeral muscular dystrophy (FSHD) [5]. Both groups suggest that fatigue is probably

underreported in these slowly progressive neuromuscular diseases.

Some caution with the role of confounding factors seems justified. The question whether patients with late onset Pompe disease and no respiratory aid have respiratory insufficiency that explains fatigue, can only be answered by actually measuring pulmonary function. In FSHD impaired shoulder function that is difficult to quantify, will affect quality of life and could explain fatigue.

Nevertheless, the notion that fatigue may be part of the phenotype of neuromuscular disease is important. To my opinion, the wear and tear from having to live a life long with the consequences of a neuromuscular disease may be one explanation for fatigue. Severe fatigue will affect participation and perceived health. Neurologists must learn to measure fatigue during history taking. Neurologists can also develop interventions. For instance, it has recently been demonstrated that simple bicycle exercise training may help to decrease fatigue in patients following Guillain-Barré syndrome [6].

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