IMPAIRMENT OF RAPID MOVEMENT IN HUNTINGTON’S DISEASE

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SUMMARY

Patients with Huntington’s disease (HD) and relatives at risk were examined with respect to their capacity to produce rapid voluntary motor activity. For this purpose, the fastest possible self-paced single isometric forefinger extensions and the fastest alternating forefinger movements were tested. In addition to these fastest voluntary performances, the time course of spontaneous hyperkinetic finger movements and the peak frequency of finger and hand tremor were analysed as a measure of the temporal characteristics of involuntary movements. Comparison of these parameters in HD patients and individuals at risk with age and sex-matched normal controls revealed a significant slowing of all types of contractions or movements in the majority (up to 95%) of the patients and in up to 40% of the relatives at risk. Reaction times were only slightly prolonged, and the abnormalities of the movement parameters showed no correlation with detailed psychometric data. Hence it is unlikely that the disturbance in the execution of rapid motor acts is due to dementia. Tremor was also slower than normal and the hyperkinesias were still slower than the fastest voluntary contractions. It appears from this study that slowness of motor performance is not only evident in Parkinson’s disease but may represent a more general dysfunction in basal ganglia disease.

INTRODUCTION

Huntington’s (HD) and Parkinson’s disease (PD) result from selective involvement of different subpopulations of striatal neurons: the dopaminergic nigrostriatal system in PD and the cholinergic intrinsic and gabaergic output neurons in HD. In spite of the affection of such closely linked circuitries the two disorders are characterized by opposite clinical features: one is hyperkinetic and the other hypokinetic. Despite these different clinical hallmarks the two basal ganglia disorders also have common aspects. Reports describing rigidity and akinesia in rare Westphal variants of HD with early juvenile onset (Westphal, 1883) or in the late stages of the disease (Bonhoeffer, 1936; Denny-Brown, 1960; Campbell et al., 1961) reveal that the clinical features typically seen in PD may sometimes also occur in HD. Besides these rare variants, Hamilton (1908), in examining 27 cases with HD, emphasized the slowness of voluntary movements, the presence of plastic rigidity.
and a flexed posture resembling PD in many of his patients. Herz (1931), in his pioneering monograph in which he introduced frame-by-frame cinematographic analysis for the description of involuntary movements, described clear abnormalities of voluntary motor activity in his patients with HD. He emphasized especially the difficulty of movement initiation, the slowness and irregularity of movement execution and the disharmonic coordination of different body parts resembling 'asynergy'. Bittenbender and Quadfasel (1962) in their review of the literature stated that 'the frequency of occurrence of the rigid form of HD is greater than generally appreciated'.

Various aspects of disturbed motor activity in HD have been revealed by neurophysiological studies. Analysis of eye movements (André-Thomas et al., 1945; Starr, 1967; Avanzini et al., 1979; Oepen et al., 1981; Leopold et al., 1982; Leigh et al., 1983) demonstrated slowness of saccades and a disturbance of smooth pursuit caused by inappropriate intrusion of saccades. Other studies in patients with HD have shown abolition of long-latency stretch reflexes (Noth et al., 1983, 1985), alteration of physiological tremor (Myers and Falek, 1979) and an impairment of postural stability (Valade et al., 1984). Only a few studies have considered voluntary motor control in HD. Petajan et al. (1979) described impairment of voluntary control of individual motor units and Starr (1967) presented one example of impairment of fast voluntary limb movements in HD in a paper otherwise concentrating on oculomotor disturbances. Recently Koller and Trimble (1985) described alterations of gait pattern in HD.

Quantitative analysis has so far not clarified whether slowing of movement is a consistent phenomenon in HD or a rare abnormality that appears only in a few variants. This may be due to difficulties in quantifying voluntary movements. We therefore selected out of the wide range of possible movements simple motor tasks which are limited by the properties of the system itself or follow invariant principles. We have analysed two types of rapid voluntary motor performances: the fastest possible single isometric contractions and the fastest alternating movements. For comparison, two forms of rapid involuntary movements, the hyperkinesias and tremor, were also measured. All recordings were made from the index finger of the dominant hand in patients with HD, in their offspring at risk of developing the disease later in life and in an age and sex-matched control group.

**METHODS**

**Patients**

A total of 22 patients with a definite diagnosis of HD were examined. Severity of the disorder in these patients was scored according to a modified version (Lange et al., 1983) of the Shoulson-Fahn Score (Shoulson and Fahn, 1979) referred to as DIS. Table 1 gives detailed clinical data of the Huntington's disease (HD) patients in the study: age, sex, duration of choreic symptoms (DUR), severity of choreic symptoms (CHOR), disability score (DIS) and drugs. More than half of the patients (13/22) did not take any drugs, 4 were taking only substituted benzenamides such as sulperide or tiapride with dopamine receptor blocking properties but not causing extrapyramidal symptoms; 2 had a
DUR = duration of choreic movements (yrs). CHOR = severity of chorea during clinical investigation ranging from 0 to 3 (Lange et al., 1983). DIS = disability score (Shoulson and Fahn 1979; Lange et al., 1983; Lange et al., 1984). BU = butyrophenones. SU = sulpiride. PH = phenothiazines. IN = isoniazid. TI = tiapride. BZ = benzodiazepines. BA = baclofen.

Forty subjects at risk were examined, all of whom were first generation offspring of patients with definite HD. Standard neurological examination in these individuals was always normal except for the presence of indefinite ('soft') clinical signs (similar to those defined by Young et al., 1986; e.g., slowing of fast hand pronation and supination or finger flexion-extension movements) in 9 at risk subjects. None of them had any evidence of choreiform movements when entering the study. This was assessed by a careful search for the presence of involuntary movements with the at risk patients being asked to remain as still as possible while carrying out distraction tasks such as repetitive subtraction of 7 from 100 or reciting the names of the months backwards. In the 9 subjects at risk for HD with indefinite signs this search revealed some increase in restlessness above the normal values but no hyperkinesia. None of the at risk subjects was taking any drug which might interfere with motor activity except for 1 patient with a high alcohol intake. All at risk subjects with 'soft' signs are marked by special symbols in the.
figures. We also studied 1 patient with the juvenile rigid ‘Westphal’ variant of HD and 1 patient with Sydenham’s chorea. Clinical data of these patients will be given in the Results section.

