Recommendations (1)

1. Appointment time allocated for a patient referred with a provisional diagnosis of MND should be more than 30 minutes, preferably 60 minutes.

2. The body should be considered in four anatomic regions (bulbar, cervical, thoracic and lumbosacral) in accordance with the El Escorial Criteria.

3. An affected region is defined as denervation in at least two muscles supplied by two different peripheral nerves and nerve roots. The bulbar region must include examination of the tongue.

4. Signs of denervation are either acute and/or chronic neurogenic changes including fasciculations, fibrillations, high amplitude, long duration, stable/unstable polyphasic MUPs and reduced recruitment. MUP Firing rate is usually higher than 10 Hz unless there is a significant UMN component, the firing rate may be lower than 10 Hz.

Recommendations (2)

5. When a patient is referred with suspected MND, needle EMG examination should include muscle sampling in four body regions on one side of the body.

6. Recommended muscles in the bulbar region (Tongue, Trapezius/SM and Orb Oris), thoracic region (Middle thoracic paraspinals and rectus abdominis), cervical (first dorsal interoseous, EDC, biceps or deltoid), lumbosacral (Tibialis Anterior, Vastus Lateralis and Tensor Fascia Latae).

7. Examination of muscles on two sides is optional. However, it may be useful when patient presents with asymmetrical weakness to detect early denervation/reinnervation changes in clinically intact muscles.

8. Quantitative EMG analysis is optional but it may be a useful addition if it is difficult to ascertain changes in motor unit morphology by visual analysis.

Recommendations (3)

9. It is sufficient to examine two regions (cervical and lumbosacral) when patient presents with fasciculations and no denervation is found on needle EMG.

10. A definite electrodiagnosis of MND requires demonstration of denervation in four body regions

(or three body regions including bulbar, cervical and lumbosacral) with lack of conduction block and normal sensory potentials on nerve conduction testing.

11. Summary of findings /conclusion must clearly indicate the denervated region (s).

12. Avoid using suspected, possible or probable MND in the conclusion as these are clinical terms originally used in the El Escorial Criteria and their use is later discouraged.

Recommendations (4)

13. If denervation is not found in four body regions (particularly the bulbar muscles) the differential diagnosis should be wide open to include acquired or hereditary pathology of the motor neurons and/or motor axons innervating the affected muscles/body region (s).

14. A follow-up study in a few months is recommended when denervation is confined to one or two regions to check on progression. The revised El Escorial criteria emphasised the importance of progression to confirm diagnosis of MND.

15. Nerve conduction studies should be performed routinely in all patients referred with suspected MND/pure motor syndrome to exclude potentially treatable disorders (peripheral neuropathies, plexopathies or focal peripheral nerve lesions).

16. Segmental motor testing is performed on request and when patient presents with distal asymmetric upper limb weakness in a peripheral nerve distribution or whenever you deem it necessary to exclude multifocal motor neuropathy with conduction block especially when median and/or ulnar F-responses are absent or infrequent.

Recommendations (5)

17. Repetitive nerve stimulation and single fibre EMG are performed in patients with bulbar symptoms when needle EMG does not show denervation to support MND.

18. Transcranial magnetic stimulation and motor unit estimation (MUNE) are not part of the routine investigations of patients with MND. However, it may be useful to document or detect subclinical UMN dysfunction when the clinical situation is doubtful.

19. Needle EMG can be performed in patients who are taking anticoagulant or antiplatelet medications especially if the INR is below 3.0 when the benefit of getting the diagnosis outweighs the risks. This should be discussed with the patient and documented in the report. Precautions include inserting the needle just below the fascia and applying direct pressure after removal of the needle when blood comes out.

20. Muscle ultrasound can be used as an additional tool to needle EMG in the detection of fasciculations.

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