

Pathophysiological mechanisms in cerebral palsy

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SUMMARY To investigate some of the pathophysiological mechanisms in cerebral palsy, surface electromyograms (EMG) were recorded from pairs of flexor/extensor muscles during both voluntary and passive flexion/extension of upper and lower limbs of 20 patients. Elbow, knee, or ankle joint angles were measured simultaneously, as well as the force required to flex/extend the limbs passively at frequencies of 0.1–1.0 Hz. In addition, single motor units were recorded from the first dorsal interosseous muscles of six of the patients. Almost all patients showed resistance to passive movements (hypertonia). This hypertonia did not necessarily impair voluntary flexion/extension movements if alternating EMG activity was maintained in at least one of the pairs of flexor/extensor muscles involved in the movement. In six severely involved patients, there was a complete breakdown in the reciprocal relationship between reciprocally acting pairs of flexor/extensor motoneurons, which resulted in synchronous activation (co-contractions) of flexor/extensor muscles during both voluntary and passive movements. In these patients the hyperactive segmental reflex added to the disabling effects of co-contractions during voluntary movements. Single motor units recorded from patients with dystonic movements were recruited with variable delays (2–10 s) and usually discharged intermittently at high frequencies (60–120/s). This abnormal motor unit discharge pattern may relate to pathology of the basal ganglia.

Cerebral palsy is a term used to denote a wide range of motor abnormalities which result from damage to the developing or immature brain. Chronic electrical stimulation of the cerebellar cortex was introduced as a means of improving motor function in patients with motor system abnormalities, including the various forms of cerebral palsy (Cooper, 1973; Cooper *et al.*, 1973). Clinical and neurophysiological changes after chronic cerebellar stimulation have been reported (Cooper *et al.*, 1976; Upton and Cooper, 1976; Davis, 1977; Larson, 1977; Penn and Etzel, 1977; Fisher and Penn, 1978; Gottlieb *et al.*, 1978; McLellan *et al.*, 1978). However, some of the neurophysiological changes were not related to the clinical changes, which made it more difficult to explain the effects of chronic cerebellar stimulation on the motor system. Part of the problem may be that in cerebral palsy the aetiological

factors, location of lesions, and consequent manifestations are diverse; and the clinical classifications and terminology are inadequate.

To study the pathophysiological basis of the motor abnormalities and any changes which may occur because of cerebellar stimulation, we devised a series of simple physiological tests which the patients could perform without difficulty. This paper reports on the results of these tests on 20 cerebral palsy patients and the pathophysiological bases of the abnormalities.

Methods

A simple protocol was used so that even severely involved patients could be studied. Although the co-operation of the patients was necessary, their full attention was not required in all the tests. The procedures were designed to test the "reflex" responses of the upper and lower limbs to passive movements and how they might be related to the voluntary movement deficits and the involuntary movements.

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MEASUREMENT OF ELECTRICAL AND MECHANICAL PARAMETERS

During voluntary movements, surface electromyograms (EMG) from a pair of flexor/extensor muscles and the joint angles were recorded; the force required to move the limb was also recorded during passive movements.

Joint angles

The patient was first comfortably seated in a chair. A goniometer employing a linear potentiometer for measuring joint angles was positioned around a joint (elbow, knee, or ankle), and secured with Velcro straps.

Force

The force required to flex and extend the elbow or knee was measured with a strain gauge bonded on a metal bracket. This "force-bracket" was strapped around the wrist or ankle and occasionally the foot. The limb was then manually moved by gripping the handle on the force bracket.

EMG recording

A pair of circular disc surface electrodes filled with conducting cream were attached to the bellies of pairs of flexor/extensor muscles, using adhesive rings and surgical tapes. During movement around the elbow joint, EMGs were recorded from the biceps/triceps, during flexion/extension of knee joint, EMGs were recorded from quadriceps/hamstring muscles, and finally when the ankle was dorsiflexed/plantarflexed, EMGs were recorded from triceps surae/anterior tibial muscles. The EMGs were amplified by a Tektronics 2AG1 unit with 60–600 Hz bandwidth, and filtered by a third order low-pass filter with 100 Hz cut-off.

Voluntary and passive movements

Patients were asked to flex/extend their elbows at slow rates (0.1 to 0.5 Hz) 10 times and at their fastest rates: EMG from biceps/triceps and elbow joint angles were recorded on a Hewlett-Packard (HP3964A) tape recorder. The elbow was then passively flexed/extended 10 times at frequencies of 0.1, 0.2, 0.5, and 1.0 Hz and the angle, force, and EMG activity measured. The frequencies were restricted to less than 2 Hz because the patients were not capable of voluntary movements at faster rates. The procedure was then repeated at the knee joint. The effect of muscle length on passive responses was studied in a few patients by varying the knee joint angle. In patients who could dorsiflex/plantarflex their ankles voluntarily, voluntary and passive movements around the ankle joint were investigated. Each recording

session lasted one to one and a half hours, depending upon the extent of motor disability, the co-operation, and mood of the patient.

Motor units

Single motor units were recorded from six patients aged 15 to 27 years with a TECA (TG-4) EMG apparatus, an FM tape recorder, and a signal discriminator. The patients were seated in a wheelchair with one arm resting on a board on top of a bed. The force transducer (Grass FT.03) was clamped rigidly to a metal rod fixed to the board. The output from the transducer amplifier (Grass 7P-122) was connected to a calibrated digital multimeter so that the force exerted on the transducer could be read directly. A bipolar needle electrode (DISA 13K80) was inserted into the first dorsal interosseous muscle and positioned so that the discharge of single motor units would be distinguished at a moderate level of voluntary contractions. In one patient motor unit potentials were recorded from tibialis anterior muscle.