A group of 26 normal volunteers recruited from the hospital staff and from outpatients at our institution with vascular headaches, compressive radiculopathy of the lower limbs or minor nonneurological disease served as controls. Controls were matched by age and sex with the patients and the at risk group: each patient was matched for age and sex by a single control subject but the same control subject could match multiple at risk subjects.

Experimental Procedures and Data Analysis

Analysis of tremor. A light-weight Philips type (PR 9366 E/20) accelerometer was taped to the distal phalanx of the index finger of the dominant hand. Patients were seated in a reclining chair and asked to keep their hands outstretched at 90 deg anteversion of the shoulder with the elbow kept straight, the hand pronated and the fingers stretched out horizontally. The ongoing tremor recorded by the accelerometer for a period of 25 s was fed into a Nicolet (Med-80) computer and sampled at a frequency of 80 Hz. The resulting signal was stored on magnetic discs for off-line data analysis. The total period was subdivided into 8 segments and for each of these segments a power spectral density function was computed. The 8 resulting spectra were averaged. From this average spectrum the peak frequency was determined. Series of partially overlapping power spectra were plotted to visualize the time course of variation of peak frequency (for further details regarding filtering, see Homberg et al., 1987).

Analysis of fastest alternating movements. For analysis of fastest alternating index finger movements the accelerometer remained at the distal phalanx of the index finger as described in the preceding section. Subjects were instructed to keep the forefinger outstretched with the wrist stabilized by the contralateral hand, the forearm semipronated and the elbow flexed at 90 deg. In one or two warm-up trials they were trained by the experimenters to perform fastest possible flexion-extension movements at the metacarpophalangeal joint with the index finger kept outstretched. Between the warm-up trials, pauses of at least 2 min were provided to avoid muscular fatigue. The resulting accelerometer signal was digitized as described above at a sampling frequency of 60 Hz. A total recording period of 34 s was subdivided into 16 segments. For each segment the power spectrum density-function was calculated and its peak frequency determined. The maximum of these 16 frequencies was defined as the frequency of maximal voluntary alternating movements. In addition consecutive partly overlapping segments of 4.2 s duration were plotted in the form of hidden line plots to visualize dynamic changes of alternating frequencies.

Fastest isometric voluntary index finger extensions. Subjects and patients were seated in the reclining chair as before. The forearm was fixed in full pronation on a flat support. The index finger was attached to the axis of a bidirectionally sensitive Schaewitz type FTD force transducer by means of a concentric ring adjustable in diameter to individual finger size at the proximal interphalangeal joint. To avoid movements around the wrist, leather straps were attached across the middorsum of the hand, just proximal to the wrist joint and at positions 15 and 30 cm more proximally in the forearm. Bipolar surface EMG electrodes were fixed over the extensor indicis and the flexor digitorum superficialis muscles. Optimal location of electrodes was achieved by obtaining a maximal signal amplitude in both flexion and extension at a given constant force and by ensuring that slowly alternating flexion-extension of the metacarpophalangeal joint resulted in reciprocal activity in agonist and antagonist.

Subjects were instructed to press their index finger against the force transducer as quickly as possible after the occurrence of a 1000 Hz tone of 50 ms duration. The amplitudes of the contractions performed in this simple reaction time paradigm could be chosen deliberately by the subjects, but it was emphasized that the amplitudes were varied from trial to trial, so that a broad distribution of amplitudes was produced. The force signal and the rectified demodulated surface EMG were sampled at a rate of 1 kHz for trial epochs of 1.024 s duration starting 50 ms before stimulus onset, and stored on a magnetic disc; 20 to 30 trials were recorded in every subject.

From the force records the following parameters were extracted by means of an interactive cursor
display: (1) reaction time (RT), i.e., the time between tone onset and start of the contraction; (2) contraction time (CT), i.e., the time between start and peak point of the contraction; and (3) contraction amplitude (AM). The remainder of the data analysis will be explained in the Results section.

Recordings of involuntary isometric contractions. In some HD patients the hyperkinetic movements of the fingers occurred with such large amplitudes that they could be compared with voluntary single contractions. For this purpose the patients remained coupled to the force transducer in the same way as described above for the isometric recordings and were told not to move voluntarily and to relax completely. Hyperkinetic fluctuations in isometric force were recorded continuously for periods of several minutes. Since the force transducer used was bidirectionally sensitive, both flexion and extension forces of the hyperkinetic movements could be recorded. From the data stored on discs these involuntary force changes were analysed off-line in the same way as described for rapid isometric contractions in the preceding section.

Other EMG recordings. In some HD patients, the EMG activity during fastest isotonic elbow extension movements and during alternating wrist flexion-extension movements of the dominant arm were studied. Surface EMG electrodes were attached over biceps and triceps brachii or flexor carpi ulnaris and extensor carpi radialis muscles, respectively. The patients were instructed to perform the fastest possible elbow extensions after a tone cue (1000 Hz, 50 ms duration, 70 dB SPL) starting from a resting position of 90 deg flexion of the elbow with the forearm fully supinated and the shoulder elevated to 90 deg. Onset of movement was detected by an accelerometer taped to the forearm. For alternating wrist movements the wrist was held in semipronation, the elbow in 90 deg flexed and the forearm resting on a flat support. Patients were told to move their wrist at a frequency of about 2 Hz as regularly as possible.

In 1 HD patient at a very early stage of disease in whom hyperkinesia was still restricted to the toes on the right, the EMG pattern of these hyperkinetic movements was recorded; surface electrodes were fixed over extensor digitorum brevis and flexor digitorum brevis. An accelerometer was taped to the distal dorsal phalanx of the big toe to detect onset of movements. During the recording, the patient was asked to relax completely with both legs resting on a bench.

