Characteristic Alterations in Responses to Imposed Wrist Displacements in Parkinsonian Rigidity and Dystonia Musculorum Deformans

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ABSTRACT: The amplitude and temporal modulation of the segmented EMG activity in flexor carpi radialis, evoked by imposed angular wrist extension, was studied with respect to the level of pre-existing background activity in rigid parkinsonian (PK) and dystonia musculorum deformans (DMD) patients. The interdependence of the evoked M1 and M2-3 segments on pre-existing background EMG activity and initial velocity of imposed displacement was established previously for a normal population. Individual responses of 21 parkinsonian and 12 dystonic patients were compared to the established normal “response volume”. The augmented magnitude of the M2-3 segment in rigid PK patients, which correlates to the measure of rigidity, could not be accounted for by the low level of pre-existing EMG activity. Therefore, increased descending facilitation does not impinge directly on alpha motoneurons. Paradoxical excitation in the shortened muscle and resetting of tonic tremor of the stretched muscle by the imposed wrist extension are two other demonstrated abnormalities which may also contribute to PK rigidity. In contrast, DMD patients demonstrated normal amplitude modulation of the M1 and M2-3 segments, but exhibited a disturbance of normal temporal mechanisms that result in constant duration of the M1 and M2-3 responses with imposed force step loads.

RESUME: Nous avons étudié chez patients qui souffraient de la rigidité parkinsonienne ou de dystonie musculaire déformante, l'amplitude et la modulaton temporale de l'activité segmentaire EMG du muscle "flexor carpi radialis", évoquées par l'extension imposée angulaire du poignet, à l'égard du niveau de l'activité d'arrière-plan pré-existant. L'inter-dépendence des segments évoqués M2 et M2-3 sur l'activité EMG pré-existant fut décrit antérieurement chez une population normale. Les réponses individuelles du 21 patients parkinsoniens et 12 patients dystoniques furent comparées à les variations de témoins normaux. La magnitude augmentée du segment M2-3 chez les patients rigides parkinsoniens, ce qui bien correspond à la quantité de rigidity, ne pouvait pas s'expliquer par le niveau bas d'activité EMG pré-existant. Ainsi que la facilitation descendante augmentée n'affecte pas directement les motoneurones alpha. L'excitation paradoxale dans le muscle raccourci et la restauration du tremblement tonique du muscle tendu par l'extension imposée du poignet sont deux autres anomalies montrées qui pourraient contribuer aussi à la rigidité parkinsonienne. Par contre, les patients dystoniques montraient la modulation d'amplitude normale des segments M1 et M2-3, mais exhibaient une anomalie des mécanismes temporaux qui résultent à une durée constante des réponses M1 et M2-3 à les charges imposées.

activity, as compared to younger normals, apparently being unable to relax adequately.

In a companion paper, "response planes" which relate the magnitude of the M1 and the M2-3 segments simultaneously to the background or tonic level of EMG activity in the stretched muscle and the initial velocity of the imposed joint displacement were presented. The planes provide a means of quantitatively assessing the relative effect of pre-existing levels of EMG activity in the stretched muscles on the segmented responses (Bedingham and Tatton, 1984). "Response volumes" have also been calculated for populations of normal subjects which should enclose all of their individual response planes, and can potentially be used to determine if the magnitude of the response segments recorded in individual subjects differs significantly from the normal population. The normal studies also showed that the segmented responses to wrist displacements imposed by force step loads, although jointly amplitude modulated by the mechanical parameters and pre-existing EMG levels, are of constant duration despite several fold variations in the duration of the imposed movements.

These approaches for quantitative analysis of amplitude and duration control of the segmented reflex responses provide a means of testing the proposal of Evarts et al. (1979) regarding the long latency segments in parkinsonian patients. Accordingly, we examined the EMG responses to imposed wrist displacements in a series of rigid parkinsonian patients. In order to obtain comparative results we also examined both response amplitude modulation and duration control in patients with another extrapyramidal disorder, dystonia musculorum deformans (DMD).

The study established that increased levels of tonic EMG activity cannot account for the increased M2-3 segments in parkinsonian rigidity and response duration control is intact in this form of rigidity. In contrast, patients with DMD show an apparent disturbance of the mechanisms that result in constant duration of the segmented responses in normals while amplitude modulation appears normal.

METHODS

A total of 21 parkinsonian (PK) patients ranging in age from 35 to 72 years together with 12 DMD patients (14 to 40 years of age) were examined. All the patients were independently assessed by the same neurologist prior to electrophysiological evaluation. The PK patients were chosen because their predominant deficits were bradykinesia and rigidity. Rest tremor sufficient to prevent the patient from maintaining the wrist in a constant angular position precluded analysis (see below). The majority of the PK patients (18/21) were being treated with L-Dopa.

