The role of quantitative electromyography in inclusion body myositis

Thomas H Brannagan, Arthur P Hays, Dale J Lange, Werner Trojaborg

Abstract

Objective and methods—Inclusion body myositis is said to have both myopathic and neurogenic features on electrophysiological tests. Twenty one studies from 20 patients with biopsy defined inclusion body myositis, 13 of whom had quantitative electromyography (qEMG), were reviewed to determine if this technique added diagnostic specificity (one patient had both needle EMG and a later study with qEMG before muscle biopsy).

Results—Excessive numbers of polyphasic motor unit potentials (MUPs) (>12% per muscle) were seen in 11 of the 13 patients. In 10 of 13 patients, mean MUP duration was abnormally reduced (26% to 48%). In three patients, mean MUP duration was abnormally reduced only after polyphasic MUPs were excluded. In all 13 patients, the simple MUP duration was reduced. Myopathy was unequivocally diagnosed in all 13 studies that included qEMG; of the remaining eight patients, the conclusions of the electrophysiological studies without qEMG was myopathy (one), neurogenic (four) or non-diagnostic (three).

Conclusions—There is no evidence of a neurogenic component in inclusion body myositis if qEMG is used. Quantitative EMG is often necessary to make an electrophysiological diagnosis of a myogenic disorder in patients with inclusion body myositis.

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Keywords: inclusion body myositis; quantitative electromyography; motor neuron disease; fasciculations; motor unit potentials; myopathy

Inclusion body myositis is a chronic inflammatory myopathy, clinically characterised by both proximal and distal limb weakness and a poor response to steroid treatment. Pathologically, the condition is defined by the presence of rimmed vacuoles, filamentous inclusions, and intracellular amyloid deposits. Inclusion body myositis is said to have both myopathic and neurogenic features on electrophysiological tests as summarised in tables 1 and 2. Some patients were referred with the diagnosis of ALS and although they did not have definite upper motor neuron signs such as a Babinski’s sign or clonus, reflexes were preserved in weak and wasted limbs. All biopsies showed rimmed vacuoles and endomysial fibrosis. Inflammatory cells and groups of atrophic fibres were seen in most cases. Target fibres or fibre type grouping were not seen in any biopsy.

Sensory and motor conduction velocities were normal in all but two patients. Quantitative EMG (qEMG) was performed in 13 patients. Excessive numbers of polyphasic MUPs were seen in 11 patients (15% to 60%). In 10 of these patients, the mean duration of all
MUPs was abnormally reduced by 26% to 48%. In three patients, the mean duration of all MUPs was normal, but it was abnormally reduced if only simple MUPs were considered (34% to 46% below the normal mean; table 3).

