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Motor Unit Territories in Juvenile Myoclonic Epilepsy Patients

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Abstract—**In this study, the scanning EMG technique was implemented to investigate electrophysiological cross-sections of the motor unit (MU) territories in healthy volunteers and in subjects with juvenile myoclonic epilepsy and spinal muscular atrophy. Measurements were taken intramuscularly by means of two concentric needle electrodes from biceps brachialis muscles. 3-D maps of the MU territories were plotted for each MU to determine the lengths of MU cross-section and the maximum amplitudes of each MU. There was evidence of a preponderance of large MUs in patients with juvenile myoclonic epilepsy.**

I. Introduction

Motor unit action potentials (MUAPs) detected by EMG using concentric needle electrodes (CNE) in clinical routine examinations are used in the diagnosis of neuromuscular diseases [1-4].

Juvenile myoclonic epilepsy (JME), a disorder characterized by myoclonic jerks with generalized tonic-clonic seizures is genetically linked to HLA region on chromosome 6, which is also responsible for the development of the spinal cord [5-7].

Subclinical anterior horn cell involvement was proposed in JME from a previous study performed with conventional EMG and the turn/amplitude analysis [8]. Large macro MUAPs with slightly increased fiber density (FD) found with macro EMG and decreased number of motor units found with the motor unit number estimate (MUNE) method suggested the preponderance of normal large motor units [9].

Above techniques provide only an estimate to MU size and do not give an insight about MU unit territory [9]. Therefore, a scanning EMG technique that reflects spatial and temporal characteristics of the MU is considered to reveal the preponderance of large MUs.

II. Measurement Procedure

The study involved 9 patients with juvenile myoclonic epilepsy (JME), 3 patients with spinal muscular atrophy (SMA) and 10 healthy subjects (NC).

An experimental EMG scanning system based on step by step recording of the electrical activity by means of two CNEs drawn through the MU territory was designed and implemented (Figure 1).

One of the electrodes (Medelec ELITE Disposable) was used as the scanning electrode and the other as the trigger electrode. An EMG instrument (Keypoint v.5.09) equipped with a D/A converter board was used.

The band-pass characteristics were 2 kHz-10 kHz for the trigger channel; and 10 Hz–10 kHz for the scanning channel.

The data acquisition system (NI-USB-6009, National Instruments) with a sampling rate of $48x10^3$ samples/second and 14-bit resolution was used to digitize the EMG signals.

Linear movement of the scanning electrode was obtained using a linear actuator (T-LA60A). Data acquisition and scanning processes were controlled with a computer. The scanning electrode was inserted into a specially manufactured needle attachment to affix it to the lead-screw of the linear actuator and connected to EMG instrument via a needle holder. The actuator was placed on the right arm of the subject lying in supine position.

Measurement procedure is as follows:

1. A concentric needle electrode was moved downward inside biceps brachialis muscle until an electrical activity of a MU was detected,
2. The trigger electrode was inserted as close as possible to the scanning electrode in a fashion that a time-locked single fiber action potential was detected on the display. The distance between the electrodes was 10 mm,
3. Scanning electrode was pulled linearly upwards in a 100-µm step size until no electrical activity was observed any further.
4. Scanning and trigger signals were digitized and stored in a text-file in the computer at each step.
5. Electrodes were inserted into different positions to record electrical activity from a different MU site.
6. This procedure was repeated for each MU.

