Electrophysiological study of Writer’s cramp

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Abstract
Background: Writer’s cramp is a movement disorder characterized by prolonged involuntary muscle contractions, when the subject begins to write. In this study, We set out to assess various reflexes on both the affected and non-affected side in Writer’s cramp patients as well as in healthy controls to determine the pathophysiological basis of this condition.

Material and methods: A total 14 patients with Writer’s Cramp including 10 patients with simple Writer’s cramp and 4 patients with dystonia Writer’s cramp and 14 normal persons as control were evaluated with Electrophysiological tests performed on a Dantec EMG machine and unpaired t test was used for statistical analysis.

Results: The results of various reflexes studied were analyzed between the patients and the normal controls. There was no statistical difference in the latencies of Long Latency Reflex (LLR) 1,2 and 3 between affected and normal side as well as controls (P>0.05), with Blink Reflex (P>0.7304), Somatosensory evoked potential, Brain Stem auditory evoked potential(BAEP), Visual Evoked Potential (VEP).

Conclusions: A long latency reflex measurement is appears to be a reliable test to assess the functional impairment in writer’s cramp.

Key words: Writer’s cramp, ElectroPhysiological studies.

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1. Introduction

The word ‘Cramp” is used in many different ways in neurological practice. Cramp in the most common usage has been defined as “sudden involuntary and painful shortening of muscle attended by visible or palpable knotting of muscle, often with abnormal posture of the affected joint”. Writer’s cramp is a movement disorder characterized by prolonged involuntary muscle contractions causing twisting actions and abnormal postures of the hand, sometimes including the wrist and elbow, when the subject begins to write. From the early descriptions of Writer’s Cramp 4, 5 to the end of the nineteenth century the syndrome was considered an “organic” disease 4, 5 in the first half of the twentieth century, perhaps under the influence of developing psychoanalytic theories, it was often attributed to psychoneurosis 6, 7. Later it was thought to be due to a combination of physical and psychological factors. 8 In the last decade, evidence points again to an organic origin: occasional familial occurrence 9 and with other focal dystonias. 9 Writer’s cramp is broadly classified into two types 8

1. Simple writer’s cramp - appears only during writing
2. Dystonic writer’s cramp - appears during other activities also like eating and sewing etc.

Objective
The objective of the present study is to evaluate the neuro-physiology in patients of Writer s Cramp by the study of the following parameters in patients and comparing them with normals:

1. Long latency reflexes,
2. Somatosensory evoked potentials,
3. Brain stem auditory evoked potentials,
4. Blink reflex and
5. Visual evoked potentials.

Physiology Of writer’s Cramp

Evidence of motor abnormalities:
A neural circuit for motor control has been delineated in which cortical motor areas (primary motor cortex, premotor, supplementary motor and prefrontal areas) project to the putamen. 11 Direct and indirect pathways form a closed loop through the internal pallidum and thalamic nuclei back to the cortical level. 12 Neuronal activity in striatum and pallidum has been shown to reflect adaptation during sensory motor learning 13 and cognitive aspects of the performed movement respectively 14
A progressive disability in individual finger movement control with prolonged writing is often seen in Writer’s Cramp. The hand moves en bloc in an excessive grasping pattern. This grasping pattern could be due to a loss of cortical control of sub cortical structures. Specific neurons in the premotor cortex discharge in the sequencing of reach/grip movements, but not in isolated grip or reach movements. Task specific neurons are present in the supplementary motor area.

The critical mechanism appears to be an impairment of striato-pallido-thalamic pathways with disinhibition of excitatory thalamo-cortical neurons resulting in cortical over flow and dystonic postures. A dysfunction of the supplementary motor area in dystonic patients has been indicated by the findings of an enlarged amplitude of the N30 component of the somatosensory evoked potential and the attenuated blood flow in SMA increase during vibratory stimulation of the hand. Many studies using PET and magnetic stimulation demonstrated that there is cortical disinhibition with over activity of supplementary motor area causing dystonic postures in Writer’s cramp.

