Clinical Neurophysiology 130 (2019) 1405–1406

Contents lists available at ScienceDirect

Clinical Neurophysiology

journal homepage: www.elsevier.com/locate/clinph

Editorial

Hyperreflexia as an upper motor neuron sign in amyotrophic lateral sclerosis

See Article, pages 1455–1459

The Awaji criteria modified the acceptable criteria for lower motor neuron involvement (de Carvalho et al., 2008), and these changes have improved our ability to make an earlier diagnosis of motor neuron disease/amyotrophic lateral sclerosis (MND/ALS). However the report stated: "*The need for a reliable and sensitive physiological method for assessing upper motor neuron (UMN) disorder is evident by the controversy that often surrounds interpretation of apparently hyperactive tendon reflexes in wasted muscles in patients with a suspected diagnosis of ALS. Electrophysiological assessment of the upper motor neuron is therefore likely to be important but current methods require more general application before their limitations can be assessed*". The paper by Libonati et al. (2019) in this issue of *Clinical Neurophysiology* is a step forward in the neurophysiological identification of hyperreflexia.

It has been argued that H reflexes are under-utilized in diagnostic practice, particularly in suspected MND/ALS. This argument does not refer to the soleus H reflex, where it would be abnormal not to have a reflex response, but to muscles from which the H reflex cannot be demonstrated normally in subjects at rest (Burke, 2016). When H reflexes can be recorded at rest from muscles for which no reflex can normally be demonstrated, there is good evidence for hyperreflexia. In the context of possible ALS, this is an important finding. If the H reflex can be defined for one of these muscles and there is EMG evidence of chronic partial denervation in that same muscle, there is evidence for upper and lower motor neuron lesions involving a single muscle. This should raise suspicion of a degenerative disease, such as ALS, even though it would not satisfy traditional (but arbitrary) criteria based on the number of involved body regions. If we are to treat MND/ALS "before the horse has bolted", we need to start treatment before the disease process has spread beyond the initially involved region.

Heteronymous projections are quite weak in comparison to homonymous reflex projections (Pierrot-Deseilligny and Burke, 2012). Except in neonates, where there may be monosynaptic Ia excitation of antagonists as well as synergists (see Pierrot-Deseilligny and Burke, 2012), these projections normally require a voluntary contraction to be evident in surface EMG recordings (Miller et al., 1995; Meunier et al., 1996). In this respect, the finding of Libonati et al. (2019) that a heteronymous reflex can be demonstrated at rest in the temporalis muscle of most ALS patients (but not healthy subjects) on stimulation of the innervation of the masseter is significant. This finding suggests that the temporalis motoneurones were pathologically hyperreflexic in the ALS cohort.

In the report by Libonati et al. (2019), the jaw jerk ("mandibular reflex") was elicited in each patient, but the authors did not demonstrate a convincing H reflex in masseter. At first sight, this might seem surprising, but to define the H reflex would have required carefully graded stimuli, because the M wave would obscure the reflex potential, given the very short reflex pathway (latencies of ~2 ms and ~5–6 ms respectively). It is noted that the "*intensity of the stimulus was steadily increased until the best motor response (M) was elicited*", suggesting that a search for the masseter H reflex was not a specific focus of the study. In addition, the stimulus duration was 0.1 ms, a duration that does not favor stimulation of Ia afferents over α motor axons, given their different strength-duration properties (Pierrot-Deseilligny and Burke, 2012; Burke, 2016; see Lin et al., 2002). These issues are largely obviated by a feature of heteronymous reflex projections: unlike the H reflex, the "*heteronymous H reflex does not disappear when the stimulation intensity is increased*" because there is no collision between the reflex discharge in temporalis motor axons and the increasingly strong antidromic volley in masseter motor axons. Nevertheless it would be interesting to repeat these studies using a stimulus duration of 1 ms, much as was used by Godaux and Desmedt (1975), to see if both the masseter H reflex and the heteronymous temporalis reflex could be defined better.