Only patients appearing to have the classic form of DMD (Marsden et al., 1976) were included in the results presented in this paper. Five of the twelve patients had undergone thalamotomy. In the case of unilateral thalamotomies, recordings from the wrist musculature contralateral to the thalamic lesion were not utilized for the results presented in this paper.

The patients were studied using the same apparatus, recording methods and computer analysis techniques described in detail for a normal population in the companion manuscript (Bedingham and Tatton, 1984). Briefly, angular step load displacements were imposed on the wrist joint by a computer controlled torque motor. Electromyographic activity was differentially recorded by surface electrodes placed over flexor carpi radialis (FCR) and extensor digitorum communis (EDC). Wrist position records and EMG activity were simultaneously recorded and analyzed. The magnitude of evoked reflex EMG activity above the pre-existing background level was normalized with respect to maximum voluntary activation (% MVA). As a measure of imposed mechanical input, the initial velocity (IV) of the displacement was computed (i.e. mean velocity during initial 35 ms) prior to the onset of the earliest reflex activity. Latencies and terminations of the EMG segments were measured from the points of intersection of the 3.0 standard deviation level above mean background EMG with the averaged traces. The mean magnitude of each reflex segment (i.e. M1 and M2-3) was plotted against the pre-existing level of background EMG activity and the IV of the displacement. This interdependent relationship was then compared to the range of values established previously for a normal population, which was represented graphically as a volume relating these three variables.

RESULTS

Figure 1 presents the age distribution for the thirty normal subjects studied in the companion paper (Bedingham and Tatton, 1984) together with that for the nine selected parkinsonian patients whose data will be used to illustrate our findings. The nine were chosen (with one exception indicated by an asterisk) because of their relatively young age in order to avoid the potential interpretive difficulties related to Evarts et al. (1979) suggestion that the increased M2-3 segments were a feature of the responses in the elderly.

![Figure 1](https://example.com/figure1.png)
Figure 2 presents a typical set of records for wrist displacements stretching FCR. The values at the left end of the position averages show the IVs. Background EMG (BKGD%) values are expressed as percentage of maximum voluntary activation (MVA) at the left end of the EMG averages, while M1 and M2-3 segment magnitudes above background level are shown at the right end of each record (RFLX%). The traces show that the M1 segments are small (increasing from 1 - 4% MVA) in keeping with the low background levels (1.6 - 2.0% MVA) as would be predicted by the M1 response planes constructed for the population of normal subjects (see Bedingham and Tatton, 1984). In contrast, the M2-3 segments are extremely prominent and demonstrate minimal amplitude modulation for IVs exceeding 93% sec. when compared to the monotonic grading found for normals over the IV range of 46 to 188% sec. In short, the responses for the rigid parkinsonian illustrated in figure 2 show M2-3 segments with amplitudes exceeding those found in normals at background levels of as high as 14% MVA while the M1 segments amplitudes appear to be within the range of normals for the background levels.

The parkinsonian patients demonstrated two other abnormalities to the imposed wrist displacements as described previously (Lee and Tatton, 1975; Tatton et al., 1979). These are illustrated in figure 5. The averaged records in this figure have a 630 msec time base after the onset of the randomly-timed wrist displacements. The first abnormality is a phasic increase in EMG activity in the shortened muscle (in this case EDC) at about the same time as the increased M2-3 segment in the muscle being stretched (FCR). This “paradoxical” excitation (termed as the Westphal phenomenon) can be seen in some normals, although less prominently. The second abnormality consists of a six to ten Hz oscillation in the EMG activity subsequent to the increased M2-3 segment in the stretched muscle (the first burst between 30 and 105 msec is the M1 and M2-3 segments, while in this case the peak to peak interval between the subsequent three cyclic bursts in the average is 160 msec). The bursts typically “damped out” after four to six cycles. Since the imposed step loads were presented randomly in time, the oscillatory activity represents

In order to explore the relationship between M2-3 amplitude and clinical rigidity the nine young PKs presented in the age distribution were separated into two groups according to the severity of rigidity that could be detected clinically in the wrist musculature. Patients with +4 rigidity were not included in this group since the degree of rigidity was sufficient to prevent an adequate range of IVs from being examined. A scale of 0-4 was used according to the following criteria:—

0 = Absent.
+1 = Slight or detectable only when activated by contralateral wrist movements.
+2 = Mild to moderate.
+3 = Marked, but full range of movement easily achieved.
+4 = Severe, range of motion achieved with difficulty.

