Quantification of Electromyographic Activity in Stiff Leg Syndrome-Adding to the Diagnostic Tool Box

Dear Editor,

Stiff person syndrome (SPS) is a rare disorder of the central nervous system, characterized by rigidity affecting the lumbar, trunk and limb muscles.[1,2] More recently, stiff limb syndrome (SLS) has been described as a distinct focal variant, with symptoms limited to one or both lower limbs. SLS is diagnosed based on clinical features, serum anti-glutamic acid decarboxylase (GAD65) antibody, and electrophysiology testing.[1,3] The classic electrophysiological finding described is continuous motor unit activity (MUA) on needle electromyography (EMG) in the stiff muscle that increases with contraction of the antagonist muscle

and decreases with treatment.[1,3] Root mean square (RMS) amplitude is a surface EMG processing technique that has been useful in quantification of abnormally increased MUA in certain disorders,[4,5] but has not been studied in SLS or SPS, to the best of our knowledge. We describe a case of SLS, in whom we used RMS amplitude to reliably detect and quantify the increased activity in the symptomatic limb. Written, informed consent was obtained from the patient and this does not contain any personal information that could lead to the identification of the patient. None of the authors have potential conflicts of interest to be disclosed.

Eight months prior to presentation, a 48-year-old lady developed constant stiffness in the right distal leg with painful spasms 10-15/day, each lasting 10-20 seconds. A few days prior to presentation, she also developed oscillopsia and binocular, diagonal double vision in lateral gaze. Neurological examination showed a right foot drag, increased tone in the right distal leg and normal deep tendon reflexes. On ocular examination, there were ocular cerebellar signs in the form of mild downbeat nystagmus worse on lateral and downgaze, an alternating skew deviation, and saccadic smooth pursuits. There were no systemic cerebellar signs and the rest of the neurological examination was normal. At the time of the clinical examination, no focal stiffness or spasms were apparent. Magnetic resonance imaging of the brain, orbits and spine with contrast were normal.

Nerve conduction studies, slow rate repetitive nerve stimulation test, F-wave latencies and routine needle electromyography of muscles of the lower limbs were normal. Though the patient complained of multiple spasms and constant stiffness, during the EMG examination we were able to record only one episode of increased MUA with a full interference pattern in the right tibialis anterior (TA) muscle, coinciding with a painful spasm.

We proceeded with surface EMG with RMS processing. The equipment used was Natus Ultrapro EMG, and a power spectrum software with fast Fourier transformation. The standard "turns -amplitude" program provided in the electromyograph was used to measure the RMS amplitude. Two sets of surface recording electrodes were placed on the right TA and gastrocnemius muscles, respectively. The active electrode was placed over the muscle belly, reference electrode 3 cm away, and the ground electrode was placed nearby. RMS amplitude was calculated over a 1-s epoch from each muscle (i) at rest, (ii) with active contraction (iii) passive activation of the antagonist muscle and (iv) during a spasm [Figure 1]. The RMS amplitude from the right TA was much higher than that of the unaffected side when the gastrocnemius was actively or passively contracting. The ratios of increase in activity over the right TA with passive and active movement of the gastrocnemius muscle, which we call RMS (p) agonist/antagonist and RMS (a) agonist/antagonist, were calculated and were high (15.1 and 14.3, respectively). The ratios from the unaffected limb were much lower at 2.3 and 5.0, respectively.

The patient tested positive for anti GAD65 antibodies and was diagnosed with GAD65 neurological autoimmunity with combined SLS and oculomotor cerebellar phenotype. She was started on treatment with oral diazepam 5 mg twice a day and experienced an improvement in symptoms. Re-evaluation in the EMG lab 1 week after initiation of treatment revealed that the RMS (p) agonist/antagonist and RMS (a) agonist/antagonist were decreased [Table 1]. We propose that RMS (p) and (a) agonist/antagonist ratios could be a useful marker for electrophysiologically diagnosing SLS, as well as assessing response to treatment, in conjunction with the clinical suspicion and serological findings. SLS has been mistaken for hysterical posturing in the past, since voluntary limb contraction can appear electromyographically identical. RMS amplitude could be a useful additional to the diagnostic tool kit as it detects and quantifies the abnormal increase in the muscle activity, even in the inter-spasm periods, that would be missed by needle EMG. Calculating a ratio is important, since a normal muscle at rest also demonstrates a positive, though small RMS amplitude value. Surface electrode recording makes it a comfortable test for the patient and the software required is already present or an easy addition to most labs. This novel technique, however, needs to be studied in a systematic manner with calculation of sensitivity and specificity of the test as it has been done for treatment of co-contractions in birth brachial plexus lesions.[5]

