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Dystonia is characterized by sustained muscle contractions, frequently causing twisting and repetitive movements or abnormal postures. For various reasons many patients with dystonia have been considered to suffer from a psychogenic disease rather than an organic one. The dyskinesias often have a bizarre nature and may appear only on certain characteristic actions, while other motor actions employing the same muscles are carried out normally. Furthermore, they may be relieved by certain inexplicable tricks.

In recent years insight into the pathophysiology of dystonia has grown due to clinical neurophysiological, radiological and nuclear investigation tools (chapter 1). In the studies in this thesis neurological features and soleus H-reflex tests have been explored to improve the understanding of pathophysiological mechanisms involved in dystonia and to facilitate the diagnosis of dystonic features. The soleus H-reflex is evoked by electrical stimulation of afferent fibers (Ia fibers) from muscle spindles running through the posterior tibial nerve and appears at lower stimulus intensities and has a longer latency than the direct muscle potential (M-potential) evoked by activation of motor nerve fibers. Ia afferents may mono- and oligosynaptically excite homonymous motoneurons resulting in an electromyographic response of the soleus muscle, called the Hoffmann (H)-reflex (chapter 2). The soleus H-reflex studies in this thesis concern the ratio of the maximal H-reflex response to the maximal direct muscle potential (H/M ratio), the amount of inhibition of the soleus H-reflex response during vibration of the Achilles tendon, and the recovery functions of a second H-reflex elicited at variable time intervals after a preceding H-reflex response. The H/M ratio reflects the excitability state of the motoneuronal pool, and vibratory inhibition mainly the autogenic axo-axonal presynaptic inhibition of the Ia fiber terminals. The various phases of the recovery curve of the soleus H-reflex probably correspond with activity of cutaneous afferents and polysynaptic pathways either in the spinal cord or along long-loop pathways and presynaptic inhibitory mechanisms as well. In chapter 3 soleus H-reflex results obtained in patients with leg dystonia are compared with those obtained in healthy controls and in patients with dystonic features without involvement of the leg. In patients with leg dystonia, vibratory inhibition of the soleus H-reflex response is depressed compared to findings in healthy subjects, whereas
recovery features of the soleus H-reflex were clearly enhanced. These findings suggest that spinal inhibitory mechanisms are diminished compared to those present in healthy control subjects. The motoneuronal pool excitability as reflected by the H/M ratio was normal. The normal H/M ratio in dystonia contrasts with the increased H/M-ratio in spasticity. In patients with only arm involvement soleus H-reflex tests in the leg fell into the normal range, which suggests that the soleus H-reflex abnormalities were associated with clinical involvement of the extremity under study. With combined soleus H-reflex test variables most patients could be discriminated correctly from healthy controls. In three patients with dopa-responsive dystonia (chapter 4) a clear relationship could be demonstrated between changes in soleus H-reflex test responses and the disappearance and reoccurrence of dystonia. The clinical improvement due to levodopa treatment paralleled normalization of abnormal soleus H-reflex features. In the one patient in whom treatment was discontinued, dystonic signs reoccurred after 5 days concurrent with reoccurrence of soleus H-reflex abnormalities. These findings provided further evidence that alterations in soleus H-reflex tests are allied to dystonia.

In some patients dystonic movements occur after a peripheral trauma (chapter 5). Dystonia after a peripheral trauma is a debated entity and is also considered to be of a psychogenic origin. In contrast to patients with a presumed central origin of dystonia most of these patients show a fixed dystonic posture. The soleus H-reflex test abnormalities seen in five of these patients did not differ from those seen in patients with dystonia of a presumed central origin. However, similar soleus H-reflex test abnormalities were found in healthy controls who were asked to mimic the dystonic posture. As such the soleus H-reflex tests could not discriminate mimicked dystonia from trauma induced dystonia or from dystonia of central origin. These findings falsify the hypothesis suggested by us earlier, that soleus H-reflex tests might be helpful in discriminating psychogenic dystonia from organic dystonia. However, during the subsequent studies the distinction between organic and psychogenic had become increasingly unclear. The hesitance was caused by a number of factors. Firstly, in primates with dystonia after repetitive strain, studies showed plastic changes in the brain as a result of repetitive sensory stimulation. In these primates dedifferentiation of receptive fields occurred in the sensory cortex. Dedifferentiation of cortical fields, however, may not be limited to the sensory cortex itself. Reorganization in the motor cortex in dystonic patients are suggested to occur by observations obtained in
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transcranial magnetic stimulation studies. The observation of a patient with ‘mirror’
dystonic movements during writing also suggested that dedifferentiation of motor
fields are related to the development of dystonic symptoms (chapter 7). Secondly,
the study performed in six patients with a dystonic or myoclonic movement disorders
after trauma who repeatedly showed a beneficial although temporary effect after electro-
acupuncture (chapter 6). Patients in this study did not respond to various interventions
or medical treatment, making a specific effect due to acupuncture more likely. In
these patients soleus H-reflex abnormalities also improved after acupuncture. Electro-
acupuncture has been demonstrated to reorganize prominently the sensory cortical
receptive fields. Therefore, beneficial effects of electro-acupuncture may further signify
the importance of sensory input in the generation of movements. Thirdly, sensory
tricks were not only helpful in alleviating dystonic movement but also appeared to
improve normal movements. This was demonstrated in a patient with writer’s cramp,
whose writing with the uninvolved left arm improved by manipulation of the affected
right arm (chapter 8). As such, not only in dystonia induction or fluctuation of
dystonic symptoms may mainly depend on various external factors, but it may also be
true in apparently normal voluntary motor action. The diagnosis of a psychogenic
movement disorder is largely based on the variability of signs and symptoms under
influence of environmental conditions (chapter 9). However, this variability does not
discriminate in itself a psychogenic from a dystonic movement disorder. Furthermore,
it implies an arbitrary distinction between mind and brain. As such it may be hardly
possible to interpret a movement disorder to be of psychogenic origin.