Psychometry. For all of the HD patients and at risk subjects in this study, detailed psychometric data were obtained within a period of two weeks around the motor studies. The various psychometric procedures have been described in more detail before (Lange et al., 1983; Hömberg et al., 1986). Briefly the following psychometric tests were performed:

1. The WIP score (Dahl, 1972). This is a short condensed intelligence score encompassing the 'information', 'similarities', 'picture completion' and 'block design' subtests of the German version of the WAIS.
2. The German version of the Raven standard progressive matrices (Raven, 1938; Kratzmeier and Horn, 1980). This test was used to assess reasoning and concept formation independent from language-related abilities and time pressure. In essence this test requires that incomplete visually-presented patterns be completed by selection of 1 of 6 or 8 stimuli. The tasks are ordered in increasing difficulty, with 60 trials presented in 5 blocks of 12 items.
3. The multiple choice-vocabulary test (MWT-B, Lehrl, 1977). This has been shown to be a reliable indicator of the premorbid level of verbal intelligence (Merz et al., 1975). The test requires selection of 1 proper word out of groups of 5 words which always contain 4 nonsense words; 37 of these word groups rank ordered in increasing difficulty are used.
4. The Syndrom-Kurz Test (SKT) (Erzigkeit, 1977). This is a sensitive indicator of impairment of memory, attention and concentration of patients with cognitive impairment. It consists of naming, reproduction of visually-presented material after various time intervals freely or out of a second list of items, ordering of numbers, counting of symbol categories and an interference task where all instances of the letter A have to be read aloud as B and vice versa. This measures 'disposition rigidity' and vulnerability to distraction. All subtests imply time pressure.
Statistical analysis. For group comparisons conventional Student's t tests for uncorrelated means were applied after testing for homogeneity of variances using the F-max test. The relationships between various motor parameters age, psychometry or disability scores were assessed using linear regression analysis and Pearson product-moment correlation coefficients. The normal range for age-dependent movement parameters was defined by 2 SD above and below the age-related regression line for each parameter in the normal population. Covariation ellipses were calculated from the Ciba Geigy Wissenschaftliche Tabellen (Statistik), pp. 219-221.

RESULTS

Most Rapid Single Isometric Index Finger Contractions

Contraction times. The fastest voluntary single index finger extensions are prolonged in patients with HD, as can be seen even in single trials. Fig. 1 shows a single trial of the reaction time paradigm for fastest possible voluntary isometric index finger extensions in a normal subject and an HD patient. In the normal subject a smooth contraction curve results (fig. 1, second row, left side). The extensor (fig. 1, third row) and the flexor EMG (fig. 1, bottom row) both show a single burst with the flexor already being coactivated about 20 to 30 ms after the extensor. This coactivation is typical for rapid isometric contractions (Freund and Büdingen, 1978; Sanes and Jennings, 1984). In the HD patient (fig. 1, right side) the contraction curves show a marked prolongation before peak force is reached. This is caused by a slowing of the corresponding build up of EMG activity. The coactivation pattern of agonist and antagonist is, however, maintained. Similarly the normal 'triphasic' pattern (Wachholder and Altenburger, 1926; Hallett et al., 1975) for fastest isotonic

![Fig. 1. A single most rapid index finger extension in a normal subject (left side) and in a patient with HD (right side). In the normal subject the contraction starts 120-170 ms (= reaction time, RT) after the tone signal and reaches peak amplitude in about 100 ms (contraction time, CT). The EMG shows cocontraction of agonist (EXT) and antagonist (FLEX) muscles. In the HD patient RT, CT and the EMG burst duration are prolonged. The normal coactivation pattern is maintained. AM = amplitude.](image-url)
elbow extensions and the normal reciprocal pattern for alternating wrist movements are maintained. It therefore appears that the proper selection of muscle groups for these simple types of contractions and movements is not impaired in HD.

The calculations of mean contraction times for the different populations would be the next step of data processing. This is, however, not possible at this stage because, as will be shown, contraction time varies with contraction amplitude in many HD patients. Since the task was to perform the contractions as fast as possible at freely selectable amplitudes, for the comparison of contraction times the variation of contraction amplitudes has to be taken into account. It is therefore necessary to establish the contraction time-amplitude relationship first.

*Relationship between contraction time and amplitude.* In fig. 2 several most rapid contractions of a normal subject (left side) aligned for contraction onset are compared with the most rapid contractions plotted in the same way for 2 typical HD patients representing various stages of the disorder: the example in the middle is from Case 20 (37 yrs; F; CHOR = 1.0; DUR = 3; DIS = 0.4) whereas the example on the right is from the more affected Case 17 (52 yrs; F; CHOR = 2, DUR = 4; DIS = 1.2). In the early stages of HD, contractions are already prolonged. In the normal subject contraction times remain approximately constant irrespective of amplitudes (Freund and Büdingen, 1978). In milder affected cases with HD the variation of the rate of rise of tension may be well preserved and hence contraction time remains independent of amplitude. With further progression of the disease, however, the ability to increase the rate of rise of tension is reduced so that a clear increase of contraction time with amplitude results (fig. 2, right side).

To quantify the impairment in the speed control of single voluntary contractions that normally keeps contraction times almost constant in individual subjects, the relationship between contraction time as the dependent variable (y axis) and
amplitude being the independent variable (x axis) was determined by linear regression analysis of 20 to 30 trials (AM-CT regression). In fig. 3A such a regression line is presented for a normal subject and in 3B for an HD patient (Case 17; cf. fig. 2, right side). The slope of the regression line of the normal subject was close to zero, whereas in the HD patient the slope of the regression line was considerably steeper. Furthermore, the scatter of the single trial values around the AM-CT regression line was much larger in the HD patient than in the normal subject.

Each AM-CT regression line can be described by its slope and one reference point (e.g., the intercept). Since some HD patients had difficulties in performing rapid contractions of small amplitude the contraction time corresponding to an amplitude of 2N (CT2N) was chosen instead of the y intercept as reference value. Thus for each subject and patient the AM-CT relationship is characterized by one pair of values: the y value of the AM-CT regression line at 2N (CT2N) and the slope of the AM-CT regression line (AM-CT slope) (see fig. 3B).

