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Gordon's Reflex Phenomenon in Huntington's Disease

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Summary. The patellar tendon jerk was examined quantitatively in 8 cases of Huntington's disease and in 27 controls using surface electrodes over the quadriceps femoris and biceps femoris muscles. The purpose was to investigate whether Gordon's patellar phenomenon was caused by a lack of antagonistic innervation or by late agonistic tonic reflex activity in Huntington's disease. Specific results could indicate that the patellar tendon jerk could be used as a diagnostic aid, as is the long loop reflex of hand muscles. A late tonic stretch response was found, appearing between 160 and 260 ms after the tendon tap. This response appeared in 6 out of the 8 Huntington's cases, even when Gordon's phenomenon was not observed. Therefore, it could be assumed that the tonic quadriceps activation gives rise to the clinically observed Gordon's phenomenon.

Key words: Gordon's phenomenon – Huntington's disease – Long latency reflex

Introduction

A monosynaptic phasic stretch reflex of the quadriceps femoris muscle (Patellar Tendon Jerk PTJ) can be elicited by a short tap on the infrapatellar tendon. The stretch reflex is followed by a tonic polysynaptic myotatic response (Henatsch 1986). The PTJ can be elicited using electrical or mechanical stimuli. The latency is 14 ms for electrical and 18–24 ms for mechanical stimuli (Brown 1984; Chan et al. 1979; Gassel and Ott 1973; de Weerd and Jonkman 1986). It decreases in cases of facilitation. The PTJ has been examined quantitatively in radiculopathies (de Weerd and Jonkman 1986) and in central motor disturbances (Altenburger 1937).

In about one-third of Huntington's cases the tendon jerk is very brisk (Bruyn 1968; Gordon 1908). Bing wrote in 1932 that one could sometimes find that the elevated foot does not go down immediately, but remains elevated for a few seconds because of repeated quadriceps contractions (Bing 1932). This tonic reflex response has been called "hung up" or Gordon's phenomenon (Bing 1932; Grinker et al. 1960; Hassler 1953). It can be used to confirm the diagnosis of Huntington's Chorea. Hassler explained the phenomenon as the result of a pathological synkinesia (Hassler 1953). Bruyn described a disturbed reciprocal muscle innervation with paradoxal contraction due to simultaneous activation of agonistic and antagonistic muscles as the reason for the phenomenon (Bruyn 1968).

The loss of long latency muscle responses on the hand has proved to be a valuable criterion for the early diagnosis of Huntington's disease (Noth et al. 1985). Therefore, it was of interest to examine the obviously affected PTJ quantitatively in Huntington's disease. The question was whether Gordon's phenomenon is due to hypoactivity of antagonistic muscles or to hyperactive tonic responses of the quadriceps femoris muscle.

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Materials and Methods

A total of 27 controls (age 17–75 years, average 36 years, height 162–192 cm, average 177 cm) and 8 patients suffering from clinically manifest Huntington's chorea (age 22–71 years), average 50 years, height 167–182 cm, average 174 cm) were examined. Despite the manifest symptoms the patients were still able to move independently. The diagnosis was confirmed in every case by the slowly progressing hyperkinetic symptoms and atrophy of the caudatum head seen on cranial CT or by positive family anamnesis (Folstein et al. 1986).

For examination the subjects lay comfortably on a reclining EEG chair. The recording was done from both legs consecutively using surface Ag-AgCl electrodes (diameter 5 mm, distance 50 mm). The electrodes were fixed (cathode distally) over the bellies of the lateral vastus muscle and the biceps femoris muscle, 20 to 25 cm proximal of the cleft of the knee joint. The PTJ was elicited by a tap with a reflex hammer done as equally as possible on the infrapatellar tendon. The two channel derivation was triggered by the signals of a piecoelectric pressure gauge via a bridge amplifier. The muscle potentials were rectified and 32 runs were averaged (200 Hz–10 kHz, 200–500 μV/D input sensitivity, 500 ms sweeptime, 1024 signal points per sweep). The six examinations were done in a randomized order: right and left side, resting leg, stretched and bent knee joint. The muscle innervation of knee flexors was monitored on a scope to maintain about 30% of the maximal strength. The power of knee extensors was defined by the weight of the lower leg in the sitting position. Every patient was able to actively hold the lower leg in an extended position. The values were read using an electronic cursor on the screen of the oscilloscope. We did not calculate areas because sometimes the borders of the potentials were not distinct. Statistical analysis was done using Fisher's test.