The application of long latency reflex testing in clinical electro physiology has so far been largely restricted to the polysynaptic brain stem reflexes and the monosynaptic Hoffmann reflex of the leg muscles. Since 1968, Marsden et al have performed a series of extensive studies on the electro physiologic effects of muscle stretch on the long flexor of the thumb in humans. They demonstrated the LLR in the thumb muscles and postulated a servomechanism of control of voluntary activity by the human motor cortex (Marsden 1972, 1976a). A comparison of the latencies of the LLR in various muscles led them to postulate the transcortical hypothesis (Marsden 1976b). Through a study of the LLR in various nervous system lesions they established further indirect evidence for a transcortical pathway (Marsden 1977a, b). While many hypotheses have been put forth to explain the basis of the LLR, the one that appears the most plausible is a transcortical polysynaptic reflex pathway, at least for responses recorded in the distal hand muscles (Fig 1). If the transcortical hypothesis were true, the LLRs would be altered in a variety of upper motor neuron lesions. Preliminary studies do suggest characteristic abnormal patterns in various motor lesions.

These reflexes may thus be a valuable tool in the evaluation of various CNS disorders. Long-latency reflexes (LLR) have been successfully used to test afferent sensory and efferent motor pathways traveling via the cortex. While the path physiology was still uncertain, studies in symptomatic dystonias demonstrated lesions of the lentiform nucleus, caudate nucleus and thalamus, indicating that dystonic disorders may be associated with basal ganglia dysfunction. Scanty reports on LLR testing in idiopathic focal dystonia patients have so far produced inconsistent findings. Roth well et al 1983, found no alterations of stretch-evoked LLRs on the clinically affected side.

Long Latency reflexes are believed to assist in voluntary control of skilled hand movements and assist rapid compensation of unexpected disturbances by a ‘servo mechanism’. Since, writing is a skilled movement using hand muscles, we selected long latency reflexes in writer’s cramp.

Somatosensory evoked potentials:
There are only few reports on somato sensory evoked potential testing in the evaluation of writer’s cramp. The N30 component of the median nerve somatosensory evoked potential is a variable component with a controversial site of origin. It is clearly influenced by motor behavior and found a decrease in N30 amplitude in patients with Writer’s cramp. Contrarily, Reilly and co-workers (1992) have found an enhancement of the amplitude of N30 in patients with writer’s cramp. The somatosensory evoked potential studies are difficult to interpret at this time but do suggest, as do the PET studies, that central sensory processing in dystonia may be Electrophysiological tests were performed on a Dantac EMG machine impaired. There is evidence of disordered spinal cord and brain stem reflexes in several types of focal dystonia.

We set out to assess Long latency reflexes, Somatosensory evoked reflexes, Blink reflex, Brain stem auditory evoked potentials, and Visual evoked potentials, on both the affected and non-affected side in Writer’s cramp patients as well as in healthy controls to determine the pathophysiological basis of this condition.

II. Material and methods
We investigated 14 patients with Writer’s Cramp including 10 patients with simple Writer’s cramp and 4 patients with dystonia Writer’s cramp. In all patients symptoms were confined to right hand. The diagnosis of Writer’s cramp was based on clinical presentation with focal dystonia without other neurological symptoms, and a normal neurological examination except focal dystonia. Throughout the text, the side showing the focal dystonia will be referred to as the ‘affected side’. Conversely the non-affected side denotes the side contralateral to the clinically symptomatic side. All 14 controls were healthy volunteers. All investigations were done with informed written consent of the patients and controls. All patients and controls underwent median nerve conduction studies including CMAP, MCV, M latency and F latency and SNAP and SCV on both the sides. Subjects with abnormal nerve conduction were excluded from the study.

Long latency reflexes:
Short latency reflex (SLR) and long latency reflex (LLR) muscle responses were elicited by stimulating the median nerve at the wrist and recorded from thenar muscles on the affected and non-affected side using surface electrodes. The patient was asked to contract thenar muscles by opposing thumb to the fifth finger so that a full EMG interference pattern could be seen on the screen. During electrical stimulation muscle contraction was maintained at ~ 20% of maximum force. The median nerve was stimulated (pulse duration
0.5ms) at the wrist at a rate of 3Hz. Stimulus strength was gradually increased to near motor threshold (1-5mA), until a small compound muscle action potential could be recorded. The signal was filtered (1-3000Hz) and averaged up to 500 times, which were repeated at least twice to ensure reproducible recordings. Amplitudes of all reflex components were measured peak to peak on non-rectified traces. Onset latencies of the brief response at ~ 30ms (corresponding to the H reflex). LLR 1 at ~ 40ms, LLR 2 at ~ 50ms and LLR 3 at ~ 75 ms were measured. Absolute latencies and amplitudes of all components were compared intraindividually and were also compared with control subjects.