The electrical activity was (i) amplified (input impedance 100 megohms), (ii) filtered with a low frequency cut-off of 32 Hz and a high frequency cut-off of 3.2 kHz, (iii) fed through a delay line and trigger level, (iv) fed into a discriminator which gave out a pulse any time the selected motor unit discharged, (v) displayed on an oscilloscope, and (vi) recorded on the FM tape recorder.

Patients were provided with audiovisual feedback of the electrical signals and were asked to maintain a continuous discharge of the single motor unit for about 30 s and then to increase the firing frequency of the same motor unit by increasing the force. At each recording session which lasted 30 minutes, 5 to 10 motor unit potentials were recorded at different force levels.

The DC force, the motor unit impulses recorded with the needle, and the discriminator pulses were played back through a photosensitive chart recorder (Kodak Linograph 1895), and the discharge frequencies computed. The figures from the motor unit potential studies were retraced because the direct photographic prints of the traces on the photosensitive paper proved unsatisfactory.

Patients

There were three sources of patients: (i) six patients who were already undergoing chronic cerebellar stimulation, (ii) five patients who were possible candidates for cerebellar implants, and (iii) nine patients from schools and hospitals for handicapped children. In all cases the protocol

was presented in writing to the staff of the hospitals and schools, the patients (as appropriate), and their families. Furthermore, a clinical research co-ordinator fully discussed the details of the procedure with staff, patients, and families. Before testing, consent forms approved by our Medical Center Committee on Human Investigation were signed by the adult patients. In the case of minors and the severely involved, the consent forms were signed by a parent or legal guardian.

Terminology

In view of the diversity and complexity of "spasticity" and "rigidity" (Landau, 1974), the use of any of these terms will be restricted only to the initial neurological classification.

Results

Data from only six patients will be presented because their combined data completely represent all the pathophysiological mechanisms that were observed in the 20 patients studied.

CASE 1

This 26 year old man (DO) was quadriplegic, athetoid, dystonic, and unable to walk. He was being treated with chronic cerebellar stimulation, but the stimulator was turned off 12 hr before testing. This patient was so severely involved that any improvement due to the stimulation will not be taken into consideration.

Upper limb movements

The patient was asked to flex/extend his elbow voluntarily and Fig. 1A shows the resulting elbow angle, biceps and triceps EMGs. Compared with the normal subject (Fig. 1B) two abnormalities were obvious: (i) the movement was not continuous, but jerky and decomposed, and (ii) the phasic EMG activity in the biceps/triceps did show abundant coactivation. In order to get a better understanding of the nature of this abnormal electrical discharge pattern, single motor unit potentials were recorded from this patient, which will be presented later.

The elbow was flexed/extended manually using the metal bracket attached to the wrist at fre-

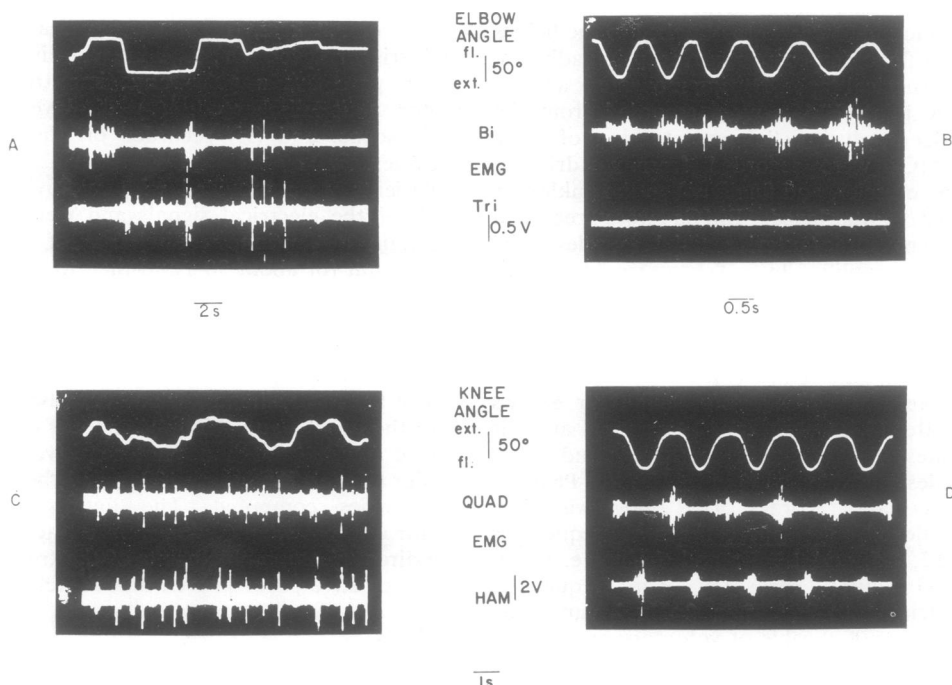


Fig. 1 Voluntary flexion/extension of elbow joint: (A) patient DO, (B) normal subject. Voluntary flexion/extension of knee joint: (C) patient DD, (D) normal subject. Upper traces are joint angles, middle and bottom traces are EMGs from pairs of flexor/extensor muscles. In A and C note the synchronous EMG activity in pairs of flexor/extensor muscles; in D there was reciprocal EMG activity between quadriceps and hamstrings. Note the EMG calibration was after amplification of 1000 and filtering.

frequencies of 0.2, 0.5, and 1.0 Hz. The force required to move the limb through an angle of 10° was computed as a measure of the resistance to passive movement. Figure 2A and B shows the elbow angle, force and biceps EMG recordings from this patient at 0.2 and 0.5 Hz. The mean force was computed from the peak-to-peak amplitudes of 10 force traces; the resistances to passive movement were $260 \text{ g}/10^\circ$ at 0.2 Hz and $320 \text{ g}/10^\circ$ at 0.5 Hz.

The distortion of both the angle and force traces at 0.5 Hz was due to the involuntary movements, which suggests that the dystonic phenomenon may be potentiated by a hyperactive segmental reflex mechanism depending upon the frequency of movement. This effect was observed in five other patients.