Table 1. Average values and SD of the reflex examination of 27 controls and 8 Huntington cases. The derivation was done in three conditions: at rest, with bent and stretched knee joints. The results of the statistical analysis with the Mann-Whitney test and Fishers test are shown when significant.

	Leg examined		Reflex examination					
			Controls ($n = 27$)			Huntington's disease ($n = 8$)		
			Right	Left	One or both sides	Right	Left	One or both sides
At rest	PTJ Latency (ms)	n	27	27	27	8	8	8
		$\bar{x}$	19.1	19.0		18.9	18.8	
		SD	2.25	2.54		2.18	2.59	
	Mann-Whitney	U				/	/	
	Amplitude (mV)	$\bar{x}$	0.65	0.59		0.37	0.32	
		SD	0.68	0.68		0.16	0.16	
	Mann-Whitney	U				/	/	
	Duration (ms)	$\bar{x}$	27.1	27.8		30.0	31.3	
		SD	5.23	3.56		2.27	3.05	
	Mann-Whitney	U				1.68	2.40 $P = 0.02$	
	✻ 160–260 ms	n	4	0	4	5	4	6
	Quariceps response latency (ms)	$\bar{x}$	206.0	/		192.4	199.0	Fisher $P = 0.004$
		SD	21.79	/		22.99	21.37	
	Mann-Whitney	U				/	/	
	Amplitude (mV)	$\bar{x}$	0.01	/		0.02	0.01	
		SD	0.004	/		0.02	0.007	
	Mann-Whitney	U				/	/	
Bent	✻ 160–260 ms	n	4	2	4	6	6	6
	Quadriceps response latency (ms)	$\bar{x}$	159.0	183.5		172.8	176.4	Fisher $P = 0.006$
		SD	27.53	10.61		19.99	28.08	
	Mann-Whitney	U				/	/	
	Amplitude (mV)	$\bar{x}$	0.01	0.01		0.02	0.01	
		SD	0.01	0.004		0.02	0.009	
	Mann-Whitney	U				/	/	
Stretched	Silent period (ms)	n	27	27	27	8	8	8
		$\bar{x}$	120.8	121.8		122.9	124.5	
		SD	33.74	32.63		33.77	29.06	
	Mann-Whitney	U				/	/	
	120–200 ms	n	9	11	12	7	7	8
	Quadriceps response latency (ms)	$\bar{x}$	141.2	137.9		137.9	135.9	
		SD	31.17	23.93		29.24	27.51	
	Mann-Whitney	U				/	/	
	Amplitude (mV)	$\bar{x}$	0.01	0.01		0.02	0.02	
		SD	0.009	0.007		0.012	0.007	
	Mann-Whitney	U				/	14 $P < 0.05$	

The significance of differences was examined using the Mann and Whitney non-parametric test.

Results

The latencies of the phasic reflexes (PTJ) correlated with body height, as had been expected (Gassel and Ott 1973) ($Y = 1.5X + 148.5$, $r = 0.41$, $P < 0.01$). The PTJ (latency right side: 19.05 ± 2.25 ms, left side: 18.97 ± 2.54 ms) was followed by different potentials. The range of variability was very high for the PTJ amplitudes (Table 1), and therefore the values in the patient group did not diverge significantly. The duration of the PTJ was prolonged in the patients only on the left side. The PTJ could also always be seen in the antagonistic biceps femoris muscle (Fig. 1). A tonic flexor activation at a latency of 80 ms (7 probands, 6 patients, Fig. 2) and a tonic extensor reponse after 80–100 ms (2 probands, 0 patients, Fig. 1) were seen inconsistently. But otherwise the pattern of tonic muscle responses following the PTJ was relatively constant.