Figures 3 and 4 present data from four PK patients with minimal rigidity (grade 0, +1) over-plotted on normal response volumes in the left sides of the figures (labelled NON RIGID) while the plots on the right sides are for five PK patients with mild to moderate rigidity (grade +1, +2 or +2, +3; labelled RIGID). The response volumes were calculated for the M1 and M2-3 segments in the normal population (Bedingham and Tatton, 1984). Both figures present average response values of five different BKGD-IV measurements for each patient. Different symbols for each patient are plotted relative to the normal volumes.

All M1 segment values fell just below or within the volumes for both the RIGID and NON RIGID patients with one exception. Three of the values for the 72 year old patient just exceed the upper surface of the normal volume (the reason for this patient’s inclusion is that she was the only PK studied to date whose M1 values have exceeded the upper surface). In marked contrast all of the M2-3 values for the RIGID PKs exceed the upper surface volume while those for the NON RIGID patients are either just beneath or within the volume.

In particular, note that the M2-3 values exceed the normal response volumes for all values of background EMG, even those below 4% MVA. This clearly demonstrates that increased tonic levels of EMG activity in the stretched muscle are not a prerequisite for increased M2-3 segments as proposed by Evarts et al. (1979). Furthermore, the M1 segment values are not increased above the normal volumes in the RIGID PKs for the same background levels of activity.
Figure 3 — Response volumes for a normal population taken from Bedingham and Tatton (1984). Five BKGD-IV measurements for the FCR M1 segments in NON RIGID and RIGID PK's are superimposed.

Figure 4 — Similar to figure 3 with the values plotted for the M2-3 segments.

DMD patients showed a consistent abnormality different from that found in the rigid PKs. A typical example is presented in figure 6. The labelling conventions are the same as those in figure 2. This patient shows moderately high background EMG levels (5.4% - 10.9%). The M1 segment has a normal latency and grades monotonically from 13 - 23% MVA for IVs varying from 87 to 204°/sec. Although the mean level of long latency activity (labelled M2/M3p) is similarly graded (increasing from 20 - 29% MVA for the same IV values), it is prolonged and appears to continue as long as the imposed movement is in progress. This
continued activity is in sharp contrast to the constant segment terminations found for step load imposed displacements in both normals (Bedingham and Tatton, 1984) and rigid PKs (see figure 2).

The prolonged long latency activity makes the data inappropriate to superimpose on response volumes for the normal M2-3 segments. Input-output plots show that the slope of the graded responses increases with BKGD levels in a manner similar to normals (Bedingham and Tatton, 1984). Typical examples are presented for four DMDs in figure 7 for both the M1 and the prolonged long latency activity labelled M2/3p.

Figure 8 best illustrates the characteristic duration abnormality found in the DMD patients. Although the M1 segment latency remains constant to within ±2 msec as in normals (the shaded regions show the range of values for normal subjects taken from Bedingham and Tatton, 1984), the duration of summed M1 and M2-3p activity (labelled M1/2/3p duration) decreases almost proportionally to the duration of the imposed displacement (as shown by comparing the plots for displacement duration and M1/2/3p durations). This is in contrast to normals whose M1/2/3 duration is constant within ±2 msec irrespective of the displacement duration.

DISCUSSION

The response volume analysis of M2-3 segments for rigid parkinsonian patients establishes that the increased M2-3 segment in these patients is not merely a reflection of high tonic levels of alpha motoneuron firing. Figure 9 schematizes the experimental paradigm, the wrist joint, and the various populations of muscle, joint and cutaneous mechanoreceptors that

Figure 5 — Record illustrating the paradoxical excitation in the shortened muscle and the 6-10Hz oscillation in the stretched muscle seen in rigid PKs.

Figure 6 — Average records for wrist displacements in a patient with DMD. labelling and other conventions are identical to those in figure 1.

Figure 7 — Input-output plots relating M1 and M2-3p magnitude to initial velocity in four DMD patients. Background levels are shown at the right end for each plot.
likely are activated by the imposed angular joint displacements. The three arrows labelled 1, 2 and 3 are included to indicate the different abnormal mechanisms that have been proposed to underlie parkinsonian rigidity (for details and references see Tatton et al., 1979): (1) Increased descending drive to alpha motoneurons. The present study rules against this mechanism as the major cause of rigidity. High levels of background EMG would have been expected and the M1 and/or the M2-3 segment would not have exceeded the upper surface of the volumes particularly at low background levels. In fact, we found that most of the parkinsonians examined demonstrated low resting background levels in contrast to the elderly normals studied by Evarts et al. (1979). (2) Increased descending drive to static gamma motoneurons. Recordings of single la fibers from muscle spindles in rigid parkinsonians using neurographic techniques (Burke et al., 1977) have not supported this mechanism. Furthermore, if the M1 segment receives a contribution from spindle afferent evoked motoneuron activity and static gamma drive was increased then both the M1 and the M2-3 segments would be expected to be similarly increased in magnitude. Complex scenarios could be proposed according to which increased gamma drive was phasically modulated so as to selectively alter the M2-3 segment but these are beyond the scope of our non-invasive studies to confirm or deny. (3) Increased excitation of interneurons in polysynaptic pathways activated by one or more of the mechanoreceptor populations offers the simplest mechanism to account for the increased M2-3 segment.