The resistance to passive elbow movements of this patient and another patient with similar abnormal behaviour are compared with two normal subjects in Fig. 2C. In all four subjects the force required to move the limb increased with

increasing velocity of stretch, but the forces were more than twice as large in the two patients.

Lower limb movements

Voluntary flexion/extension of the knee was jerky, and continuous EMG activity was seen in both quadriceps and hamstrings; in addition, multiple periods of phasic discharges occurred (Fig. 1C). When the resistance to passive movements was compared with that of a normal subject an increase in resistance with increasing velocity of passive stretch was demonstrated in both, although the forces were again twice as large in the patient.

Motor unit activity recorded from the above patient (DO) and a second patient (PP) will be presented here. Motor unit potentials recorded from the other four patients with no dystonic movements fired normally at 6–15/s.

In patient DO the characteristic discharge of the few motor units recorded from the first dorsal interosseous muscle was as follows (Fig. 3A): (i) motor units fired in bursts of high frequencies

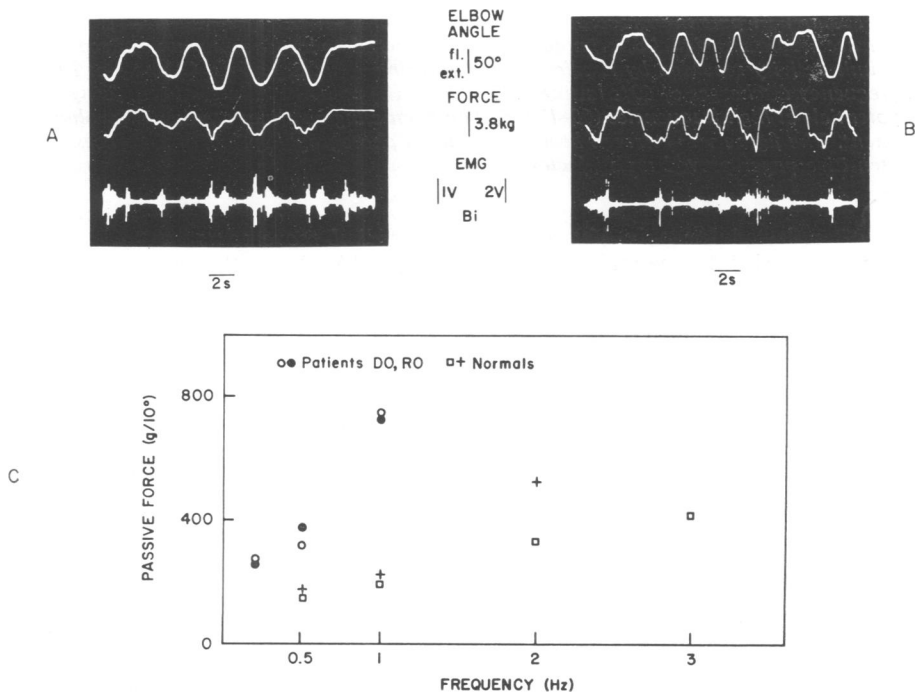


Fig. 2 Measurement of resistance to passive movement (patient DO). The upper traces are the elbow joint angles, the second traces, the force required to flex/extend elbow continuously at 0.2 Hz (A) and 0.5 Hz (B), and the third traces are EMGs recorded from biceps. (C) Plot of force required to flex/extend elbow passively at different frequencies; two patients and two normal subjects. Patients' frequencies were 0.2, 0.5, and 1.0 Hz; normal subjects' frequencies were 0.5, 1.2, and 3 Hz. Force computed at 1.0 Hz was 190 and 230 g/10° for normal subjects, 730 and 750 g/10° for patients.

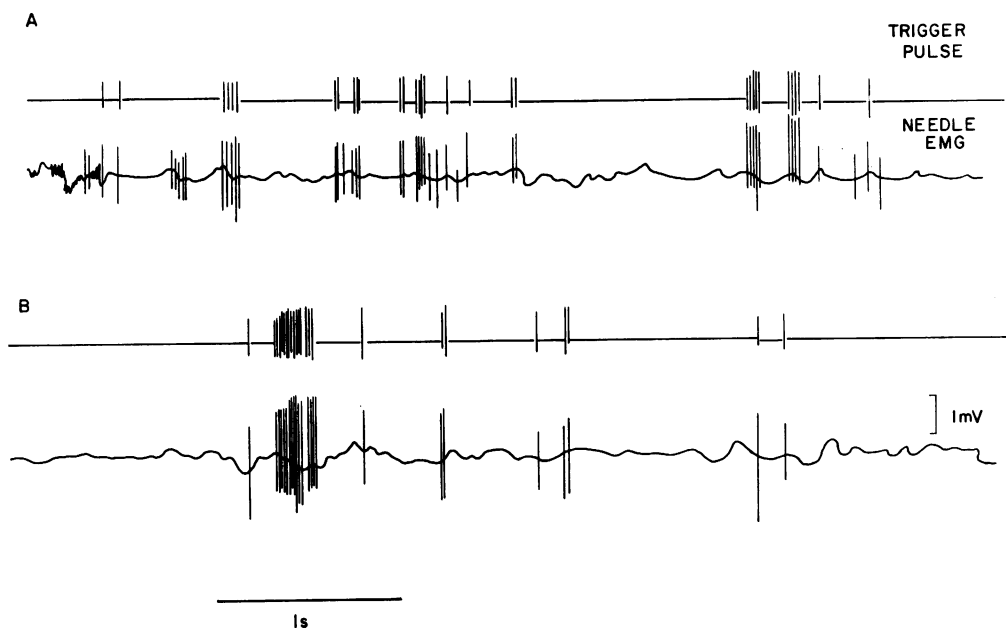


Fig. 3 Motor unit potentials recorded from the first dorsal interosseus muscles of two patients. Upper traces represent signal discriminator pulses triggered by larger motor units. Second trace is motor unit activity recorded by needle electrodes during voluntary effort. (A) patient DO shows high frequency discharges of 60/s lasting only 100 ms. (B) patient PP: intermittent firing of motor unit potentials at high frequencies (60–120/s). The fluctuating baselines are due to involuntary movement. The figure was retraced because the direct photographic prints of the traces on the photosensitive paper proved unsatisfactory.