**Stimulus parameters**

- **Site**: Wrist
- **Strength**: To obtain weak M response
- **Type**: Rectangular pulse
- **Duration**: 0.5 ms
- **Frequency**: 3 Hz
- **Total No of responses**: 500

**Recording parameters**

- **Site**: Thenar muscle
- **Band filter**: 1-3000 Hz
- **Sweep speed**: 200 ms
- **Sensitivity**: 200 pV

**Blink Reflex:**

The blink reflex was evoked by stimulation of the supraorbital nerve. Stimuli of 0.1ms duration and 3-20mA intensity were applied percutaneously at intervals of 10-20 seconds. The orbicularis occuli muscle responses were recorded by surface electrodes with the active electrode over the lower eyelid half way between the inner and outer edges of the orbit and reference electrode over the ipsilateralala of the nose. The blink reflex was considered abnormal based on the criteria (Hop.F 1991):

**R1 abnormalities:**

1. Absent R1
2. Latencies > 12ms
3. Side differences of > 1.2ms

**R2 abnormalities:**

1. Loss of R2
2. Latency >42.4ms
3. Side difference of > 5 ms

**Somato sensory evoked potentials (SSEP):**

Active surface electrodes were placed over the ipsilateral Erb’s point over second cervical spine and over the contra lateral hand area of the scalp i.e 2cms posterior and 6 cms lateral to Cz. Reference electrode was placed at Fz and ground over the wrist. Stimulation of medium nerve was done on each side using a stimuli with intensity ranging from 2-10mV with 10-20 ms duration with a frequency of 2Hz. There should be a visible painless twitch of abductor pollicis brevis. A total of 500 stimuli were averaged and repeated twice to demonstrate reproducibility. The filter was set at 30 to 3000 Hz. Latencies and amplitudes of Erb’s point potential, N 13 and N 20 potential from Erb’s point, cervical spine and contralateral hand area of the scalp respectively were recorded.

**Visual Evoked Potential (VEP):**

The electrodes were placed on the scalp according to the Queen Square system. The active midline occipital electrode was placed 5 cms above the inion. The midline frontal electrode was placed 12 cm above the nasion, which served as reference and the ground electrode was placed at vertex. The system bandwidth was set at 1 Hz to 300Hz with sensitivity of 20 microV and sweep of 30 ms. Pattern reversal stimulation of each eye was done using 12 X 16’ checkers at a frequency of 3Hz with patient sitting 1 meter away from the monitor. Averaging of 200 trials was repeated twice and were superimposed to demonstrate reproducibility.

**Brainstem auditory evoked potentials (BAEP):**

Active surface electrode was placed over the vertex, reference was placed over Ipsilateral mastoid process and ground was fixed 12 cms above the nasion. Rarefaction click stimulus was given to one ear with masking of contralateral ear, after measuring the minimum hearing threshold value for each ear. Stimulus was given 60-70 decibels above the threshold value, at a frequency of 10 Hz. The wave forms (I, II, III, IV and V) were recorded at a sweep speed of 1 ms duration, and 10 (iV sensitivity. Fiter was set between 10 -3000 Hz. Averaging of 2000 stimuli was repeated twice and were superimposed to demonstrate the reproducibility.
Statistics
The latencies and amplitudes of the LLR, Blink reflex, SSEP, BAEP and VEP are normally distributed, and so the unpaired t test was used for statistical analysis.

III. Results

Controls
All 14 who served as controls were healthy volunteers and all are males and right-handed individuals. Age of these subjects ranges from 25-56 yrs, with a mean of 39.07±9.9 yrs and a median of 39.5 yrs. The mean height was65.57±4.64 cm (157-173) and a mean arm length was 94.71+3.77 cm (87-100).