![Fig. 3. Linear regression line analysis between contraction amplitude (abscissa) and contraction time (ordinate). The slope of the regression line (AM-CT slope) is much steeper in an HD patient (B) than in a normal subject (A). In order to display this slope for the different populations examined such regression lines were determined for each subject. They can be expressed by the AM-CT slope and the contraction time at a force level of 2N (CT2N). For the normal subject CT2N = 107 ms, AM-CT slope = 1.7 ms/N; for the HD patient CT2N = 95 ms, AM-CT slope = 24.1 ms/N.](image)

Therefore the AM-CT regression line of each subject can be represented by one point in an x/y plane taking CT2N as the x value and the AM-CT slope as the y value; 95% of the normal values could be encompassed by an ellipse as shown in fig. 4A. When plotting the HD patients against this normal ellipse it appears that all the data points of HD patients lay outside or at the border of this normal range (fig. 4A, triangles on left side). The 2 HD patients at the border of the normal range were only mildly affected (Case 7, 30 yrs, M, DUR = 5; CHOR = 1.5, DIS = 0.3; Case 14, 43 yrs, M, DUR = 5, CHOR = 1.5, DIS = 0.5). Also a high percentage of subjects...
Correlation between the increase of contraction time with amplitude and disability. Adopting a criterion of 9 ms/N as the upper normal limit for the slope of the AM-CT relation (Hömberg et al., 1984) the HD patients could be divided into subgroups with normal and abnormal slopes. The mean disability scores for these two subgroups showed a highly significant difference with a mean score of 0.75 ± 0.65 in the HD patients with normal and 1.52 ± 0.47 in those with abnormal slopes. This indicates that the increase in the AM-CT slope was associated with more advanced disease. This was also reflected in a significant positive correlation between the AM-CT slope and the disability score (fig. 5), (r = 0.47, P < 0.05, two-tailed test).

Contraction time variability. In each subject the scatter of the contraction times around the AM-CT regression line was expressed as the mean value of the absolute distances of individual contraction times from the AM-CT regression line. In normal subjects the mean value of this deviation from the AM-CT relation (CTV) is about 10% of the mean contraction time. The mean value for CTV in normals was 12.8 ± 4 ms. As can be seen in fig. 6A (left side) there is no significant influence of age on CTV in normal subjects (slope = −0.021 ms/yr, r = −0.059, n.s.). In patients at risk for HD lay outside this normal range (crosses in fig. 4B; squares indicate at risk subjects with 'soft' signs).
with HD, however, (fig. 6A, middle) contraction times vary to a much larger extent than in normals: the mean value of CTV in patients with HD was 55 ± 30 ms, which was significantly larger than in normals (P < 0.01). Plotting individual CTV values of HD patients against the normal range revealed only 2 HD patients with a nearly normal scatter of the contraction times around the AM-CT relation. These 2 subjects were already mentioned as having a normal AM-CT relationship. No influence of age on CTV was observed in HD patients (slope = 0.039 ms/yr, r = 0.0128, n.s.). A considerable percentage of people at risk (14/37 at risk subjects) also showed a significantly higher CTV value than the normals (fig. 6A, right side).

![Graph](image)

**Fig. 5.** Relationship between the slope of the amplitude-contraction time (AM-CT) regression line and the disability score. This shows a significant positive correlation, the more affected patients having a larger increase in contraction time with amplitude.

*Standardized contraction time.* The foregoing calculations have established the basis for the comparison of contraction times between normal subjects and patient groups. Since most of our subjects produced contractions with a mean amplitude of about 5N we selected the point corresponding to the 5N-value of the AM-CT regression line in order to define in each individual a contraction time suitable for intergroup comparisons. This will be referred to as standardized contraction time (SCT). In normal subjects there was no significant effect of age (fig. 6B, left side) (slope = −0.305 ms/yr, r = −0.159, n.s.) on SCT. In HD patients, however, a clear
Fig. 6. A, relationship between variation in mean contraction time (CTV) from the individual contraction time-amplitude regression line and age in normals (left side), HD patients (middle) and individuals at risk (right side); squares indicate those with 'soft' signs. B, relationship between standardized contraction time (SCT, taken from individual contraction time amplitude regression lines at 5N) and age in normals (left side), HD patients (middle) and individuals at risk (right side); squares indicate those with 'soft' signs. C, relationship between mean simple reaction time (RT) and age in normals (left side), HD patients (middle) and individuals at risk (right side); squares indicate those with 'soft' signs.
increase of SCT with increasing age can be observed (slope = 3.12 ms/yr, r = 0.453, P < 0.05) corresponding to the progression of HD with increasing age. The group mean value for SCT in normals was 114 ± 23.7 ms in contrast to a mean value of 217 ± 53.3 ms in HD patients (P < 0.001, t test, two-tailed).

Plotting individual SCT values for the patients against the age-related regression of normals revealed that all of the values of the HD patients lay outside the normal range (fig. 6B, middle). In the at risk group, 8 subjects had abnormal results (fig. 6B, right side). This parameter thus separated the disease group most clearly from normals.

Both slowing of contraction times and increase of contraction time variation seem to be early signs for a deficit in the regulation of rapid single voluntary contractions in HD patients and at risk subjects. The influence of disease duration on these parameters will be discussed later, and is summarized in Table 3.

**Reaction time.** In normal subjects reaction time slightly increases with age (fig. 6c) (slope = 0.545 ms/yr, r = 0.226, n.s.). The mean RT value for the normal population is 163 ± 28.1 ms. In patients with HD the mean RT was 252 ± 74.2 ms and thus significantly larger than the normal value (P < 0.01). Most patients show reaction times in the upper normal range. Reaction times covary with patient age (fig. 6c, middle) (slope = 3.912 ms/yr, r = 0.4990, P < 0.05). There are some patients with normal reaction times even with long-standing disease. But only about one-third of the HD patients show RTs which are prolonged by more than 2 SDs beyond the

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**Fig. 7.** Comparison between voluntary isometric index finger extensions (A) and involuntary hyperkinetic extensions (B) in a 32-yr-old male HD patient with predominantly distal hyperkinetic movements. Involuntary contractions of comparable amplitude to voluntary contractions have even longer contraction times (C). ● = hyperkinetic contractions, + = voluntary contractions.
normal age-RT regression line. Only 6 of the subjects at risk show prolonged reaction times (fig. 6c, left side). It appears, therefore, that abnormal RTs in HD patients and at risk subjects are rarer than the abnormalities in force production.