(60/s), which often lasted for only 100 ms; (ii) although the patient tried to maintain a steady voluntary contraction variable periods (0.5–2 s) of motor unit inactivity were observed; (iii) variable delays (2–10 s) in motor unit recruitment occurred.

Patient PP is a 27 year old man, quadriplegic with rigidity and dystonic movements. The firing pattern of his motor units was investigated from the first dorsal interosseus and anterior tibial muscles. Figure 3B shows a 5 s recording of multiple motor unit potentials from the first dorsal interosseus muscle while the patient was asked to exert a continuous force on a transducer placed between his thumb and first finger. The abnormal firing characteristics were: (i) a delay in motor units recruitment of 2–5 s, (ii) intermittent firing of motor units, and (iii) brief (0.25 s) high frequency (60–120/s) phasic bursts. The possible relationship between this abnormal firing pattern and lesions in the basal ganglia will be discussed.

CASE 2

This 27 year old woman (SS) was spastic, quadriplegic, and unable to walk.

Upper limb movements

Figure 4A shows the angle, biceps, and triceps EMGs during voluntary flexion/extension (Fl/Ext) of her elbow at 0.2 Hz. Triceps EMG showed alternating activity during extension while biceps EMG was continuous and random during both flexion/extension. There were also periods of simultaneous increase in electrical activity in both biceps and triceps during extension, indicating co-contraction of this pair of flexor/extensor muscles.

Resistance to passive flexion/extension of the elbow at 0.5 and 1.0 Hz were 230 g/10° and 300 g/10° respectively; these were larger than corresponding average values of 150 g/10° and 190 g/10° recorded from normal women of comparable size and age. A comparison of the reflex EMG responses of the flexors and extensors indicated that, while the threshold frequency of stretch for eliciting a reflex response in the extensors (triceps) was less than 0.2 Hz, negligible reflex EMG was recorded from the flexors (biceps) at 1.0 Hz. Hence in this patient there was an increase in extensor motoneurone reactivity com-

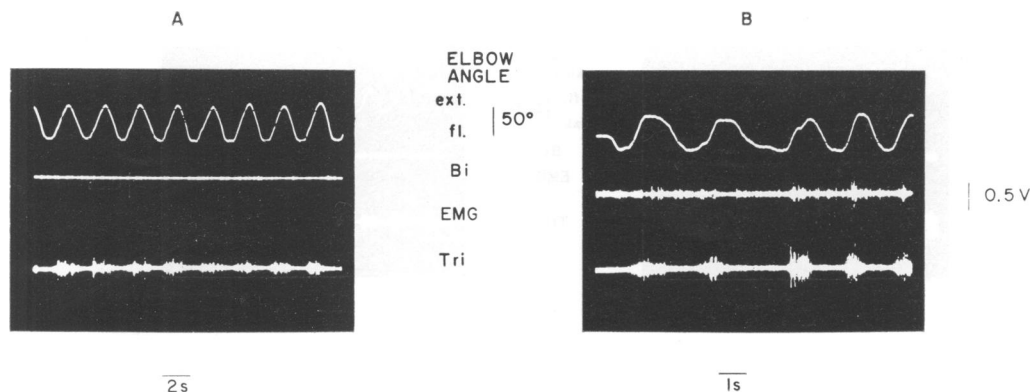


Fig. 4 Patient SS. First (top) traces are elbow angles, second and third traces are biceps and triceps EMGs respectively. (A) Voluntary flexion/extension of elbow, (B) passive flexion/extension at 1.0 Hz. Note the lack of reciprocal activity in biceps, with periods of synchronous EMG activity in both biceps and triceps during extension.

pared to normal subjects as well as a relative increase in extensor reactivity compared to flexors.

Lower limb movements

Contractures limited knee movements to 10° , and little EMG activity was recorded from the quadriceps and hamstring muscles.

The knee joint could be moved passively through only 20° . The resistance to movement was $2.3 \text{ kg}/10^\circ$ at 0.5 Hz (cf $1.0 \text{ kg}/10^\circ$ recorded from normal subject). The lack of significant reflex EMG responses from quadriceps and hamstrings indicated that the increased resistance was due to the contractures, not to hyperactive stretch reflexes.

CASE 3

This 10 year old boy (TP) was quadriparetic, mildly "spastic," and able to walk.

Upper limb movements

The patient did not have any difficulty with voluntary flexion/extension at the elbow (Fig. 5A). However, overlapping EMG activity between biceps and triceps muscles was recorded during extension. Such an alternating pattern of EMG activity from the triceps is normal; but, at a frequency of 0.5 Hz, the electrical activity was excessive compared with normal subjects (Fig. 1B), suggesting hyperactive extensor motoneurone activity.

Figure 5B shows elbow angle, biceps, and triceps EMG during passive flexion/extension at 1.0 Hz. Triceps EMG activity occurred during passive flexion only, the biceps showed a random continuous pattern of EMG activity. The resistance

to passive movements of $330 \text{ g}/10^\circ$ at 1.0 Hz, was greater than the corresponding $200 \text{ g}/10^\circ$ computed for normal subjects. The resistance (computed as the force required to move the limb through 10°) showed the normal pattern of increase in resistance with increasing frequency of passive movement (Fig. 5C).