Recent research has unambiguously established that the long latency segments in the distal upper limb flexors in the primate are dependent on motor cortical integrity for their evolution (Lenz et al., 1983; see Wiesendanger and Miles, 1982 for a recent review) and that single motor cortical neurons do contribute to the generation of EMG activity during the M2 segment in monkey wrist muscles (Cheney and Fetz, 1984; see Tatton et al., 1983 for a review comparing homology between reflex responses in monkey and human wrist muscles). Furthermore, Filion (1979) has shown increased levels of spontaneous activity in the globus pallidus internus (GPI) of monkeys rendered rigid and bradykinetic by midbrain lesions. The GPI projects to neurons in areas x and VLo of the thalamus which in turn send axons to the supplementary motor area (SMA) and the arcuate premotor cortex (APA, see Wiesendanger 1981 for review of the connections and functions of the association and supplementary motor area). Both APA and SMA project in turn to the motor cortex.

It was previously proposed that increased GPI output reaching area 4 through the thalamus might abnormally facilitate the polysynaptic transcortical reflex (Tatton and Lee, 1975) and, thereby, result in increased M2-3 segments. Recent evidence firmly establishing the posit that transcortical reflex activity contributes to the generation of the long latency segments in the primate distal upper limb makes it tenable to consider that the interneuron in figure 9 receiving the increased drive to be located in the motor cortex. Yet, this suggestion is at best speculative until the functional operation of the linkages between GPI and area 4 are better defined. In fact, the interneuron(s) could be just as easily located within spinal polysynaptic circuits which receive increased descending facilitory drive and terminate predominantly on the “fast” motoneuron population (see Bawa and Tatton, 1979). In fact, the increase in the M2-3 segments in rigid PK patients may not reflect an abnormality in the mechanisms which generate the M2-3 segments in normals. The M2-3 activity may be summed with appropriately-timed activity which is not evoked in non-rigid subjects.

Lee and Tatton (1982) showed that tendon taps at the wrist evoke activity over the interval of the M1 segment while angular displacements evoke both the M1 and M2-3 segments. Mortimer and Webster (1979) reported a high correlation between a mechanical measure of parkinsonian rigidity and the relative
size of the M2-3 segment. The normal or even slightly decreased M1 segments is in keeping with the usual clinical finding of normal tendon jerks in rigid parkinsonians, and it could be proposed parkinsonian rigidity is almost entirely due to mechanisms manifesting the increased M2-3 segments. This view should be taken with some caution since the Westphal phenomenon and the 6-10 Hz oscillatory activity would also be expected to contribute to the muscle tension production underlying rigidity.

The continuation of evoked EMG activity in the DMD patients as long as the displacement continues is in sharp contrast to normal subjects and rigid parkinsonians who respond with constant duration responses to imposed step loads, independent of displacement duration (Bedingham and Tatton, 1984). Previously, H reflex studies have failed to reveal a characteristic abnormality in DMD (Sax et al., 1976). The failure of the activity to terminate in a manner similar to normals may indicate dysfunction in an inhibitory mechanism which limits the duration of the reflex response. This may be one aspect of a generalized failure in sensorimotor mechanisms designed to limit the time course of motoneuron activity. Yet, other abnormalities such as increased sensitivity to one or more movement variables must be also considered. Studies using other imposed inputs with variable acceleration profiles (see Bedingham and Tatton, 1984) may serve to better define this apparent reflex abnormality and provide information regarding the mechanisms of rigidity.

In any case, the two extrapyramidal disorders, both characterized by abnormal muscle tone, show strikingly different abnormalities in their EMG responses to step imposed wrist displacements. In rigid parkinsonian patients there is an abnormality in reflex amplitude control, specifically increased sensitivity of the mechanisms generating the M2-3 segment to one or more parameters of the imposed movement, while duration control remains intact. DMD patients have normal reflex amplitudes but present a failure in duration control.

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