*Time characteristics of isometrically recorded hyperkinesia.* In some HD patients who presented with marked hyperkinesias restricted to the distal upper limbs, we compared their most rapid voluntary contractions with those resulting from involuntary hyperkinesias. A typical example is shown in fig. 7. Voluntary isometric contraction curves (fig. 7A) aligned at onset are compared with involuntary contractions (fig. 7B) in a 32-yr-old male HD patient (DUR = 11, DIS = 1.5). Most of the involuntary hyperkinetic contractions are slower than the voluntary contractions of comparable amplitudes. In fig. 7C the AM-CT relationship of voluntary (crosses) and involuntary (dots) contractions are shown. The AM-CT slope is steeper for the involuntary than for the voluntary contractions. Comparisons between different patients show that with more pronounced hyperkinesias the contraction times also increase and become more dependent on amplitudes.

The EMG pattern of hyperkinesias are illustrated in a 31-yr-old patient with early HD, who presented with low amplitude irregular hyperkinesias restricted to his right toes. Fig. 8 shows recordings of EMG activity from the short toe extensors (EDB) and flexors (FDB) and an accelerometer trace from the big toe. Using a long

![EMG recordings](image)

**FIG. 8.** Different EMG patterns in the extensor digitorum brevis (EDB) and the flexor digitorum brevis (FDB) muscles in a 31-yr-old male subject during hyperkinetic movements of the right toes. The burst durations vary randomly (A, long time constant). Short myoclonic bursts (B) as well as tonic discharges (C) and complex agonist/antagonist patterns (D) may occur. ACC = accelerometer trace.
time base (A) an irregular sequence of EMG bursts of highly variable duration (40–400 ms) is apparent in both muscles. Most bursts are restricted to the EDB but occasionally ordered sequences with an EDB burst followed by an FDB burst may occur (D), whereas coactivation of EDB and FDB is not observed. Therefore, even in early chorea the observation by Hallett (1983) applies, that in chorea a wide variety of different EMG patterns underlie choreic hyperkinesias ranging from short myoclonus-like bursts (as in B) to long ‘tonic’ discharges (as in C). Occasionally (as in D) ordered sequences of agonist and antagonist can be observed resembling, for example, the ‘triphasic’ pattern characteristic of fastest voluntary isotonic movements.

Observations in other choreic disorders. A young man (29 yrs, DUR = 6, DIS = 2.25) was studied who suffered from the akinetic Westphal variant of HD. His contraction parameters were the most prolonged of all HD patients studied: his

![Graph A](image)

**Fig. 9.** A, most rapid isometric index finger extension of a 29-yr-old HD patient suffering from the akinetic Westphal variant of HD (note the change of the time scale compared with fig. 1). Oscillations are superimposed on his contractions. These oscillations are similar to those observed in parkinsonian patients. They are consistently found both in patients with tremor at rest (example in B) and in akinetic-rigid parkinsonian patients without tremor at rest (example in C). In all 3 cases a 10–13 Hz action tremor can be observed during the contractions. This kind of action tremor present in the Westphal variant of HD is typical for Parkinson's disease and different from the smoothly slowed shape of contractions observed in HD.
SCT (332 ms) rather underestimates the actual amount of slowing because he was unable to produce most rapid contractions of smaller amplitudes (minimal amplitude 6N). His efforts to produce a most rapid small contraction always elicited series of EMG bursts which he was unable to stop (fig. 9A, left side). Recordings of the most rapid single index finger extensions in patients with PD show a similar action tremor superimposed on the voluntary contractions. Two typical examples are shown in fig. 9B and c. This 10 to 13 Hz action tremor can be observed in PD patients with tremor at rest (fig. 9B) as well as in an akinetic-rigid PD patient without tremor at rest (fig. 9C). Voluntary alternating frequencies of the patient with the Westphal variant were also the slowest observed in this study (2.6 Hz). In contrast to other HD patients his postural tremor was clinically apparent. It showed a higher peak frequency (7.7 Hz) and considerably higher amplitudes. Obvious bursts in the EMG suggested that this tremor was due to neural synchronization. The presence of a synchronized postural tremor and the repetitive EMG bursts superimposed on the contraction curves are two features more typical of PD than of HD.

A 30-yr-old female with Sydenham’s chorea due to recurrent streptococcal infections was also studied. All movement parameters showed similar slowing as in HD patients (tremor frequency = 2.8 Hz, voluntary alternating frequency = 3.9 Hz, SCT = 210 ms, DEV = 32 ms, RT = 185 ms ± 49). The slowing of the fastest isometric contractions was caused by a smooth slowing of build-up of EMG activity indistinguishable from the EMG pattern in HD patients.

**Alternating Index Finger Movements**

The **fastest voluntary serial movements**. After having presented the analysis of single isometric contractions we will now demonstrate the impairment of voluntary alternating movements in HD. In fig. 10A the acceleration signal of most rapid alternating index finger movements in a normal subject (left side) and that in an HD patient (right side) are presented. The spectral composition of these signals (fig. 10B) shows that the normal subject can move his index finger initially at an alternation frequency of about 8 Hz. After a few seconds the peak frequency gradually declines to values around 4 Hz. The sharp peaks in the spectra indicate that the movement was performed very regularly. In the HD patient, however, the spectra are flat without clear peaks, which reflects the irregularity of the movements. Only initially a clear 4 Hz peak is visible. Compared with the normal subject the maximal alternation frequency is considerably slowed.

Group means of maximal alternation frequencies were 6.77 ± 0.94 Hz for normals compared with 4.91 ± 1.5 Hz in HD patients ($P < 0.001$, Student’s t test (two-tailed)). The regression between maximal alternation frequency and age was plotted for the normal population along with 2 SD intervals to define the normal range (fig. 10C). In normals the frequency of the most rapid alternating index finger movements slightly decreases with age (slope = 0.024 Hz/yr, $r = 0.258$, n.s.) (fig. 10C, left panel). In HD patients there was a clear decrease of frequency with age (slope = 0.087 Hz/yr, $r = 0.555$, $P < 0.01$). Only 7 out of 21 patients produced the
Fig. 10. Accelerometer recordings (A) and power spectra (B) in arbitrary units of most rapid alternating index finger movements in a normal subject (left side) and in a patient with HD (right side). Over a period of 34 s consecutive partially overlapping spectra are presented. In the normal subject the maximal alternation frequency is at around 8 Hz and drops to 4 Hz after some seconds (left panel). The patient with HD is unable to produce regular movements as reflected in the lack of sharp peaks in the spectra. The maximal alternation frequency is decreased to values around 4 Hz (c), age-related regression of maximal frequency of alternating index finger movements. In normal subjects maximal alternation frequencies decrease with age (left). In most of the HD patients (middle) and in some of the subjects at risk (right) maximal alternation frequencies are decreased. Squares indicate at risk subjects with 'soft' signs.
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most rapid movements at an alternation frequency within the normal range (fig. 10c, middle panel). Also in 7 individuals at risk, maximal voluntary alternation frequency was slowed beyond normal limits (fig. 10c, right side).