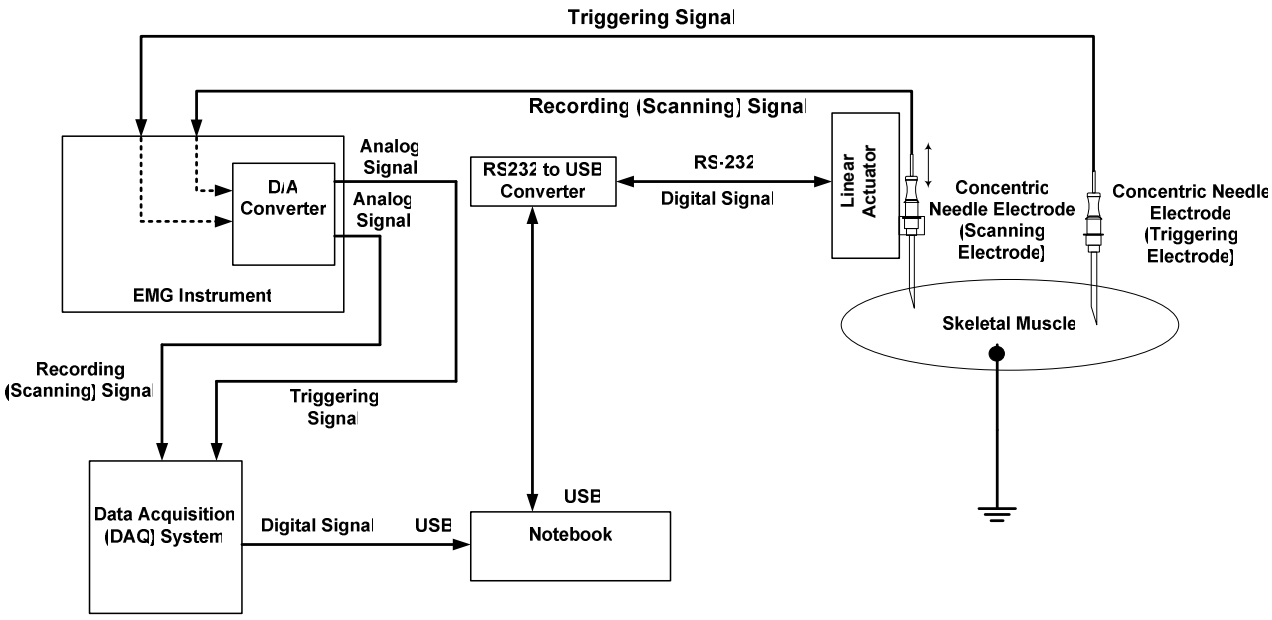

Fig 1. Block diagram of the scanning EMG system

TABLE 1. Descriptive statistics of motor units

	JME Group (n=52)		NC Group (n=51)		SMA Group (n=15)	
	LCS (cm)	Max. Amplitude (mv)	LCS (cm)	Max. Amplitude (mv)	LCS (cm)	Max. Amplitude (mv)
Min.	0.6	1.94	0.36	1.28	1.08	1.83
Max.	2.49	18.40	1.85	12.20	2.00	47.55
Mean±S.D.	1.4665±0.5	7.6843±3.17	1.1149±0.375	4.6594±2.54	1.7167±0.2434	10.1396±11.2646

LCS= Length of cross-section

TABLE 2. Descriptive statistics in terms of individual subjects

	JME Group (n=9)			NC Group (n=10)			SMA Group (n=3)		
	Min.	Max.	Mean±S.D.	Min.	Max.	Mean±S.D.	Min.	Max.	Mean±S.D.
Mean LCS (cm)	0.78	2.03	1.4538±0.44	0.71	1.50	1.1459±0.23	1.56	1.89	1.7167±0.16
Median LCS (cm)	0.74	2.04	1.4378±0.44	0.70	1.62	1.1455±0.28	1.53	1.90	1.7067±0.18
Maximum Amplitude Mean(mV)	4.82	12.30	7.56±2.48	1.76	7.30	4.4266±2.1	2.89	16.96	10.139±7.04
Maximum Amplitude Median (mV)	4.32	11.32	7.05±2.29	1.68	7.68	4.4585±2.26	2.90	10.81	7.8905±4.34

An M-File in MATLAB v.7.2 was created to extract scanning EMG signal data to a text-file. The data are time-locked with the trigger data to plot 3-D maps of the electrophysiological cross-section of the MU territory. The maximum amplitudes of the MUAPs were also detected by this subroutine. DC offsets were removed by median filtering through this subroutine. The steps where any electrical activity was present was counted on the plot and multiplied by the 100-µm step size to find the length of cross-section.

