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Added value of electromyography in the diagnosis of myopathy: A consensus exercise

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HIGHLIGHTS

- In a European multicentre study, electrodiagnostic medicine (EDX) results increased the consensus probability of myopathy.
- Adding EDX results increased the diagnostic certainty of myopathy in one third of 195 patients.
- The highest diagnostic impact of EDX was seen in myopathies of unknown aetiology.

ABSTRACT

Objective: Currently, neurologists may primarily rely on blood biomarkers, muscle biopsy, MRI, and genetics in the diagnostic work-up of suspected myopathy. Using expert consensus as diagnostic reference standard, this study addressed the added value of electrodiagnostic medicine (EDX) in diagnosis of myopathies.
Methods: One hundred ninety-four EDX evaluations of patients with a peer-review consensus diagnosis of myopathy were collected by seven European centres. Each patient was given three different consensus diagnoses: (1) the EDX diagnosis solely based on EDX results, (2) the pure clinical diagnosis based on all available information except EDX results, and (3) the final diagnosis including EDX and all additional information. The myopathies were grouped as muscular dystrophy (45), inflammatory myopathy (46), other aetiology (36) or unknown aetiology (67).
Results: Higher diagnostic probabilities for myopathy were seen in the final diagnosis compared to the pure clinical diagnosis ($p < 0.001$). Adding EDX information increased the diagnostic probability of myopathy in 67 patients (34.4%). The greatest increase was seen for myopathies of unknown aetiology.
Conclusions: EDX has a major impact in the diagnosis of myopathies of unknown aetiology. In genetically or biopsy proven myopathies, EDX generally supports the diagnosis.
Significance: EDX is still a useful tool in the diagnostic work-up of most patients with suspected myopathy.

1. Introduction

It is widely accepted that the electrodiagnostic (EDX) evaluation of muscle disorders is useful in detecting myopathic changes in the muscles and to exclude differential diagnoses (Fuglsang-Frederiksen, 2006; Liguori et al., 1997; Schoser, 2016). However,

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neurologists may not refer patients with suspected myopathy to EDX, but rely on results from blood biomarkers (as creatine kinase), muscle biopsy, genetic testing, and magnetic resonance imaging (MRI) (Anthonisen et al., 2005; Paganoni and Amato, 2013).

Using electromyography (EMG), several muscles in different limbs can be examined with an instantly available result, and myopathic changes may also be detected in clinically unaffected muscles. The characteristic EMG findings of myopathy are polyphasic motor unit potentials (MUPs) of short duration. and a full interference pattern of low amplitude (Fuglsang-Frederiksen, 2006). As the EMG needle electrode has a relatively small uptake area, it is recommended to record from multiple sites evenly distributed over the muscle. At least 20 different MUPs or 10 turns-amplitude analyses of the interference pattern are considered to be representative of the pathophysiological state of the muscle (Fuglsang-Frederiksen and Månsson, 1975; Rosenfalck and Rosenfalck, 1975) While muscle biopsy and genetic testing are also valuable tools in the diagnosis of myopathy, especially in the search of a specified aetiology, these techniques cannot determine the distribution of muscles affected. Muscle MRI, including whole body-MRI (Elessawy et al., 2016), seems to be a promising evaluation in the diagnosis of myositis, but it is an advanced and costly procedure still not routinely used in many places (Maurer and Walker, 2015; Fischer et al., 2016).

Several reports have documented that a myopathic EMG is predictive of finding myopathy or specific myopathy on muscle tissue biopsy (Buchthal and Kamieniecka, 1982; Cardy and Potter, 2007; Shaibani et al., 2015). However, there are not many reports comparing clinical findings and EMG. A study of 37 patients with genetically proven facioscapulohumeral dystrophy (FSHD) showed no correlation between electrophysiological findings and clinical features (Dorobek et al., 2013), while another study on muscle biopsies showed that abnormal EMG had no impact on the outcome in 104 patients with asymptomatic or minimally symptomatic hyperCKemia (Fernandez et al., 2006).