Since the forearm was not fixed these measurements may be influenced by involuntary movements of the forearm or the whole arm. Single hyperkinetic movements, however, would yield a low frequency spectral component below 2 Hz. Hence interference by single hyperkinetic movements cannot explain the occurrence of peaks in the spectra of voluntary alternating movements in the range between 4 and 6 Hz.

Involuntary alternating motor activity: postural tremor. Analogous to the analysis of single isometric contractions, we recorded not only voluntary but also involuntary alternating motor activity (tremor) in order to examine whether the involuntary alternating tremor movements are slowed in HD. The outstretched hand of a normal subject shows some low amplitude postural tremor (fig. 11A, left side). The frequency spectra of such a physiological tremor are shown from bottom to top for a 25 s epoch on the left side of fig. 11B. The right side of fig. 11B presents the time course of the spectral composition of tremor of a typical HD patient. In the normal subject, spectra are characterized by fairly broad spectral maxima at frequencies around 9 Hz. In contrast, in the HD patient the spectral maxima are shifted to lower frequencies (around 6 Hz). They also show a broad-based peak similar to that in normal physiological tremor. The average peak frequency in the HD group was 5.32 ± 2.25 Hz compared with 8.64 ± 1.13 Hz in the age and sex-matched control group (P < 0.001, t test (two-tailed)). To identify abnormalities of tremor of peak frequency in individual cases the age-tremor peak frequency regression of the normal population was computed. As illustrated by the left of the three panels of fig. 10c, peak frequency does not increase significantly with age over the range from 16 to 58 yrs (slope = 0.011 Hz/yr, r = 0.104, n.s.). In the HD patients, however, there is a clear dependence of peak frequency on age (slope = −0.146 Hz/yr, r = −0.636, P < 0.01). In 14 of 22 patients with HD peak frequency lies below the normal range of peak frequency (fig. 11c, middle panel). Nine subjects at risk had an abnormally low tremor peak frequency (fig. 11c, right panel).

The tremor measurements may have been contaminated by involuntary movements from the nonsupported proximal parts of the upper extremity. With the arm kept outstretched, however, movements around the elbow joint were diminished and the resonance frequency of the whole arm lies certainly below 3 Hz.

Summary of Proportions of Abnormalities Over All Parameters

Table 2 summarizes the incidence of the abnormalities found in HD patients and at risk subjects for the various parameters. It is evident that the measurements of single isometric contractions (AM/CT; CTV) are most frequently impaired whereas tremor peak frequency and peak frequencies of fastest voluntary alternating movements are abnormal only in about two-thirds of the HD patients and reaction times are prolonged only in one-third of all HD patients. A similar distribution of
Fig. 11. Accelerometer recordings (A) and power spectra in arbitrary units (B) of postural tremor of the dominant hand recorded over a period of 25 s in a normal subject (left side) and in a patient with HD (right side); 48 consecutive partially overlapping power spectra calculated from periods of 4.2 s duration are plotted to visualize variation of the spectral composition of the tremor. In the HD patient tremor peak frequency is decreased. (c), age-related regression of tremor peak frequency in normal subjects and normal variation as defined by 2SD above and below the regression line (left panel). Most of the 22 patients with HD (middle panel) show a tremor peak frequency below the normal range. Also some of the people at risk (right side) have lowered tremor peak frequencies. Squares indicate at risk subjects with 'soft' signs.
Table 2: Incidence of Abnormalities of Movement Parameters in Patients with Huntington's Disease (HD) and Relatives at Risk (HR)

<table>
<thead>
<tr>
<th></th>
<th>HD</th>
<th>HR</th>
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<tbody>
<tr>
<td>TRE</td>
<td>14/22 (64%)</td>
<td>9/40 (23%)</td>
</tr>
<tr>
<td>VAM</td>
<td>14/22 (64%)</td>
<td>7/39 (18%)</td>
</tr>
<tr>
<td>AM/CT</td>
<td>20/22 (91%)</td>
<td>14/37 (38%)</td>
</tr>
<tr>
<td>CTV</td>
<td>20/22 (91%)</td>
<td>14/37 (38%)</td>
</tr>
<tr>
<td>RT</td>
<td>8/22 (36%)</td>
<td>6/37 (16%)</td>
</tr>
</tbody>
</table>

TRE = tremor peak frequency, VAM = maximal alternation frequency of voluntary alternating index finger movements, AM/CT = increase of contraction time with contraction amplitude, CTV = contraction time variability, RT = reaction time.

Abnormal findings across these parameters appear in the at risk group. All percentages observed lie well below the maximal expectable value of 50% gene carriers.

Covariation of Movement Parameters with Disability Score and Disease Duration

The correlation between AM-CT slope and disability has already been described. Furthermore, in the HD patients' other motor parameters and RT have been correlated with disability scores (DIS) and duration of the disease (DUR) (Table 3). Only RT showed a significant ($P < 0.01$) correlation with the disability score. No other parameters were significantly correlated with disease duration.