In the bent condition the silent period of the biceps femoris muscle was interrupted by an 80–160 ms response in patients as well as in probands (20/27 controls, 8/8 patients; latency: controls right 79.9 ± 11.8 ms, left 80.5 ± 11.6 ms, patients right 72.5 ± 15.3 ms, left 70.1 ± 14.3 ms) (Fig. 1, line 4, Fig. 2, line 4). There was no significant difference between the groups

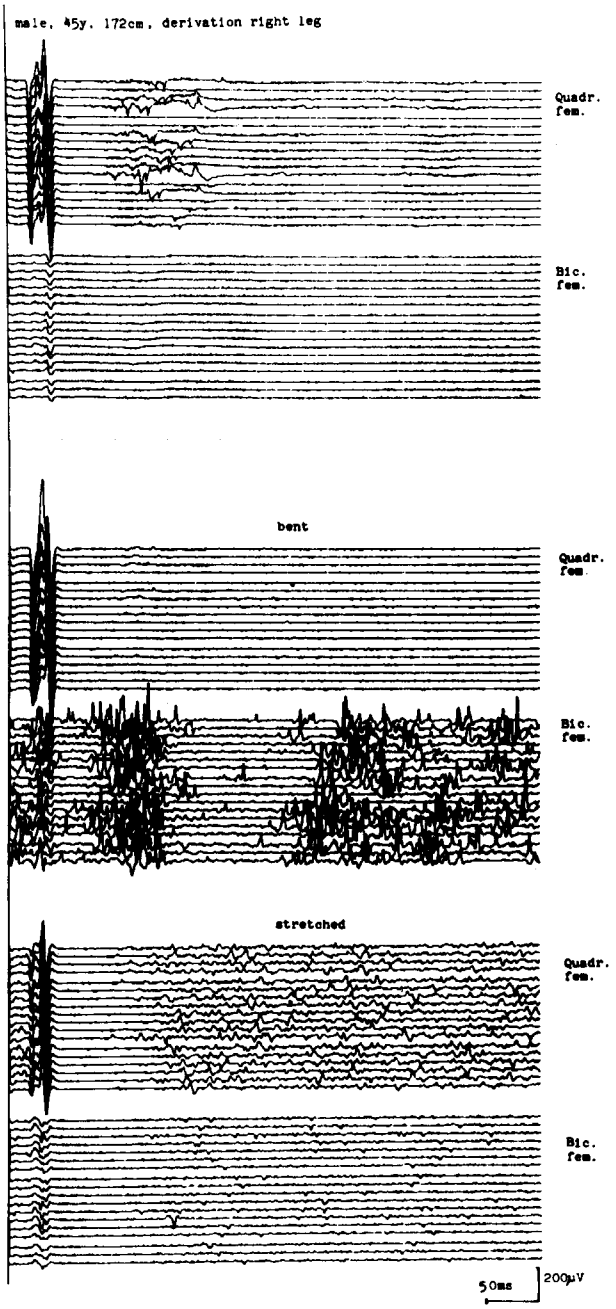

Fig. 1. One example of the knee tendon jerk examination of a control subject in rest, bent and stretched conditions. The derivation of the antagonistic muscles is shown in cascades of single sweeps. The phasic patellar tendon jerk (PTJ) response appears highly synchronised, with a latency of 19 ms. In the rest condition a tonic extensor response can sometimes (2/27 controls) be seen after 80 ms *(upper traces)*. In the bent condition the flexor silent period is interrupted by a tonic activity after about 80 ms *(middle traces)*. In extension the PTJ is followed in the quadriceps muscle by a silent period of 140 ms duration *(lower traces)*