Table 2 Electrodiagnostic results from 20 patients with inclusion body myositis

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Referring diagnosis</th>
<th>EMG diagnosis</th>
<th>MNC</th>
<th>SNC</th>
<th>Muscle</th>
<th>MUPs (n)</th>
<th>Prolonged MUPs (%)</th>
<th>Duration (µs) (normal)</th>
<th>Duration of simple MUPs (µs) (normal)</th>
<th>Amplitude (µV)</th>
<th>Spontaneous activity</th>
<th>Recruitment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>IBM</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Tib ant</td>
<td>20 15</td>
<td>9.5 (62)</td>
<td>10.2 (84)</td>
<td>11.3 (73)</td>
<td>11.7 (114)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>2</td>
<td>ALS</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Tib ant</td>
<td>20 30</td>
<td>9.5 (62)</td>
<td>8.8 (57)</td>
<td>10.0 (84)</td>
<td>10.8 (84)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>3</td>
<td>MND</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>24 0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>4</td>
<td>ALS + N</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>V lat</td>
<td>17 12</td>
<td>11.9 (96)</td>
<td>11.4 (93)</td>
<td>11.4 (93)</td>
<td>11.4 (93)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>5</td>
<td>PM</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>20 10</td>
<td>11.3 (73)</td>
<td>10.6 (69)</td>
<td>11.3 (73)</td>
<td>11.3 (73)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>6</td>
<td>Myopathy</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>24 0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>7</td>
<td>Myopathy</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>20 50</td>
<td>11.3 (73)</td>
<td>10.6 (69)</td>
<td>11.3 (73)</td>
<td>11.3 (73)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>8</td>
<td>PM</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>20 50</td>
<td>11.3 (73)</td>
<td>10.6 (69)</td>
<td>11.3 (73)</td>
<td>11.3 (73)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>9</td>
<td>Myopathy</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>20 50</td>
<td>11.3 (73)</td>
<td>10.6 (69)</td>
<td>11.3 (73)</td>
<td>11.3 (73)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>10</td>
<td>SMA + myopathy</td>
<td>MND</td>
<td>4a</td>
<td>N</td>
<td>CV(m, u, s)</td>
<td>Long</td>
<td>1-2+fibs/</td>
<td>1-2+fascies/CRDS</td>
<td>SMA myopathy</td>
<td>Not done</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>11</td>
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<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>V lat</td>
<td>24 0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
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</tr>
<tr>
<td>12</td>
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<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>V lat</td>
<td>24 0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>13</td>
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<td>MND</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>20 50</td>
<td>11.3 (73)</td>
<td>10.6 (69)</td>
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<td>11.3 (73)</td>
<td>1-2+fibs/</td>
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<td>N</td>
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<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>1-2+fibs/</td>
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<td>N</td>
<td>N</td>
<td>V lat</td>
<td>24 0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>16</td>
<td>Myopathy</td>
<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>Biceps</td>
<td>20 50</td>
<td>11.3 (73)</td>
<td>10.6 (69)</td>
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</tr>
<tr>
<td>17</td>
<td>PM</td>
<td>Non-diag</td>
<td>N</td>
<td>N</td>
<td>V lat</td>
<td>24 0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>18</td>
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<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>V lat</td>
<td>24 0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>1-2+fibs/</td>
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</tr>
<tr>
<td>19</td>
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<td>Myopathy</td>
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<td>N</td>
<td>V lat</td>
<td>24 0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
<tr>
<td>20</td>
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<td>Myopathy</td>
<td>N</td>
<td>N</td>
<td>V lat</td>
<td>24 0</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>10.2 (84)</td>
<td>1-2+fibs/</td>
<td>N</td>
</tr>
</tbody>
</table>

a=amplitude; ab=absent; ALS=amyotrophic lateral sclerosis; CRDS=complex repetitive discharges; fascics=fasciculations; fibs=fibrillations and positive sharp waves; CV=conduction velocity; I=increased; IBM=inclusion body myositis; m=median nerve; MND=motor neuron disease; N=normal; np=neuropathy; p=peroneal; PM=polyneuropathy; R=reduced; r=sural; SMA=spinal muscular atrophy; SMN=sensornotor myopathy; t=tibial nerve; u=ulnar nerve; t=full recruitment on maximal effort; t=full recruitment on submaximal effort; f=fasciculations; f=fasciculations/myopathy; CRDS=rare fibs/CRDS;

Therefore a myogenic disorder was diagnosed in all 13 studies in which qEMG was done. Of the remaining eight studies, when formal analysis of MUP form was not undertaken the findings were interpreted as neurogenic (four), including three as motor neuron disease or non-diagnostic (three). In only one of eight patients was a myogenic disorder correctly diagnosed (table 3). In one patient (10), the EMG was interpreted as showing motor neuron disease, without using quantitative EMG; when repeated five months later, qEMG disclosed short duration MUPs compatible with myopathy.

Eleven of the 20 patients with inclusion body myositis were referred to us with the diagnosis of motor neuron disease or neuropathy. A myogenic disorder was diagnosed in all five in whom qEMG was done. In only one of the six patients tested without qEMG was the myogenic nature of the illness identified using physiological methods.

Two patients (6 and 13), with EMG evidence of fasciculations, had biopsies of the motor nerve to the gracilis muscle to consider the possible coexistence of a motor neuropathy.
Reduced recruitment is a qualitative measurement, but is also non-specific because it may be seen in myopathies when loss of fibres is so extensive that whole motor units drop out. The presence of long duration polyphasic MUPs seen in myopathies correlate with regenerating fibres. Patients with neuropathic, myopathic, or normal EMG may have individual MUPs that could be considered “neuropathic” (>16 ms) or “myopathic” (<6 ms). Only the mean duration of a sufficient number of simple MUPs correlates with the presence of a neuropathy or myopathy. In one report of eight patients with inclusion body myositis, no patients showed neuropathic changes using quantitative EMG. Studies using single fibre EMG and macro-EMG in patients with inclusion body myositis also found no evidence of a neuropathic process or reinnervation as a cause of the long duration polyphasic motor units seen.