Patients
All 14 patients were right-handed individuals and all are males. Age of these patients ranges from 15 to 52 yrs with a mean of 28.57±12.02 yrs and a median of 39.5 yrs. The mean height of the patients was 165.79±7.26 (148 -174) cm and mean arm length was 94.50±4.74(85-104) cm.

Long latency reflexes

Controls
Details of components of the long latency reflex can be obtained from table 1 (latencies) and table 2 (amplitudes) (Fig 2). LLR 2 and 3 responses could be elicited from all 14 control subjects. LLR 1 was absent in 4 subjects of which, bilateral in 2, right side in one and left side in another.

Patients
Results of LLR measurements from the affected side and normal side in all 13 patients were given in table 3 (latencies) and table 5 (amplitudes) (Fig 3). Bilateral SLR could be recorded in all patients. Bilateral LLR 2 and LLR 3 were found in all patients. LLR 1 could be elicited bilaterally in 7 of 13 patients. LLR 1 could be elicited in all except two patients on the affected side. The LLR 1 amplitude was enhanced in all patients when compared to normal side, as well as in controls. In one LLR 1 was larger than normal side.In addition in some patients the LLR 1 abnormality was linked to LLR 2 abnormalities. LLR 2 amplitude in 11 of 13 patients was low in the affected side when compared to normal side (Table 5) as well as in controls. Amplitude ratio of LLR2 of the patients also was significantly different from the controls. LLR 2 amplitude was more in 2 patients where as equal on both the sides in one. No statistical difference was found in the amplitude of LLR3.There was no statistical difference in the latencies of LLR 1,2 and 3 between affected and normal side as well as controls (P>0.05).

Blink reflex:
Results of bilateral R1 and R2 measurements of all the 13 patients and control subjects were tabulated (table 8). Both R1 and R2 could be elicited from all subjects. There is no statistically significant difference in the latencies and interside difference of R1 and R2 in patients or controls (P=0.7304).

Somatosensory evoked potentials:
Results of bilateral Epi, N14 and N20 measurements in all the 13 patients and controls were given in table 9 (amplitudes) and table 10(latencies). There is no statistically significant difference in the latencies and amplitudes of patients and controls, either intra-individually or between the groups.

Brainstem auditory evoked potential:
Results of bilateral 1,3 and 5 potential measurements of all patients and controls were given in table 10 (amplitudes) and table 12(latencies). All 3 potentials could be recorded in all patients as well as control subjects. Statistically significant difference was not found in the latencies and amplitudes either intra individually or between patients and control groups.

Visual evoked potential:
Results of bilateral P100 measurements of all patients and control subjects were given in table 13. No statistically significant differences were observed in either latencies or amplitudes of P100 potential between patients and controls and also between right and left sides.

IV. Discussion

This study on LLR, Blink reflex, SSEP, BEAP and VEP in 13 focal task specific dystonia (Writer’s cramp) patients indicated abnormalities of LLR 1 in 85.71% of patients and LLR 2 in 78.57%.

Long latency reflexes:

LLR 1 abnormalities
The LLR1 was more frequently seen on the affected side than on the normal side. Frequency of occurrence of LLR1 on the unaffected side in patients was similar to the control group.

In 1997 Markus Naumann et al studied 36 focal dystonia patients, which included 7 patients of Writer’s cramp showed increased amplitude of LLR 1 in 24% of patients on the affected side when compared to normal side as well as to controls. Results of the present study are consistent with the above observations.

In the present study, in most cases LLR 1 alterations occurred on the clinically affected side (12 patients), but LLR 1 responses were also abnormal on the non-affected side in one patient. This is in keeping with radiological and electrophysiological findings in idiopathic dystonia showing that basal ganglia may be affected bilaterally despite unilateral clinical presentation.40, 44-47 At present the significance of such LLR 1 alterations is not clear. So far, LLR1 abnormalities were seen mainly in patients with extra pyramidal disorders.46 Our results extend these findings to other extra pyramidal disorders.
Long latency reflex studies in normal subjects have shown that the presence of a LLR 1 response is always associated with a well-defined LLR 2 48 in contrast LLR 1 abnormalities in focal task specific dystonia were combined with an absent or low LLR 2.44 Our findings are consistent with that of above observations. Sources for smaller or delayed LLR 2 responses are heterogeneous and include disorders of the afferent sensory or efferent motor pathways (Marsden et al 1977 a,b,Claus etal,1985).Normal SSEP in our study suggest that a efferent pathway disorder is more likely.SMA is a major cortical target for basal ganglia output. Lesions of the supplementary motor area (SMA) have resulted in increased LLR 2 responses in the animal model 49