Using expert consensus diagnosis as the diagnostic reference standard, this study aimed at addressing the added value of EMG in the diagnosis of myopathy. The study was carried out in the European multi-centre project ESTEEM (European standardised Telematic tool to Evaluate electrodiagnostic Methods), an ongoing collaboration since 1992, where eight physicians from seven European neurophysiological centres collect samples of their patient examinations for peer review (Vingtoft et al., 1995).

2. Methods

2.1. Data collection

One hundred and ninety-five examinations were collected by eight physicians from seven European centres as sets where each physician was asked to submit 10 or 12 consecutively performed studies of patients diagnosed with myopathy by the examining centre. The examinations were performed in the period 1995–2010, with the majority (190) done after year 2000. The patients were evaluated using the diagnostic strategy and examination techniques routinely used in the laboratory doing the examination, i.e. some patients were examined with quantitative methods and others with qualitative. In 145 of the patients at least one quantitative technique (quantitative MUP analysis, turns-amplitude analysis of the interference pattern, or macro-EMG) was used, and an average of 2.9 muscles per patient was examined. The remaining 50 patients were examined with qualitative methods solely, with an average of 4.5 muscles examined per patient. The diagnosis of myopathy was given by the examiner submitting the examination to the database and was based on EDX and all available clinical information, including muscle biopsy, genetic tests etc. There was no minimum standard for the extent of the evaluation of the patients, but a detailed patient history, clinical findings, and EDX data were obtained in all. The eight physicians each contributed with 3, 16, 18, 18, 33, 34, 35, and 38 examinations.

2.1.1. Standardised data format

Data were entered into a standardised data structure implemented in PC program (Johnsen et al., 1994), either manually or by automatic transfer from Keypoint (Dantec, Skovlunde, Denmark) EMG equipment. Only the derived parameters and not the digital recordings were included. Briefly, the standardised data structure comprises three levels: (a) General data including patient demographics, history, clinical evaluation, and paraclinical tests; (b) EDX data represented as the measured value with local reference values. Muscle force as 0, 1, 2, 3, 4−, 4, 4+, 5 according the Medical Research Council (MRC) scale with 0 indicating no movement and 5 indicating normal muscle strength (Medical Research Council, 1976), atrophy as 0, +, ++, +++ indicating no atrophy and mild, moderate, or severe atrophy, and patient cooperation were additionally stated; (c) Inferred data as *test conclusions* (Normal, Neurogenic, Myopathic, Myotonic, Unspecific, Impossible) for muscles and nerve segments, and one or more *diagnoses* with a categorical probability (definite, probable, possible, excluded) and a supplementing visual analogue scale (VAS) from 0 to 100, where 0 is indicating no possibility for the diagnosis and 100 is indicating definite diagnosis. For at detailed description of the standardised data format see (Johnsen et al., 1994).

2.1.2. Peer review

Each examination was given two different blinded diagnoses by consensus (Fig. 1):

- The pure clinical consensus diagnosis based on all available information except EDX results.
- The final consensus diagnosis based on all available information, including EDX.

For peer review all information except the measured EDX data was removed and the examination was re-evaluated independently by all physicians for their blinded interpretation. For the discussion of each patient at the consensus meetings, the physicians' individual diagnoses were displayed on a screen as a starting point for an open discussion among the group with the aim of obtaining consensus on the diagnosis. First a consensus diagnosis based solely on EDX data was established. After this clinical and additional laboratory information was revealed to the consensus group for the final consensus diagnosis. For this project a second diagnosis, the pure clinical consensus diagnosis, was obtained by reviewing all clinical and laboratory information, except EDX data. This step-process was done at separate sessions on ten different consensus meetings from 2006 to 2012, with the investigators blinded to the former diagnoses (Fig. 1).

2.2. Patients

Initially records from 201 patients were included in the database. Four of these did not achieve consensus due to conflicting data and two were discarded because they were not diagnostic studies, i.e. one was a follow-up study and one was an EDX testing for carpal tunnel syndrome in a patient with known myopathy. Thus 195 cases with a final consensus diagnosis of definite (139), probable (35), possible (20), or excluded (1) myopathy were left for analysis.