Association of Motor Parameters with Psychometry in HD

Table 4 summarizes the correlation coefficients of the four psychometric tests and the motor parameters. Except for slight associations of CTV with the Raven and the WIP test and a slight association of the SKT with voluntary alternating movement frequencies and RTs, all other motor parameters did not show any significant covariation with psychometry.
Table 4. Associations (Pearson product-moment correlation coefficients) between movement parameters and psychometric tests (WIP, MWT, Raven, SKT)

<table>
<thead>
<tr>
<th></th>
<th>TRE</th>
<th>VAM</th>
<th>CTV</th>
<th>SCT</th>
<th>RT</th>
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<tr>
<td>WIP</td>
<td>0.0031</td>
<td>0.0847</td>
<td>-0.4319**</td>
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<td></td>
<td>(n.s.)</td>
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<tr>
<td>MWT</td>
<td>0.0377</td>
<td>0.3049</td>
<td>-0.3969</td>
<td>-0.2750</td>
<td>-0.2884</td>
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<tr>
<td></td>
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<tr>
<td>Raven</td>
<td>-0.0618</td>
<td>0.2437</td>
<td>-0.5129**</td>
<td>-0.1639</td>
<td>-0.2629</td>
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<tr>
<td>SKT</td>
<td>-0.1687</td>
<td>-0.5273**</td>
<td>0.2797</td>
<td>0.1276</td>
<td>0.4751**</td>
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* For explanation see text. Abbreviations of movement parameters as in Table 2. ** P < 0.05. n.s. = not significant.

Discussion

The principal finding of this study is that patients with HD are unable to produce motor activities as fast as normal subjects. As shown by quantitative analysis this disturbance is a consistent feature in HD patients and in a proportion of those at risk for the disease. It affects various types of rapid voluntary motor acts: single isometric contractions and serial alternating movements, as well as the involuntary tremor and hyperkinetic contractions or movements. As all of the subjects at risk and the majority of HD patients were free of any medication that might interfere with motor activity, medication effects can be excluded as a possible cause of this slowing.

In normal subjects rapid single muscle contractions are performed at approximately the same time irrespective of their amplitude (Freund and Büdingen, 1978). This independence of contraction time on amplitude is accomplished by a linear increase of the rate of rise of tension with increasing amplitude. The capacity of the motor system to adjust rate of rise of tension to amplitude is referred to as speed control. This speed control system achieves ‘isochrony’ not only for the fastest possible contractions or finger movements but also for a wide range of learned ‘automatic’ everyday movements such as writing (Viviani and Terzuolo, 1980), drawing, typing or even such complex tasks as weight lifting (Enoka, 1983). This stable relationship between contraction time and amplitude is lost in more severely affected HD patients. The more severely affected is the patient, the slower the rapid contractions and the larger the increase in contraction time with contraction amplitude. Thereby the patients lose an invariant feature of motor control.

One possible neuronal mechanism underlying the impairment of the speed control system is the disturbance of firing rate modulation in the final common pathway. The generation of rapid movements requires short high-frequency bursts.
released from the motoneurons. At the present time there is no information about single unit recordings from HD patients during rapid contractions, but it seems likely that these patients are unable to produce firing rates as high as required for the generation of normal rapid contractions. In PD there is direct evidence from motor unit recordings (Dietz et al., 1974) that the discharge rates are slower than in normals.

The impairment in speed control demonstrated for HD patients in this study resembles the slowing of motor activity that is well documented in many quantitative studies of patients with the more common basal ganglia disorder of Parkinson's disease (Lance et al., 1963; Draper and Johns, 1964; Barbeau and de Groot, 1966; Brumlik and Bosches, 1966; Flowers, 1975, 1976; Hallett and Khoshbin, 1980; Teräväinen and Calne, 1980; Evarts et al., 1981; Baroni et al., 1984). This suggests that slowing of motor activity may be caused by pathological mechanisms common to both basal ganglia disorders. Closer inspection of the fine structure of these abnormalities, however, especially for the fastest contractions, discloses considerable differences. As demonstrated by Hallett and Khoshbin (1980) and Teräväinen and Calne (1980) the EMG records of PD patients performing single rapid voluntary movements are characterized by a lack of increase in the size of the first agonist burst with increasing amplitude and, to compensate for this deficit, by series of subsequent bursts alternating from agonist to antagonist. Analysing the fastest isometric index finger extensions in PD patients with and without tremor at rest in the same way as described here for HD patients, we invariably found oscillations superimposed on the contraction curves caused by repetitive EMG bursts occurring simultaneously in the agonist and antagonist with a frequency of 10 to 13 Hz (Hefter et al., 1985; Hömberg et al., 1985). This kind of action tremor measured under isometric conditions has already been analysed by Lance et al. (1963) and probably parallels the oscillations seen in the isotonic recordings of Hallett and Khoshbin (1980) and Teräväinen and Calne (1980). The latter authors also emphasized that these EMG oscillations during voluntary movement were independent of the presence of resting or postural tremors in their PD patients. The changes in EMG activity underlying the slowing of motor activity are different in HD as compared to PD although the resulting slowing as measured by mean values of contraction times may be similar. In HD we find a deficit in building up EMG activity causing prolongation of EMG bursts and slowness of contraction without evidence for tremor such as bursting in the EMG or oscillations superimposed on the force trajectory as in PD.

The comparison between samples of superimposed tremor beats recorded isometrically from the finger of a PD patient and some superimposed finger hyperkinesia of similar force amplitude recorded from a HD patient show that the time course in both kinds of different contractions is nearly the same. Thus the only difference between both kinds of involuntary activity remains that the tremor in PD is regular and hyperkinesia in HD is irregular. An association between the slowing of movement and the occurrence of an abnormally slow tremor of high amplitude is
frequently observed in basal ganglia and cerebellar disorders. Cooling experiments in cerebellar nuclei and in the striatum (Conrad and Brooks, 1974; Cooke and Thomas, 1976; Hore et al., 1977; Vilis and Hore, 1980) have shown that the ongoing neural activity is desynchronized in both structures before cooling but becomes slowed and synchronized during cooling. This goes along with the development of coarse limb oscillations. In contrast, neither cooling nor ablating the motor cortex changes the time course of rapid plantar flexions performed by monkeys, but reduces movement amplitude (Rüegg and Juvet, 1984).

In HD the involuntary activity interferes with voluntary activity in an unpredictable way as for the impulses of a random generator. As Wilson mentioned as long ago as 1928, the involuntary movements in HD patients show similarities to voluntary movements, thus resembling ‘motor subunits’. Wilson suggested that in HD patients the motor system fails to suppress inappropriate and to select and maintain appropriate movements (Wilson, 1928; Penney and Young, 1983). The results by DeLong and coworkers shed some light on this problem (DeLong and Georgopoulos, 1979, 1981; Crutcher and DeLong, 1984a, b; Alexander and DeLong, 1985a, b): analysing single cell recordings in the caudate and putamen in monkeys, they found that the striatum receives a somatotopically-organized input from the motor cortex (see also Liles, 1979) and from frontal association and sensory areas. Microstimulation at different points in the putamen led to movements of different parts of the body, caused by contractions of ensembles of several muscles. Thus they observed the activation of ‘motor subunits’ during their stimulation experiments in the basal ganglia.