Similar findings were observed in a patient with an SMA lesion.50 It has therefore been suggested that the SMA has an inhibitory influence on afferents to or the excitability level of cortical motor neurons thereby affecting the gain of LLR 2. Based on these findings, absence or reduction of LLR 2, as observed in our cases, argues for a disinhibition of the SMA by the basal ganglia in writer’s cramp.Recently increased motor cortex excitability has been shown by transcranial magnetic stimulation studies in dystonia patients.51 This supports the current pathophysiological concept of dystonia 20 suggesting that reduced strato- pallido-thalamic inhibition may lead to cortical overflow and dystonic postures. PET studies in dystonia patients have shown decreased regional cerebral blood flow in the posterior SMA 28,52 that has a direct influence on LLR 2. There is at present no satisfactory explanation for these two apparently contradictory findings, although an inverse relationship between regional brain metabolism and neuronal activity has been observed in patients with focal epilepsy (Pawlik et al 1994).

An inverse relation between LLR1 and LLR2 amplitudes was noted in our study. This inverse relation between LLR1 and LLR2 amplitudes may suggest an alternative explanation for a reduction or loss of LLR 2. A large or enhanced LLR 1 induces a partial refractoriness in the motor neuron pool and there by reduces the LLR 2 response.53 LLR 3 is an inconstant reflex found only in up to one third of normal subjects 31,52 No LLR 3 abnormalities were seen in Writer’s Cramp patients or controls.

Somatosensory Evoked potentials:
There are only few reports on somato sensory evoked potential testing in the evaluation of writer’s cramp. All have studied the N30 component of the median nerve somatosensory evoked potential which is a variable component with a controversial site of origin possibly in SMA 21,38.

Which were contradictory?
In our study we did not find statistically significant differences in the latencies or amplitudes of Somatosensory evoked potentials, Our findings could not demonstrate abnormalities in central sensory pathways in patients with writer’s cramp.

Brain stem auditory evoked potential ami Blink reflexes:
There is evidence of disordered spinal cord and brain stem reflexes in several types of focal dystonia.19.

In order to know the Brain stem involvement we studied BAEP and Blink reflex but there was no statistical difference between patients and controls. We could not demonstrate abnormalities of brain stem on the basis of our evaluation.

Visual evoked potentials:
Though there is no literature suggesting involvement of visual pathways in writer’s cramp, we did Visual evoked potentials in all patients and controls bilaterally and compared intra-individually as well as inter-individually, but we did not find any significant statistical difference between them.

V. Conclusions
A long latency reflex measurement is appears to be a reliable test to assess the functional impairment in writer’s cramp. Distinct alterations of LLR 1 and 2 were found in the affected limb in a substantial proportion of patients with Writer’ cramp. Normal Somatosensory evoked potentials in writer’s cramp suggest that the central sensory pathways are intact. Hence we may deduce that the abnormality in writer’s cramp involves the central motor pathways.Normal Brain stem auditory evoked potentials and Blink reflex suggest intact brain stem function.Visual evoked potentials in writer’s cramp were normal.

References

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Oster. The principles and practice of medicine, designed for the use of practitioners and students of medicine: Oster, William, Sir, 1849-1919: Free Download & Streaming [online]. Available at: https://archive.org/details/ Principlespractice\000od.


Hoover JE, Strick PL. Multiple output channels in the basal ganglia. SCIENCE-NEW YORK THEN WASHINGTON-1993:259:819-819.


Cheney PD, Fetz EE. Corticomotor neuronal cells contribute to long-latency stretch reflexes in the rhesus monkey. The Journal of physiology 1984;349:249-272.


Marsden CD, Merton PA, Morton HB. Stretch reflex and servo action in a variety of human muscles. The Journal of physiology 1976;259:531-560.

Lee MS, Marsden CD. Movement disorders following lesions of the thalamus or subthalamic region. Movement Disorders 1994;9:493-507.


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