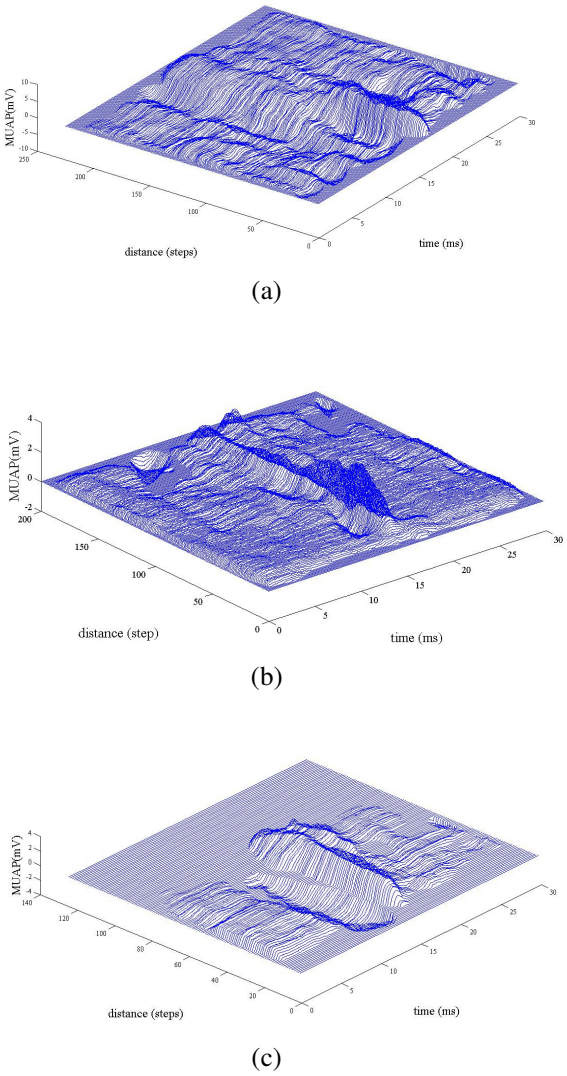

Fig 2. Three dimensional electrophysiological cross sections of MU territories of a patient with JME (a), a patient with SMA (b) and a healthy volunteer (c).

3-D plots for JME and SMA subjects and for a healthy volunteer are shown in Figure 2.

I. RESULTS

Results are given in Table 1. The difference in length of cross-sections is highly significant between JME and NC groups where as JME and SMA groups are similar (p<0.001, Tukey's honestly significant difference-HSD test). In terms of maximum amplitude, there is a significant difference between JME and NC groups (p<0.01, Tukey's HSD test), however, the difference was not significant between JME and SMA groups (p=0.19). The results are summarized in Table 2 in terms of individual subjects.

When the subjects were considered, there was a significant difference between JME and NC groups (p<0.01, Mann-Whitney and Kruskall-Wallis Methods). There is a little significant difference between JME and SMA group (p<0.1). The median lengths of cross-section are significantly different between JME and NC groups (p<0.01), however it is not significant between NC and SMA groups (p=0.16). The mean maximum amplitude is significantly different between JME and NC groups (p<0.01).

This difference is marginally significant between JME and SMA groups (p<0.1). For median maximum amplitudes, there is a significant difference between JME and NC groups (p<0.01) where as JME and SMA groups are not significant (p=0.64).

IV. DISCUSSION

Since the lengths of cross section of MUs are increased due to the reinnervation process in SMA, the similarity of JME group to SMA group both in lengths cross-section and in maximum amplitude suggest the preponderance of large MUs in JME. However, since the fiber density of MUs was found normal in a previous study, the presence of these large MUs can be considered as structural.

A correlation between the lengths of cross-section and the maximum amplitude is not expected in normal controls. This is the same for SMA group also, because, the amplitude increases due to the reinnervation even the length of cross-section does not increase. In the JME group, as the amplitude increases, the length of cross-section also increases.

The increase in the maximum amplitude with the increase in the length of cross-section was observed only in patients with JME. This may be due to the special configuration of MU territory in which the density of muscle fibers may be slightly greater than the normal configuration. This can also suggest that large MUs are structural rather than being originated from reinnervation.

The effects of age have not been shown on the amplitude and the length of cross-sections, except for the amplitude within normal subjects. Therefore, since this evidence may discard the role of progressive processes, it might be suggested that the presence of large motor units is structural.

In healthy individuals, skeletal muscles comprise motor units of various sizes. The muscle contraction

results from the recruitment of the motor units which implies the process where additional motor units are contributed to generate a muscle contraction at certain level [10][11][12]. According to Henneman's Size Principle, this recruitment generally takes place in an orderly sequence being based on the size of the motor unit as contraction increases [13][14]. The small motor units are recruited first, larger ones last to accomplish increasing gradations of contractile strength [10][14]. Hence, the fatigue is minimized. Furthermore, this size-ordered recruitment provides to perform fine motor tasks [13].

In conclusion, the motor units with large territories are dominant over the smaller motor units in JME patients. This may be present since the beginning of the life as a normal condition rather than a pathological process such as reinnervation. If the motor units with larger diameters are prominent in that muscle, difficulty in fine movement of that muscle is expected. Therefore, the clumsiness of JME patients may be related to this finding in spite of myoclonic jerks

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