Short reports

Physiological observations in Sydenham's chorea

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SUMMARY Involuntary movements in a patient with Sydenham's chorea were usually characterised by bursts of EMG activity lasting more than 100 ms, occurring asynchronously in antagonist muscles. The "hung-up" knee jerk was a choreic movement induced by the tendon tap. Exaggerated voluntary movements were produced with EMG burst lasting longer than normal.

With recent interest in the pharmacology of involuntary movement disorders physiology has often been left behind. This has occasionally led to the clinical equation of superficially similar states and apparently confusing pharmacological results. The involuntary movement disorder described by Sydenham was the first to be labelled chorea, but since then this term has been applied to several other disorders, the most prominent of which is Huntington's disease. Although Huntington¹ believed the involuntary movements in the patients he described to be identical to those in Sydenham's chorea, keen clinical observers such as Denny-Brown² have recognised differences. A recent patient with Sydenham's chorea has enabled us to make some physiological observations about this disorder, to explain some of the clinical signs and to illustrate some of the differences between the two principal types of chorea.

Case report

A nine-year-old girl presented with a ten month history of behavioural change, one right focal seizure and a three month history of involuntary movements. General physical examination was normal. On neurological examination there were quick irregular involuntary movements involving most parts of the body, rapid exaggerated voluntary movements, "hung-up" tendon reflexes, occasional postural lapses and hypotonia. CSF

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examination, CT scan and ceruloplasmin level were normal. ASLO titre was 500 TU (high value is more than 400 TU). EEG showed generalised slowing with predominant frequency of 6-7 Hz. The patient was placed on haloperidol (1 mg orally twice daily) and prednisone (20 mg orally twice daily) and showed a gradual clinical improvement over several weeks.

Physiological observations

The patient was studied a few days after the initiation of haloperidol therapy, before any clinical effect was noted. EMG with surface electrodes was recorded from muscles of both legs. When the patient sat quietly, there were only rare involuntary movements. The choreic movements in the legs were brought out by attempted voluntary movements of even distant body parts. The EMG correlate of the involuntary movements were bursts of activity usually lasting from more than 100 ms to more than one second (fig 1 (a)). In over 3 minutes of recording there were only rare bursts of activity lasting less than 100 ms and these almost always had low amplitude making quantification difficult. The bursts in antagonist muscles were often asynchronous and usually reciprocal. There was occasionally somewhat rhythmic alternating reciprocal activity in the antagonist muscles at a frequency of about 3 Hz. While recording from muscles in the right leg repetitive taps were delivered to the quadriceps tendon with a tendon hammer. With interstimulus intervals of one to several seconds, the monosynaptic reflex had constant amplitude and the knee did not "hang up". With shorter interstimulus intervals of 200-300 ms, the monosynaptic reflex had again a constant amplitude, but choreic activity was also produced which caused the knee to hang up (fig 1 (b)). This choreic activity was first seen in quadriceps muscle as a short burst of excessive activity late in the interstimulus interval. As the series of taps continued, this excessive activity became larger in amplitude, more sustained and began to appear in other muscles of the leg. When the

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taps ceased, or at times even if the taps continued, the choreic activity suddenly stopped in all muscles simultaneously and the leg fell under the force of gravity and would oscillate passively a few times. Similar choreic activity could not be produced with taps to the Achilles tendon.

Voluntary movements were studied by asking the patient to make stereotyped ballistic elbow flexion movements with methods that have been previously described in detail.³ The patient's right arm was strapped into a light splint with a low-friction joint at the elbow. With a potentiometer the angle of the elbow was converted to a voltage which was both recorded and displayed to the subject as the height of a line on an oscilloscope. The patient was asked to make as rapidly as possible a 20° movement, corresponding to a movement of the line on the oscilloscope from one place to another. Surface EMG was recorded from biceps and triceps. Normally such movements are initiated with a burst of activity in biceps lasting less than 100 ms, followed by a burst of activity in triceps, also lasting less than 100 ms. The patient's voluntary movements were characterised by

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Figure (a) EMG correlates of involuntary movements. During A, the patient lifted her left leg. During B, the patient sat up. In C, the patient was sitting and quickly extended the left knee. (b) EMG correlates of the "hung-up" knee jerk. The quadriceps tendon was tapped repetitively and produced the regular brief EMG burst seen in the quadriceps and hamstrings. Choreic activity begins first in quadriceps, becomes progressively larger in amplitude, spreads to other muscles and then suddenly ceases. (c) EMG activity during three ballistic elbow flexion movements. On verbal command the patient attempted to make 20° movements as rapidly as possible.

bursts of EMG activity with prolonged duration (fig 1 (c)). In part A, the first biceps burst is very prolonged and there is no initial triceps burst. In parts B and C, the more typical circumstance is illustrated; both initial bursts are present and they are both prolonged. In eleven attempted movements the first biceps burst averaged 139 ms (standard deviation 58 ms) with a range of 100-300 ms. The first triceps burst averaged 85 ms (standard deviation 48 ms) with a range of 0-150 ms and six of the eleven bursts lasting 100 ms or more.