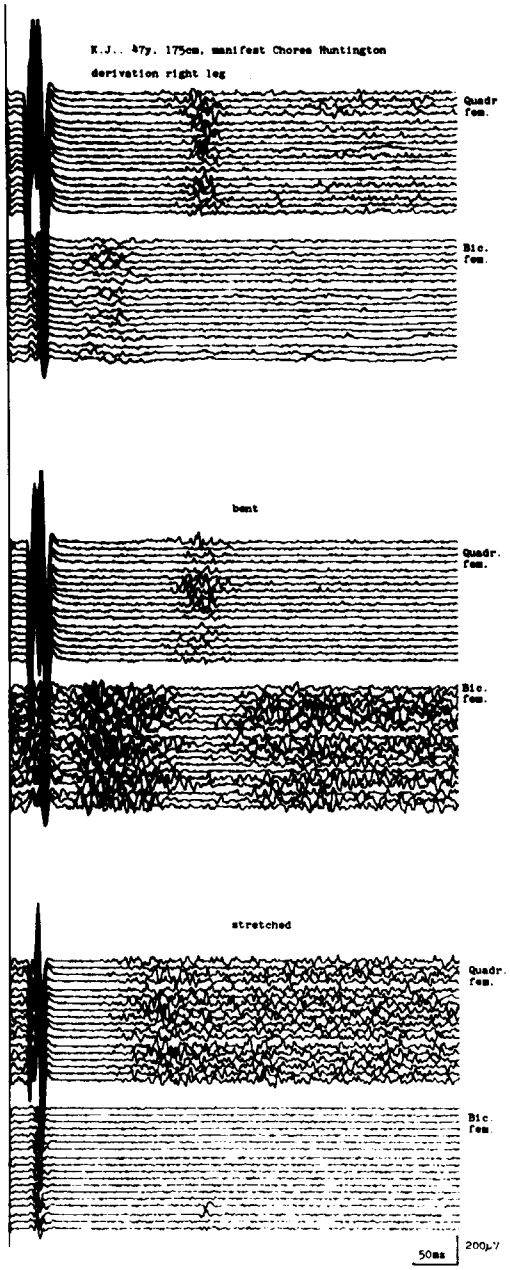

Fig. 2. Derivation in a case of Huntington's chorea, cascades of single sweeps. At rest *(above)* a tonic reflex response appears in the quadriceps femoris after 200 ms (160–260 ms potential). In the bent condition *(middle)* a tonic extensor response can also be seen after 170 ms (160–260 ms potential). In the stretched condition *(bottom)* the extensor activity is augmented at the end of the silent period (120–200 ms potential)

concerning this tonic flexor activation, which was seldom seen in the rest condition. The duration of the silent period (which appears in the flexors in flexed condition and in the extensors in extended condition) also did not vary significantly between the two groups (extensors 121–125 ms, Table 1). When examining control subjects at rest a tonic quadriceps femoris activation at 160–260 ms was seen only sometimes and then unilaterally (4 of 27, Fig. 3a above). On the other hand this response was seen clearly in 6 of the 8 Huntington's cases in the rest condition (3 times bilaterally) and also in the bent condition (6 times bilaterally, Fig. 3b bottom line). This was significantly more frequent in the patient group (rest condition Fisher $P = 0.004$, bent condition Fisher $P = 0.006$). In the derivations of the stretched legs a clearly demarcated increase in activity was sometimes seen in the quadriceps femoris muscle at the end of the silent period without any significant difference between the groups (120–200 ms response, Table 1, Fig. 4 bottom).

The substantial difference between the controls and Huntington's cases therefore seems to be a tonic extensor muscle