Quantitative EMG is necessary to accurately characterise the myogenic motor unit morphology in some cases of inclusion body myositis. Without qEMG, in the presence of spontaneous activity (fibrillations, positive sharp waves, and fasciculations), long duration polyphasic MUPs, and reduced recruitment, a pure motor neurogenic disorder, such as motor neuron disease, may be erroneously suspected.

We thank Dr Lewis P Rowland for helpful comments on the manuscript. Presented in part in abstract form at the meeting of the American Neurology Association, Washington, DC, 22-25 October 1995.

The motor nerve was morphologically normal in both and immunohistochemistry for nerve growth factor receptor was absent in one (6) and in the other there was only trace expression (13).

Discussion

Claims of a neuropathic component in inclusion body myositis have been based on features from muscle biopsy and electrophysiological testing. Group atrophy and angular fibres are seen which are suggestive although not specific for a neurogenic disorder. Fibre type grouping, which is specific for a neurogenic disorder, is generally not seen unless there is a concomitant neuropathy as occurred in three of 48 patients in one series. Long duration or “neurogenic” MUPs on EMG are also cited as evidence of a neurogenic component in inclusion body myositis. In fact, long duration polyphasic MUPs are characteristically found in chronic myogenic disease. Fibrillations and positive sharp waves are often seen in myopathy and fasciculations are also occasionally seen in various muscle diseases. One report found fasciculations in 10% of patients with inclusion body myositis. Our findings, therefore, do not support the presence of a neurogenic component in inclusion body myositis if qEMG is employed.

Nerve conduction studies were normal except for mildly decreased evoked response amplitude in five of our patients, compatible with muscle atrophy, which may result from a myopathy. Sensory and motor conduction velocities were normal in all but two. Patient (15) had a coexisting neuropathy with sensory loss, absent ankle reflexes, and a raised CSF protein content. Nerve biopsy disclosed a severe neuropathy with rare onion bulbs. The second patient (10), a 68 year old woman with no sensory complaints, had mild slowing of the median, ulnar, and sural sensory nerves and the tibial motor nerve. The amplitude of the sensory responses was normal however, and low skin temperature was the implicated cause. The abnormalities on conduction studies in these two patients, therefore, can be explained by factors other than implicating neurogenic involvement of inclusion body myositis.

### Table 3 Electrophysiological findings

<table>
<thead>
<tr>
<th>Findings</th>
<th>n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nerve conduction:</td>
<td></td>
</tr>
<tr>
<td>Reduced CMAP</td>
<td>5 (25%)</td>
</tr>
<tr>
<td>Slow motor and sensory conduction</td>
<td>2 (10%)</td>
</tr>
<tr>
<td>EMG</td>
<td></td>
</tr>
<tr>
<td>Fasciculations</td>
<td>8 (40%)</td>
</tr>
<tr>
<td>Fibrillations/positive sharp waves</td>
<td>17 (85%)</td>
</tr>
<tr>
<td>Myokymia</td>
<td>1 (5%)</td>
</tr>
<tr>
<td>Complex repetitive discharges</td>
<td>4 (20%)</td>
</tr>
<tr>
<td>Quantitative EMG in 13 patients:</td>
<td></td>
</tr>
<tr>
<td>Increased polyphasics (15-60%)</td>
<td>11 (85%)</td>
</tr>
<tr>
<td>Short mean duration - all MUPs (26-48%)</td>
<td>10 (77%)</td>
</tr>
<tr>
<td>Short mean duration - simple MUPs (34-46%)</td>
<td>13 (100%)</td>
</tr>
<tr>
<td>Quantitative EMG and diagnosis of myopathy:</td>
<td></td>
</tr>
<tr>
<td>qEMG done</td>
<td>13</td>
</tr>
<tr>
<td>qEMG not done</td>
<td>1</td>
</tr>
<tr>
<td>Diagnosis of myopathy</td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>No diagnosis of myopathy</td>
</tr>
<tr>
<td></td>
<td>7</td>
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</tbody>
</table>


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