Thus this patient showed (i) an increase in the reactivity of extensors during both voluntary and passive movements, (ii) a low threshold frequency for producing reflex EMG response in the triceps ($<0.1 \text{ Hz}$), and (iii) an increased resistance to passive movement. In contrast to case 2 the patient had no difficulty in flexing and extending his elbow. This implies that an increase in reactivity may not necessarily impair movement provided either the flexors or extensors of the particular joint retain the normal pattern of alternating EMG during voluntary flexion/extension.

Lower limb movements

Figure 6A is a record of the knee joint angle, quadriceps (extensors), and hamstrings (flexors) EMG, during voluntary flexion/extension of the knee at 0.5 Hz. While the hamstrings showed alternating EMG activity, the quadriceps had a more continuous tonic EMG activity with intermittent periods of increased electrical activity.

Passive flexion/extension at 0.1 Hz produced reflex EMG responses in the hamstrings. The resistance to passive movements at 0.5 Hz was $620 \text{ g}/10^\circ$ and within the normal range. The resistance increased with the frequency of passive movements. Figure 6B demonstrates the dynamic spindle sensitivity at 0.5 Hz, and also tonic EMG activity when the limb was held stationary for

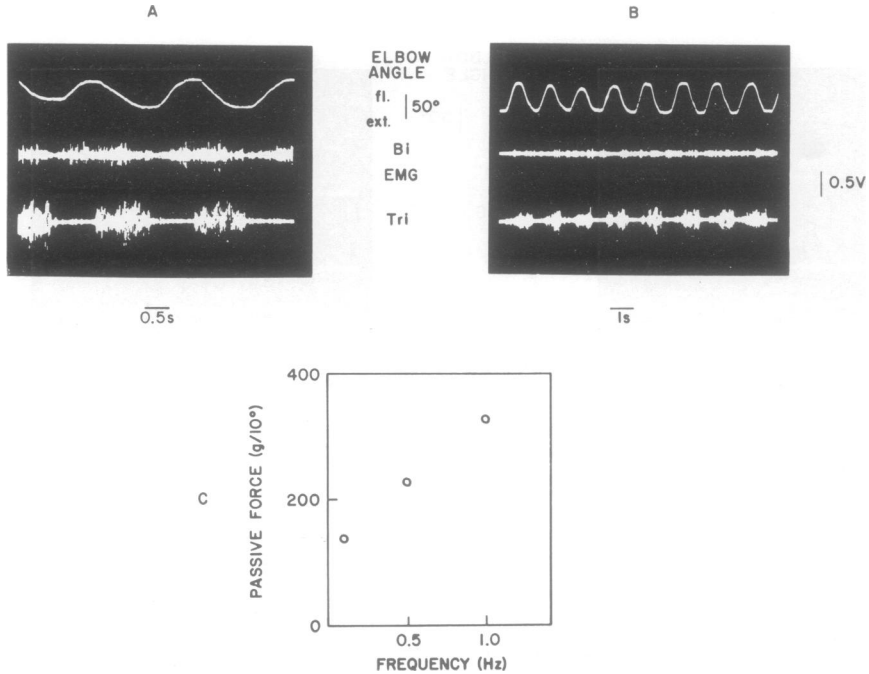


Fig. 5 Patient TP. (A) Voluntary, (B) passive flexion/extension of elbow joint. Top traces are elbow joint angles, second and third traces are the biceps and triceps EMGs. (C) plot of resistance to passive movements (computed as Force/10° angle) at three frequencies (0.1, 0.5, and 1.0 Hz). Triceps EMGs alternate but the biceps EMGs are not reciprocal, which is normal.

about 3 s; this response could be a reflection of spindle sensitivity to static stretch. At 0.8 Hz, tonic and phasic EMG responses occur suggesting increased excitability of spindle afferent nerves or flexor motoneurons or both. Indeed, F response studies have suggested a relative increase in flexor motoneurone excitability states in some cerebral palsy patients (Fisher and Penn, 1978; Fisher, 1978).

The effect of muscle length changes on reflex responses was tested by varying the knee joint angle and recording the EMG responses from quadriceps and hamstrings during passive flexion/extension (Fig. 6D). Increasing hamstrings muscle length facilitated reflex responses in the hamstrings (flexors); on the other hand in the quadriceps (extensors) a decrease in reflex response with increasing quadriceps muscle length could not be demonstrated. Assuming that these static muscle length changes are detected by spindle secondary endings, the results obtained were not compatible with the known properties of secondary endings.

Finally, test of ankle movements revealed a major deficit in voluntary dorsiflex/plantarflex movements.

CASE 4

This 17 year old youth (SH) was quadriplegic, "spastic," and unable to walk.

Upper limb movements

The patient could flex and extend voluntarily his elbow without difficulty. This was demonstrated by the continuous sinusoidal pattern of the angle trace in Fig. 7A. The biceps EMG activity had the normal alternating pattern during flexion only. The triceps EMG, however, was not reciprocally related, and periods of triceps EMG activity during flexion were seen. This was the reverse of what was observed in cases 2 and 3 (Figs. 4 and 5).

The computed resistance to passive flexion/extension at 0.5 Hz was 280 g/10°. This value was greater than the normal values of 150–180 g/10°, indicating an increased resistance to passive movement. This result further supports the earlier suggestion that this type of hypertonia may not necessarily impair voluntary movement if one of the pair of flexor and extensor muscles involved in the movement retained a normal alternating pattern of activation during contraction and re-