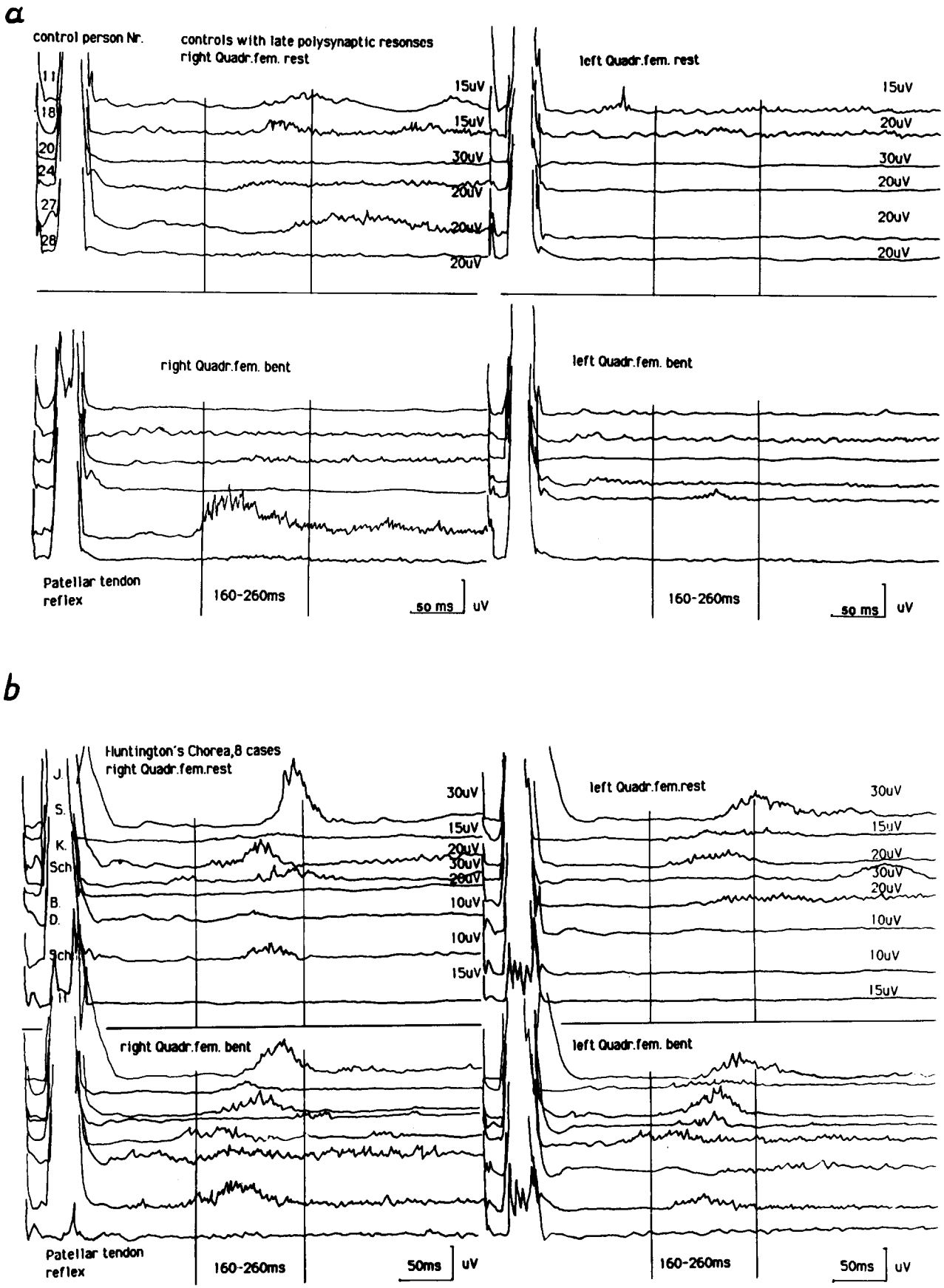

Fig. 3a, b. Recording of the rectified and averaged derivations from the quadriceps femoris muscle in rest and bent conditions. **(a)** Examinations of the 6 control subjects where tonic reflex activity appeared. **(b)** Examinations of the 8 Huntington's chorea cases. A clearly demarcated 160–260 ms potential was seen only in 1 control (number 27) in the quadriceps in the bent condition. On the other hand this tonic response appears in almost every patient's examination