Table 1: Root mean square amplitudes from the tibialis anterior muscle and ratios derived in a patient with stiff leg syndrome at initial presentation and following treatment

	Initial presentation, prior to treatment		Following diazepam oral medication for 1 week	
	Right TA	Left TA	Right TA	Left TA
RMS amplitude (μV)				
At rest, no spasm	17	9	15	9
During clinical spasm	211	-	No clinical spasms	-
With passive antagonist muscle movement	258	21	40	29
With active antagonist muscle movement	243	45	107	67
RMS (p) agonist/antagonist	15.1	2.3	2.7	3.2
RMS (a) agonist/antagonist	14.3	5.0	7.1	7.4

TA - tibialis anterior, RMS - root mean square, μV - microvolt, RMS (p) agonist/antagonist-Ratio of increase in activity with passive movement, RMS (a) agonist/antagonist - Ratio of increase in activity with active movement

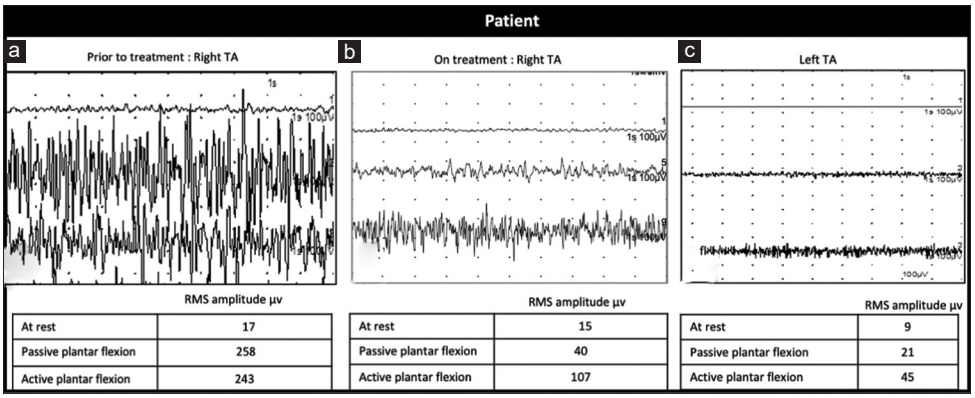

Figure 1: Surface electromyography (EMG) with root mean amplitude processing performed in the patient. (a) Increased activity in the right tibialis anterior (TA) with passive (middle) and active (bottom) plantar flexion of the right leg, compared with at rest (top). (b) Right TA activity following treatment with diazepam showing decreased amplitudes (middle and bottom) compared with pretreatment EMG. (c) Left TA activity at rest (top), with passive (middle) and active (bottom) plantar flexion of the left leg (unaffected limb)

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

Sasha A. Mansukhani, Satish V. Khadilkar[1], Madhubala Singla, Alika Sharma[2], Priyanka Chavan[2], Khushnuma A. Mansukhani[2]

Department of Ophthalmology, Mayo Clinic College of Medicine, Rochester, USA, Departments of [1]Neurology and [2]Clinical Neurophysiology, Bombay Hospital, Mumbai, Maharashtra, India

Address for correspondence: Dr. Khushnuma A. Mansukhani, Department of Clinical Neurophysiology (EMG) 2nd Floor MRC Building, Bombay Hospital, Mumbai - 400 020, Maharashtra, India.
E-mail: kilom@hotmail.com

References

1. Barker RA, Revesz T, Thom M, Marsden CD, Brown P. Review of 23 patients affected by the stiff man syndrome: Clinical subdivision into stiff trunk (man) syndrome, stiff limb syndrome, and progressive encephalomyelitis with rigidity. J Neurol Neurosurg Psychiatry 1998;65:633-40.
2. Rakocevic G, Alexopoulos H, Dalakas MC. Quantitative clinical and autoimmune assessments in stiff person syndrome: Evidence for a progressive disorder. BMC Neurol 2019;19:1.
3. Saiz A, Graus F, Valldeoriola F, Valls-Sole J, Tolosa E. Stiff-leg syndrome: A focal form of stiff-man syndrome. Ann Neurol 1998;43:400-3.
4. Farfan FD, Politti JC, Felice CJ. Evaluation of EMG processing techniques using information theory. Biomed Eng Online 2010;9:72.
5. Shin YB, Shin MJ, Chang JH, Cha YS, Ko HY. Effects of botulinum toxin on reducing the co-contraction of antagonists in birth brachial plexus palsy. Ann Rehabil Med 2014;38:127-31.

Submitted: 07-Mar-2021 **Revised:** 21-Apr-2021 **Accepted:** 21-May-2021
Published: 03-Nov-2021

DOI: 10.4103/aian.AIAN_193_21