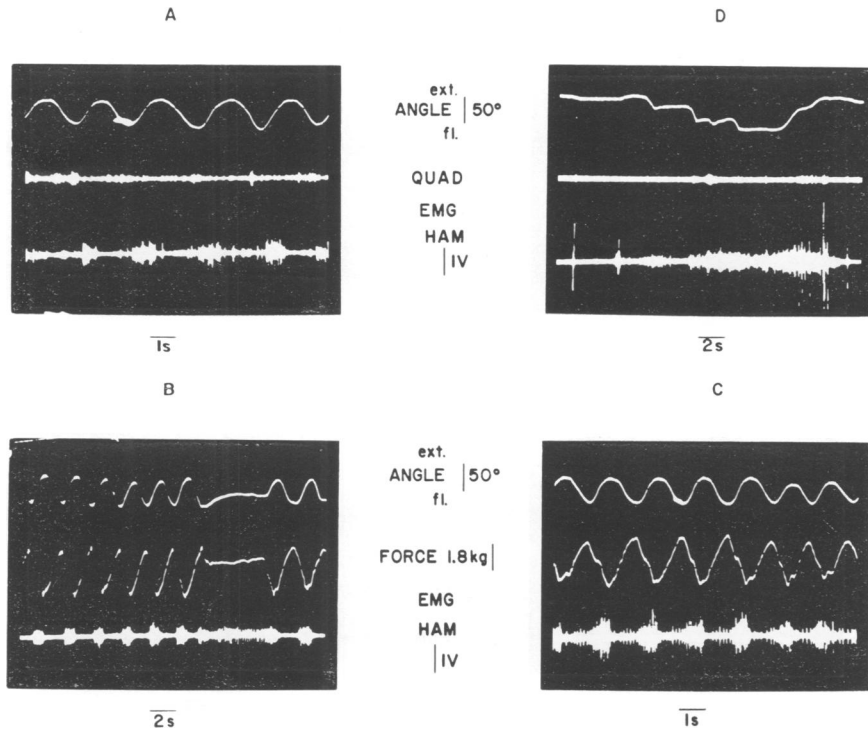


Fig. 6 Case 3, TP. Voluntary flexion/extension of knee joint (A). Passive flexion/extension of knee joint at 0.5 Hz (B) and 0.8 Hz (C). (D) Passive knee flexion/extension at different angles (muscle lengths). In (A) and (D) top trace, angle; middle trace, quadriceps EMG; bottom trace, hamstrings EMG. (B) and (C) top trace, angle; middle trace, force; bottom trace, hamstrings EMG. In A, quadriceps EMG is less prominent and not reciprocally related to hamstrings EMG. B and C demonstrate the sensitivity of the muscle spindles to dynamic and static stretch. D illustrates facilitation of hamstrings reflex responses with increasing muscle length; the quadriceps responses may reflect an abnormality.

laxation. This observation was made in five other patients.

Lower limb movements

The knee joint angle, quadriceps, and hamstrings EMG are illustrated in Fig. 7C. Little EMG activity was recorded from the quadriceps; on the other hand the hamstring EMG was continuous and prominent with irregular periods of large phasic bursts.

A highly significant increase in resistance to passive movement was found. The force required to move the knee joint through 10° was 1.8 kg, compared to average normal values of $700 \text{ g}/10^\circ$. The effect of muscle length changes on reflexes was similar to case 3 (Fig. 6D).

CASE 5

This 10 year old boy (RS) was "spastic," quadriplegic, and unable to walk.

Upper limb movements

The patient could flex/extend his elbow voluntarily without difficulty. There was alternating increased EMG activity in biceps during flexion with background activity during extension. The triceps was silent at low frequencies (0.2 Hz). The resistance to passive movement at 0.5 Hz was increased to $360 \text{ g}/10^\circ$ (normal $150\text{--}180 \text{ g}/10^\circ$). The computed forces were linearly related to frequency of movement.

Lower limb movements

He could flex/extend his knee joint voluntarily through only 20° and at a slow rate (0.1 Hz). Synchronous reflex EMG activity occurred in both quadriceps (extensors) and hamstrings (flexors) during passive movements (Fig. 8A and B). Both flexors and extensors showed reflex responses to stretch of the flexors. This reflex response pattern was observed in seven other patients.

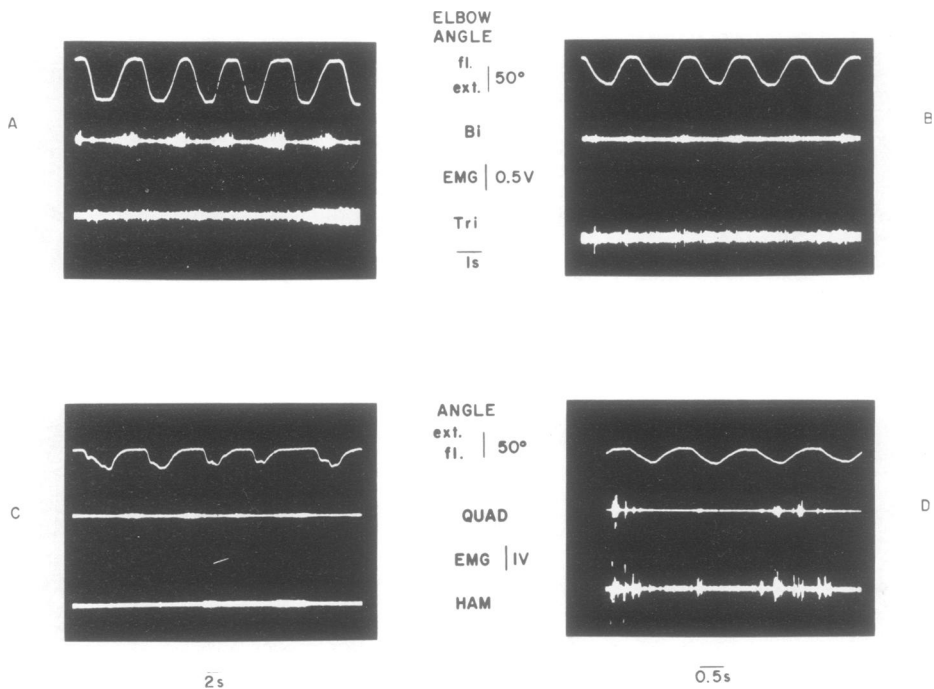


Fig. 7 Patient SH (A) voluntary, (B) passive flexion/extension of elbow joint, showing from the top trace downwards, elbow joint angle, biceps, and triceps EMGs. (C) voluntary, (D) passive flexion/extension of knee joint, showing from the top trace downwards, knee joint angle, quadriceps, and hamstrings EMGs. The synchronous phasic bursts of EMG recorded from the hamstrings and quadriceps during passive movements at 1.0 Hz (D) was the most distinct abnormal feature, in addition to the continuous triceps EMG activity.