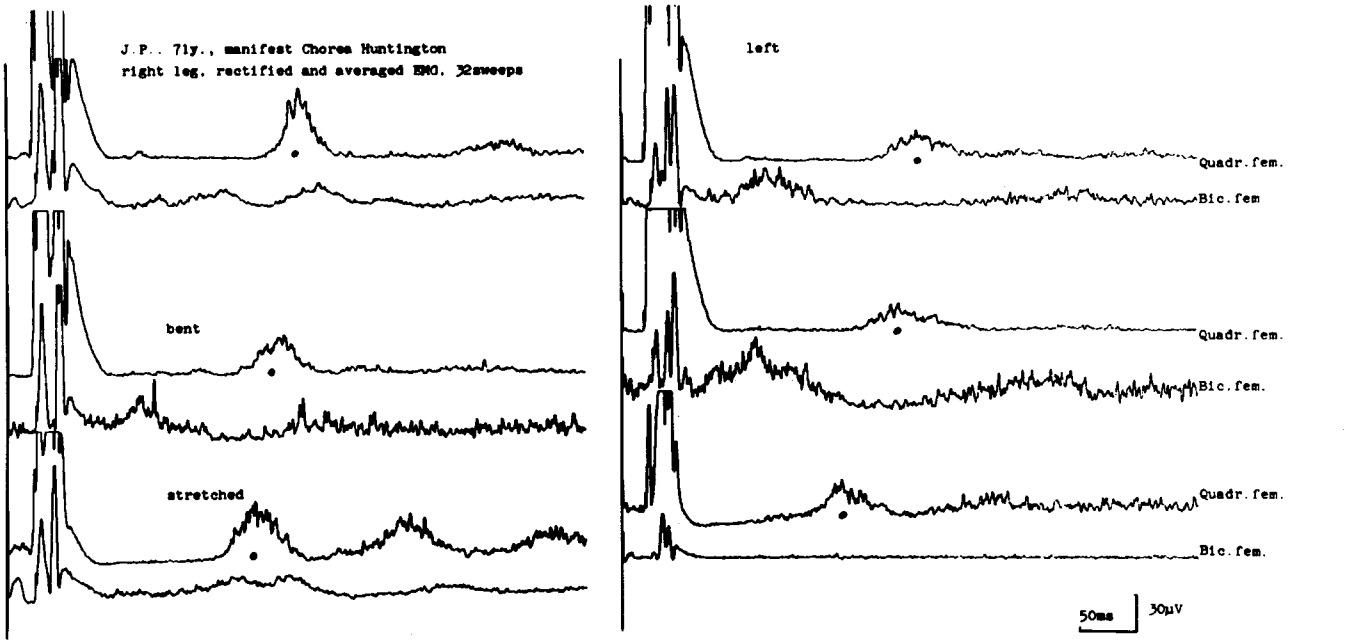

Fig. 4. Examination of a patient, rectified and averaged derivations (32 summations). In the quadriceps femoris of the right as well as left side a tonic activation can be seen in all three conditions which is marked by points (rest and bent: 160–260 ms potential, stretched: 120–200 ms potential)

response from about 160–200 ms in the rest and bent condition. This is shown in the example in Fig. 4. In the stretched condition this response appeared from 120–200 ms and was not demarcated as clearly.

Discussion

The phasic monosynaptic PTJ is not consistently exaggerated in Huntington's disease (Bruyn 1968) and, the neurophysiological parameters of this reflex remain almost totally unaltered by the disease (Table 1).

A tonic extensor response recorded in the rest condition inconstantly after 80–100 ms is supposed to be related to the functional stretch response of Chan et al. (1979). These authors assumed that the reflex is generated not only by segmental, but also by suprasegmental influences, possibly through a transcortical loop. Examinations of the gait subjected to disturbing impulses revealed a tonic reflex response (80–150 ms) of the quadriceps femoris muscle which was interpreted as a tonic polysynaptic spinal reflex (Quintern et al. 1985). Probably both spinal and supraspinal influences on late tonic reflex responses of leg muscles can be assumed (Meinck et al. 1985).

In the controls the antagonistic thigh muscles were activated tonically in an alternative order after the stretch reflex. This interaction of antagonists (Bruyn 1968) is disturbed in Huntington's chorea (Fig. 4). But, a tonic extension of the knee joint – Gordon's phenomenon – cannot be explained by simultaneous activation of antagonistic muscles alone. The knee flexor activity is not reduced in Huntington's chorea. The outstanding result of the patient examinations was a clear tonic extensor response after about 160 ms in rest and in bent conditions (160–260 ms response, Figs. 2–4). This potential was very seldom seen in controls (Fisher $P = 0.006$). The extensor response is probably not a result of voluntary innervation, because it also appeared in the bent condition when the extensors of the knee joint are not voluntarily innervated. Therefore, it is supposed that it is an involuntary reflex response.

As a conclusion, the reason for the prolonged knee extention after the PTJ in some cases of Huntington's chorea, is the increased reflectory tonic extensor response after a latency of about 160 ms (160–260 ms response). Because of the long latency the influence of supraspinal centres on the reflex is probable (Fahrenkamp 1913), but has not yet been proved. Spinal inhibition phenomena can also explain the result in part (Altenburger 1937). The mechanical stimulus affects type II afferents in addition to type Ia spindle afferents (Cody et al. 1986); Dietz et al. 1985; Eisen et al. 1984; Meinck et al. 1985). The reflex pathway of the tonic 160–260 ms extensor response remains unclear.

The extensor response was clearly increased in the 3 patients with a positive Gordon's phenomenon. However, the tonic extensor response was also clearly visible in 3 patients with a normal PTJ indicating that neurophysiological reflex examination is more sensitive than clinical testing. But it is not a highly specific finding because in 1 of the 27 controls with brisk reflexes the 160–260 ms response was also positive in rest and bent conditions. It is still not known if the 160–260 ms response is also positive in persons at risk, as is the long loop reflex examination (Noth et al. 1985). In contrast to the long loop reflex in the upper extremity, which is supposed to be generated through supraspinal nerve centres, the tonic reflex response in the lower leg does not disappear but increases in Huntington's chorea.

The results confirm Fahrenkamp's assumption (1913) that the maintained extension of the lower leg after PTJ is due to tonic quadriceps activity (Fig. 4).

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