Discussion

Simple EMG characterisation of involuntary movements is a useful first step in physiological classification. The value of this is illustrated in previous studies of myoclonus where different disorders are characterised by different EMG burst durations (as well as other distinguishing electro-physiological features). Reflex myoclonus (both reticular reflex and cortical reflex) is characterised by a burst length of

10-30 ms; ballistic movement overflow myoclonus is characterised by a burst length of 50-100 ms; and dystonic myoclonus is characterised by a burst length of more than 100 ms.⁴⁻⁶

Initial EMG characterisations of the involuntary movements in Sydenham's chorea were accomplished by Hocfer and Putnam.⁷ They showed EMG bursts asynchronous in antagonist muscles lacking reciprocal inhibition and suggested that the activity was similar to that seen in athetosis. Herz⁸ had similar findings and pointed out that in any one involuntary movement there was a tendency for one muscle of an antagonist pair to be more active than the other. Herz also commented on the variability of EMG burst durations. More recently, Rondot⁹ has again illustrated the asynchrony of antagonist muscles. The findings here confirm previous investigations and make two further points: (1) there are only rare involuntary movements with EMG burst duration less than 100 ms. This is in contrast to several patients with Huntington's chorea studied with similar techniques where more frequent bursts of 10-30 ms (similar to reflex myoclonus) and 50-100 ms (similar to ballistic movement overflow myoclonus) duration have been noted (ref 5 and unpublished observations), (2) attempted voluntary movement seems crucial for the production of involuntary movements in Sydenham's chorea where this seems less important, although still a feature, in Huntington's chorea. Another difference between Sydenham's chorea and Huntington's chorea is the tendency for synchrony of antagonist muscles in the latter disorder.⁹ It will be necessary to develop more extensive quantitative methods for analysis of involuntary EMG activity for unequivocal proof of differences between the two types of choreic movement.

The hung-up reflex in Sydenham's chorea has been a well recognised clinical sign, but there does not appear to have been any previous study of its mechanism. The findings here suggest that this phenomenon is due to a choreic movement induced by the tendon tap. It begins as a brief, localised event in the muscle stimulated and then extends both temporally and spatially. The initial involuntary movement burst has a latency (after the tendon tap) more than any of the known long-latency reflexes and hence there is no immediate insight into any possible physiologic mechanism. This phenomenon has not been described in Huntington's disease.

Ballistic voluntary movements are useful in the quantitative analysis of voluntary movement disorders. Normal persons attempting to move as rapidly as possible from one place to another usually generate a triphasic pattern with successive bursts in the agonist, the antagonist and then the agonist

again. Burst duration of the first agonist and first antagonist burst is relatively fixed at about 50-100 ms, regardless of the distance moved.¹⁰ The role of the first agonist burst is to set the limb in motion, and a role of the first antagonist burst is to help stop the limb at its endpoint. The fact that our patient produced long first agonist bursts would seem to predispose her to make excessively long movements. This would be particularly true to the extent that the braking activity of the first antagonist component is lacking. Bursts of prolonged duration have also been seen with cerebellar lesions,¹¹ where the bursts seem also to predispose to excessive movement, with pyramidal tract lesions,¹² where prolongation seems to be a compensatory mechanism for weakness, and in patients with athetosis,¹³ where the burst is at times helpful to compensate for an overactive antagonist. Ballistic movements have not been studied in Huntington's disease, so it is not clear whether this phenomenon is a distinguishing electrophysiological feature.

Before becoming too dogmatic about the results, the studies should be repeated on another patient, but such patients are uncommon as the disease seems to be disappearing.¹⁴ It does appear, however, that despite some superficial clinical similarities, Sydenham's chorea and Huntington's disease are different movement disorders. Chorea is not a single entity and cannot be treated as such for either animal models or pharmacological trials.

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