A pattern of coactivation between the finger extensor and flexor muscles in the isometric paradigm was consistently observed in all normal subjects, which confirms observations by other authors of similar coactivation in isometric forearm contractions (e.g., Sanes and Jennings, 1984). This pattern was maintained in all HD patients. The onset of agonist activity was followed as early as 20 to 30 ms by concomitant activation of the antagonist muscle. Furthermore, EMG recordings of fastest isotonic elbow extensions and rapid alternating movements at the wrist in HD patients revealed normal triphasic or alternating EMG patterns, respectively. This corroborates observations of normal EMG patterns during voluntary movements made in a few HD cases (Marsden et al., 1983) and in one patient with Sydenham’s chorea (Hallett and Kaufman, 1981). The findings indicate that the pattern of activation of muscle groups for simple voluntary movements is preserved in HD patients.

The interaction between voluntary and involuntary contractions in patients with HD was less obvious during larger contractions than during small contractions. By contrast, in PD small amplitude contractions are performed much better than large amplitude contractions. During large amplitude contractions tremulous oscillations inevitably are triggered by the voluntary activity. In monkeys, substantia nigra pars compacta neurons do not show a detectable change of firing rates during small amplitude movements or contractions. However, they change firing rates
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considerably during large amplitude movements (DeLong and Georgopoulos, 1979; Schultz et al., 1983). Thus deficits in dopamine release are more relevant for large amplitude movements than for those of small amplitude.

Reaction time in HD is impaired to a much lesser degree than the parameters related to actual movement execution. Similar observations have been made in Parkinson's disease (Wiesendanger et al., 1967; Heilman et al., 1976; Evarts et al., 1981). It also fits with the results of single unit recordings in the basal ganglia of monkeys. DeLong et al. (1983) studied the relation of neural discharge in various parts of the basal ganglia to different movement parameters and showed that the discharge of these neurons was correlated with movement parameters such as amplitude, velocity and direction. These units fired only during movement execution or just before the mechanographic onset of movement, during which EMG activity builds up. In contrast there was nearly no enhancement of single unit activity in the earlier premovement period and hence no evidence of single unit activity in the basal ganglia related to movement preparation. Our findings also match the effects observed by Horak and Anderson (1984a, b) of kainic acid lesions or stimulation in the putamen and globus pallidus of monkeys. During rapid arm-reaching movements to a visual target, lesions of the contralateral globus pallidus led to prolonged movement times caused by slowing of EMG build-up, with little or no change of reaction times. On the other hand stimulation of the globus pallidus facilitated speed of movement but left RTs unaltered.

All but one of the HD patients studied did not present with severe rigidity. This stands in contrast to earlier views regarding slowing of movement as a sequel to rigidity (e.g., Walshe, 1955) and is in accordance with Lhermitte (1923), who introduced the term 'syndrome akinetique sans hypertonie' for cases with extrapyramidal symptoms following encephalitis lethargica. Whereas our observations in the patient with Sydenham's chorea were indistinguishable from the findings in HD, the patient with the rigid variant of HD (Westphal, 1883) showed a 'parkinsonian' pattern of bradykinesia as characterized by repetitive oscillations superimposed on the force profile. Furthermore, this patient was the only one presenting with a significantly synchronized postural tremor resembling the postural tremors commonly seen in PD. It therefore appears that the kinesiological and EMG findings in the Westphal variant differ from those in 'typical' HD, but closely resemble those in PD. This finding does also suggest different pathophysiological mechanisms in the Westphal variant.

As dementia is the second major symptom of HD, it has to be considered how far the cognitive dysfunction of the HD patients may have contributed to the pathological motor results obtained. In this respect it is important that reaction times were less abnormal in HD patients than slowing of movement execution. If the cognitive or attentional impairment had caused the observed motor abnormalities, even more prolonged reaction times would have been expected. Furthermore, the slowing of motor activity was observed in a high percentage of individuals at risk not showing any evidence of cognitive impairment and also in some patients in early
stages of HD with little or no evidence of cognitive impairment. CTV was the only parameter showing a covariation with the psychometric tests. This is not surprising since CTV is not a pure motor parameter but measures reproducibility, and is probably the reason why it has a better correlation with the Raven than with the MWT test. Finally there was no significant covariation between detailed psychometric data and the other motor parameters except for only marginally significant associations between the frequency of fastest alternating movements, and reaction time for the SKT. This test actually implies motor tasks and reaction time measurements. The bulk of general intelligence tests in both verbal and nonverbal varieties, however, did not show any significant association with the abnormalities of movement parameters. Hence it appears that cognitive impairment as such is not a major factor in the slowing of motor activity observed in this study. Cognitive impairment may aggravate the motor disabilities but does not cause them.

We observed abnormalities of the motor parameters in up to 40% of clinically normal people at risk of developing the disease later in life. This may indicate that the physiological analysis used here can detect preclinical impairment in the motor system in HD and can be used to develop a motor score for early detection of HD (Hefter et al., 1986) but also for therapy control. Experiments can be repeated several times since they are not stressful for the patient. Besides their clinical usefulness, the findings are also pertinent to the understanding of basal ganglia function in motor control. In these very early stages of HD, pathology is restricted to loss of intrinsic neurons in the basal ganglia in contrast to increasing cortical involvement in later stages (Lange, 1981; Lange et al., 1984). Therefore the early stages of HD, in particular, can be regarded as a model for basal ganglia abnormality, which is not caused by a lack of dopaminergic modulation as in PD, but is due to a reduction in the number of intrinsic basal ganglia interneurons and output neurons. In this context the differences in the EMG pattern underlying the slowing of rapid contractions between PD and HD provide information about the different contribution of different circuitry in the basal ganglia on abnormalities of motor control in these two disorders.

ACKNOWLEDGEMENTS

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