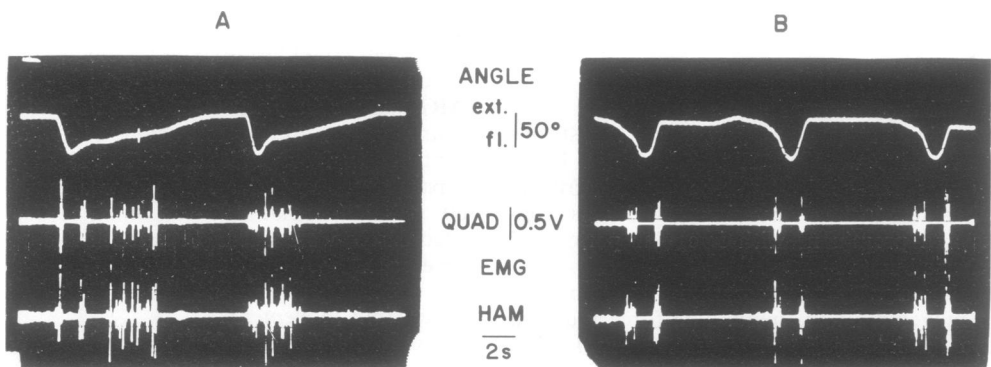


Fig. 8 Case 5, RS. A comparison between reflex responses of the flexors (hamstrings) and extensors (quadriceps) of the knee. The knee joint was continuously flexed and extended through an angle of 50° (upper trace); surface EMGs were recorded from quadriceps and hamstrings. A and B frequencies approximately 0.1 Hz and 0.5 Hz respectively. Note the synchronous EMG responses recorded from quadriceps and hamstrings.

When passive knee flexion was carried out slowly over a period of time (10 s), there was a sudden "give" (clasp knife reaction). This reaction occurred during both passive and voluntary flexion, and the patient was quite aware of his ability to bend his knees after a delay period.

CASE 6

This 27 year old woman (LG) was "spastic," quadriplegic, and unable to walk.

Upper limb movements

Figure 9A demonstrates the pattern of EMG activity in the biceps and triceps during voluntary flexion/extension of the elbow. The continuous EMG activity in the biceps during both flexion/extension was abnormal. The alternating EMG activity in the triceps appeared normal. However, at that slow rate of movement (0.4 Hz) the EMG activity was more prominent than usual and suggested a decreased threshold and increased reactivity of extensor motoneurons.

There was a decrease in the threshold frequency for eliciting a reflex EMG response to passive movements. The threshold frequency was 0.1 Hz for triceps and 0.2 Hz for biceps. The resistance to passive movements increased with increasing frequency as in normal subjects, but again the force computed was more than twice as large as normal. The increased resistance was in part the result of the increased reactivity of both flexor and extensor motoneurons, indicated by the EMG responses (Fig. 9B). In spite of the increased resistance to passive movements, the patient could flex and extend her elbow voluntarily continuously and smoothly at 0.5 Hz. This further strengthens the earlier suggestion that

hypertonia may not always impair voluntary movements.

Discussion

Several pathophysiological mechanisms were evident in these studies.

HYPERTONIA

Almost all patients demonstrated an increased resistance to passive flexion/extension movements which increased linearly with frequency of movement. Therefore, even if there was a pathological increase in fusimotor activity, the normal dynamic sensitivity of the primary endings of muscle spindles to phasic stretch was not disrupted. Indeed, in a study in which ramp stretches were used to test the sensitivity of spindles in the calf muscles, no significant difference in dynamic spindle sensitivity were observed in "spastic" as compared with normal subjects (Hagbarth *et al.*, 1973). Hence, any hyperactive stretch reflexes might be attributed to increased reactivity of spinal neurones. Although hypertonia was a common feature, the variability in the pattern of reflex EMG would suggest diversity in the underlying pathophysiological mechanisms. In most patients a threshold frequency could be defined below which no reflex EMG activity was produced as previously observed by Burke and Lance (1973). The threshold frequency varied from 0.1 Hz to 1.0 Hz in different patients, and in the same patient there were differences between upper and lower limbs and between pairs of flexor/extensor muscles. The reflex EMGs recorded from pairs of flexor/extensor muscles during passive movements also varied from completely reciprocal, through partial

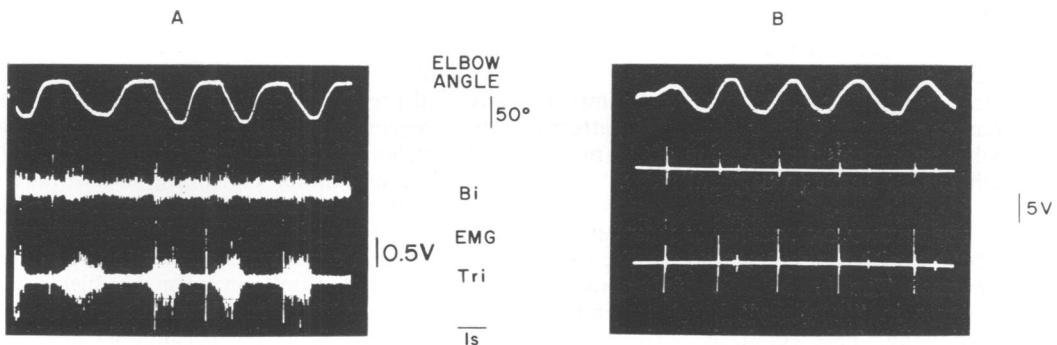


Fig. 9 Patient LG: (A) voluntary (B) passive flexion/extension of the elbow joint. Upper traces, elbow angle; middle and bottom traces, biceps and triceps EMG respectively. A, at the frequency of 0.4 Hz the EMG activity from the triceps was prominent. B, EMGs reflect increased reactivity of biceps/triceps motoneurons, as well as synchronous reflex activation.