Brisk heteronymous projections may appear superficially to provide the substrate for reflex irradiation. In patients and healthy subjects with brisk tendon jerks, percussion on a muscle tendon or bony prominence may produce reflex contractions in muscles not directly excited, even in some that are shortened by the percussion. This is clinical evidence of hyperreflexia, much as are the hyperactive heteronymous projections documented in the present paper by Libonati et al. (2019). The mechanisms underlying reflex irradiation have been addressed by Lance (Lance and de Gail, 1965; Lance, 1977), who showed that the jar produced by tendon percussion creates a vibration wave that spreads through bone to more distant muscles. The percussion-induced vibration wave excites muscle spindle endings in those muscles, thereby creating homonymous tendon jerks if the reflex circuits are sufficiently excitable. The mechanism is different in the present situation where temporalis motoneurones discharge in response to a

https://doi.org/10.1016/j.clinph.2019.05.015

heteronymous input in masseter afferents, not a volley in temporalis afferents.

Strictly speaking, the H reflex is a homonymous reflex, and it is a misnomer to refer to reflexes associated with heteronymous Ia afferent projections as H reflexes. This is a potential source of confusion because the authors sometimes refer to the H reflex without nominating that they mean the "heteronymous temporalis H reflex", not the H reflex of masseter. This paper raises other issues: *first*, was the heteronymous response in temporalis due to Ia afferents? *Secondly*, is the temporalis response monosynaptic? The short latency is compatible with fast-conducting afferents and monosynaptic transmission, but latency alone does not exclude one or two interneurons in the pathway. While it is likely that the responsible afferents were group Ia afferents and the pathways is likely to be monosynaptic, there are insufficient data in the report to be certain. Either way, these uncertainties do not negate the value that neurophysiological testing can provide in documenting evidence of upper motoneuron involvement in patients with suspected ALS.

Declaration of Competing Interest

None.

References

Burke D. Clinical uses of H reflexes of upper and lower limb muscles. Clin Neurophysiol Pract 2016;1:9–17.

de Carvalho M, Dengler R, Eisen A, England JD, Kaji R, Kimura J, et al. Electrodiagnostic criteria for diagnosis of ALS. Clin Neurophysiol 2008;119:497–503.

Godaux E, Desmedt JE. Human masseter muscle: H and tendon reflexes. Their paradoxical potentiation by muscle vibration. Arch Neurol 1975;32:229–34.

Lance JW. Mechanism of the inverted supinator reflex. J Neurol Neurosurg Psychiatry 1977;40:207.

Lance JW, de Gail P. Spread of phasic muscle reflexes in normal and spastic subjects. J Neurol Neurosurg Psychiatry 1965;28:328–34.

Libonati L, Barone TF, Ceccanti M, Cambieri C, Tartaglia G, Onesti E, et al. Heteronymous H reflex in temporal muscle as sign of hyperexcitability in ALS patients. Clin Neurophysiol 2019;130:1455–9.

Lin CS-Y, Chan JHL, Pierrot-Deseilligny E, Burke D. Excitability of human muscle afferents studied using threshold tracking of the H reflex. J Physiol (London) 2002;545:661–9.

Meunier S, Mogyoros I, Kiernan MC, Burke D. Effects of femoral nerve stimulation on the electromyogram and reflex excitability of tibialis anterior and soleus. Muscle Nerve 1996;19:1110–5.

Miller TA, Mogyoros I, Burke D. Homonymous and heteronymous monosynaptic reflexes in biceps brachii. Muscle Nerve 1995;18:585–92.

Pierrot-Deseilligny E, Burke D. The circuitry of the human spinal cord: spinal and corticospinal mechanisms of movement. New York: Cambridge University Press; 2012.

David Burke
Department of Neurology, Royal Prince Alfred Hospital, and the University of Sydney, Australia
E-mail address: david.burke@sydney.edu.au

Accepted 23 May 2019

Available online 30 May 2019