overlap, to complete synchrony. The synchronous EMG activity frequently seen suggests an abnormal situation in which the segmental reflex pathways of a pair of flexor/extensor muscles are "wired up" together. This observation has been made in triceps surae and anterior tibial muscles during sinusoidal oscillations of the ankle at 3–12 Hz, in a similar group of cerebral palsy patients (Gottlieb *et al.*, 1978). The cause of this abnormality is not clear. Possibly it is due to an abnormal synaptic action at the interneuronal and/or motoneuronal level because of abnormal descending inputs from the pyramidal system and brainstem centres, or abnormal spinal neuronal development as a result of injury at birth.

VOLUNTARY MOVEMENTS

The deficiency in voluntary motor capabilities varied considerably. The simple test of voluntary flexion/extension movements used in evaluating the level of voluntary function produced a number of diverse abnormal patterns of electrical activity in the muscles involved. Firstly, in the mildly involved limbs, normal alternating electrical activity was maintained in either flexor or extensor muscles. However, the corresponding antagonist muscle was often electrically active throughout the movements and at all frequencies. Secondly, in five patients the voluntary flexion/extension movements were performed at low frequencies (0.1 Hz) without apparent difficulty, and reciprocal EMG activity was maintained in the pair of flexor/extensor muscles being used. As the frequency of movement increased, there was a variable threshold frequency at which a complete breakdown in reciprocal innervation occurred, and co-contractions appeared in the pair of muscles involved in the movement. The overlapping contractions impaired movement. Thirdly, in six severely involved patients there was simultaneous EMG activity in pairs of flexor/extensor muscles at the initiation of movement which resulted in co-contractions of these muscles, severely impairing movements. These diverse abnormal patterns are probably only a small proportion of the motor abnormalities that occur in cerebral palsy.

EFFECT OF HYPERTONIA ON VOLUNTARY MOVEMENT

Do the above positive symptoms create the negative symptoms? The importance of hyperactive stretch reflexes in "spasticity" has been emphasised traditionally. This emphasis derives partly from the enormous physiological work on reflexes dating back to Sherrington, and partly on their usefulness in diagnosis. This study has shown the potentially complex, abnormal, segmental mech-

anisms that may be operating, and also indicated that in certain situations the resulting hypertonia may not necessarily impair smooth voluntary movements. In fact, this conclusion was reached in a study in which the relationship between stretch reflexes and voluntary movement was examined in patients with cerebrovascular lesions (Sahrmann and Norton, 1977). These observations might also explain partly why reduction of positive symptoms through medication did not improve the negative symptoms that were the prime concern of the patients (Landau *et al.*, 1960; McLellan, 1977).

The possible relationship between the abnormal segmental mechanisms and the voluntary motor deficit should depend on the pathophysiological mechanisms inhibiting voluntary movement. The mechanisms will in turn depend on the location and the extent of lesions. An important factor found in this study was the frequency dependence of the pathophysiological mechanisms. If the threshold frequency for producing co-contractions in a pair of flexor/extensor muscles during voluntary movements was comparable to the threshold frequency for eliciting synchronous reflex EMG activity in the same muscles, the hyperactive segmental reflexes would further inhibit voluntary movement. This could have occurred in some of the patients, particularly those with dystonia. Since lesions in the basal ganglia result in co-contractions of pairs of flexor/extensor muscles (Denny-Brown, 1962; 1968; Yanagisawa and Goto, 1971; Brooks, 1975), one would expect such lesions to be all the more disabling when segmented reflexes are hyperactive.

BASAL GANGLIA AND VOLUNTARY MOVEMENT

Motor unit potentials recorded from patients with dystonic movements showed a delay in recruitment and discharged intermittently at high frequencies up to 120/s. The delay in motor unit recruitment and intermittent firing have been observed previously only in Parkinsonism patients where nigrostriatal lesions are dominant (Petajan and Jarcho, 1975; Milner-Brown *et al.*, 1979). This abnormal motor unit firing pattern may well be characteristic of lesions of the basal ganglia, specifically the striatum. The high frequency discharges are in some ways similar to the discharge pattern of motor units in normal subjects during "ballistic" contractions (Desmedt and Godaux, 1977). Since motor units recruited during ballistic contractions have a lower force threshold compared to slow continuous activity, this may explain why dystonic movements (which are ballistic) impair smooth voluntary movements. The diffi-

culty which such patients have in discharging motor units in a normal continuous pattern at 8–20/s (Milner-Brown *et al.*, 1973a, b) necessary for producing slow smooth movements might be explained further by considering Kornhuber's suggested function of the basal ganglia. According to Kornhuber (1974), "the basal ganglia serves as a ramp generator for slow voluntary smooth movements." Hence, lesions in the basal ganglia, particularly involving the striatum (DeLong, 1974), would severely affect slow controlled movements compared with ballistic movements. Consequently ballistic movements—for example, dystonia—will result even when slow continuous movements are desired. During dystonic movements synchronous activations of pairs of flexor/extensor muscles occur and are potentiated by hyperactive segmental reflex mechanisms.

We may conclude from this study that a number of complex pathophysiological mechanisms are involved in cerebral palsy. When chronic cerebellar stimulation is used to try to improve motor function in these patients, such simple physiological tests are useful in delineating some of the pathophysiological mechanisms before cerebellar implant, and also in evaluating any physiological changes that may occur after chronic cerebellar stimulation. The variability in response to stimulation may relate to the specific pathology of the motor system, and by these tests we may be able to identify the patients who will respond best.

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