Dystonia. Reflexions on movement
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Citation for published version (APA):

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Introduction
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Dystonia is a syndrome of sustained muscle contractions, frequently causing twisting and repetitive movements or abnormal postures (Fahn et al. 1987). It may occur as a generalized disorder, or it may remain rather focal, e.g. in the neck (torticollis), the eyelids (blepharospasm), the mouth (oromandibular dystonia), or in the arm during writing (writer's cramp). In most instances the cause of dystonia is unknown, in which case it is referred to as primary or idiopathic dystonia. It is considered a mysterious disease (Hallett 1995). In series of patients with idiopathic dystonia up to 50 percent had been considered to suffer from a psychogenic disorder (Fahn and Williams 1988; Eldridge et al. 1969). Although it is difficult to define what psychogenic means, the interpretation has been based on a number of observations and assumptions (Marsden 1976). The dyskinesias have a bizarre nature. Their appearance frequently occurs only on certain characteristic actions, while other motor actions employing the same muscles are carried out normally. Relief is induced by certain inexplicable actions. They are sensitive to social and mental stress. No anatomical, physiological, or biochemical abnormalities were found. The belief was held that such patients show overt psychiatric disturbance, and features, e.g., eyelid closure or neck turning, were thought to have a psychopathological meaning. However, it was argued that none of these characteristics comprises decisive evidence in favor of a psychogenic origin (Marsden 1976). In addition, no differences could be found between dystonic patients and control subjects in regard to previous psychiatric history and current life adjustment or on psychiatric testing (Cockburn 1971; Riklan et al. 1976; Sheehy and Marsden 1982). Electrodiagnostic studies in patients with blepharospasm or oromandibular dystonia indicated increased motoneuronal excitability in brainstem nuclei, and thus demonstrated a physical, organic abnormality (Berardelli et al. 1985). Finally, a genetic basis was found for some of the primary dystonic syndromes (Ozelius et al. 1997; Bressman et al. 1998). Eventually, a psychogenic origin of dystonia was considered to be quiet rare (Fahn and Eldridge 1976). Still, series of patients with documented psychogenic dystonia have been described (Fahn and Williams 1988; Lang 1995; Gálvez-Jiménez and Lang 1997), and in many patients problems appear to arise as to whether the origin of dystonic symptoms is psychogenic or not.

Although we consider dystonia to be a movement disorder, there are a number of phenomena which relate to the sensory system suggesting that dystonia could primarily
be a sensory disorder (Hallett 1995). The sensory trick is a well known clinical feature in dystonia. Although in the past the beneficial effect of sensory stimuli upon the dystonic movements was regarded as indicative of a psychogenic disorder, the sensory trick is now considered to be a highly specific sign in dystonia suggesting the organic nature of the disease (Fahn 1987; Hallett 1995). An example of a sensory trick is the gentle finger touch on the skin of the face that in patients with torticollis normalizes the head position or in patients with blepharospasm opens the eyes. Contrarily, in some patients abnormal sensory input is suggested to trigger dystonia. Various authors noted the relationship between trauma to the hand and the onset of writers cramp or the onset of neck spasms in torticollis spasmodica after a recent neck injury (Sheehy et al. 1988; Jankovic 1994). However, also the relationship between a peripheral trauma and the occurrence of disordered movement is debated, and a psychogenic interference is suggested also (Wiener and Shulman 1995; Gálvez-Jiménez and Lang 1997).

In movement disorders, some of the underlying spinal mechanisms involved, can be elucidated electrophysiologically by means of the Hoffmann (H)-reflex of the soleus muscle.

Soleus H-reflex tests are used to examine inhibitory and excitatory mechanisms influencing the excitability of the motoneuronal pool at the spinal level. At the clinical neurophysiology unit of the department of neurology at the Academic Medical Center soleus H-reflex tests were initiated for the study of movement disorders in spastic patients as part of a medical developmental research project into the effects of spinal cord stimulation for treatment of spasticity in 1985. In spasticity, soleus H-reflex test results indicate an increase of motoneuronal excitability and a decrease of inhibitory mechanisms acting upon the transmission of Ia afferents from muscle spindles upon motoneurons (Angel and Hofmann 1963; Matthews 1966; Delwaide 1985; Ongerboer de Visser et al. 1989). These mechanisms may relate with clinical signs of the upper motoneuron syndrome to some extent (Koelman et al. 1993).

The availability of a new treatment for dystonia, injections of botulinum toxin in the dystonic muscles, shifted the interest of the movement disorder study group towards dystonia and the pathophysiological mechanisms involved. Soleus H-reflex test results in patients with dystonia demonstrated a spinal excitability state that is distinct from the one seen in patients with spasticity or in healthy controls (Sax et al. 1976; Bour et
Introduction

al. 1991). In the upper limb similar alterations were suggested from H-reflex response findings observed in patients with spasmodic torticollis and generalized dystonia (Panizza et al. 1990). However, how these alterations in H-reflex tests relate to clinical features of dystonia is uncertain. Furthermore it is unknown whether the etiology of dystonia influences the soleus H-reflex features, and whether they behave differently if a strong sensory component is suspected in the generation of dystonic features. In addition, it is uncertain whether soleus H-reflex relate to the presence or absence of dystonic features within the same patient and whether, in combination with clinical features they can be helpful in the discrimination of a psychogenic as opposed to an organic origin of dystonia.

Objectives

The aims of the studies in this thesis were to explore soleus H-reflex tests and neurological features to improve the understanding of pathophysiological mechanisms involved in dystonia and to facilitate the diagnosis of dystonic features in patients with movement disorders.

Outlines

A review is given on relevant anatomical, etiological and pathophysiological features of dystonia in chapter 1. Technical and physiological aspects of the soleus H-reflex tests, used in the clinical neurophysiological studies, are described in chapter 2. The relationship between abnormalities of soleus H-reflex test results and the clinical involvement of the limb under study is reported in chapters 3 and 4. In chapter 3 soleus H-reflex test results in dystonic patients with and without involvement of the leg are compared. Chapter 4 presents how soleus H-reflex test results relate to the presence and disappearance of dystonic features in patients with dopa-responsive dystonia. In chapter 5 studies are presented that investigate whether soleus H-reflex test results relate to the origin of dystonic movements. The results of soleus H-reflex tests obtained in patients in whom dystonic features were apparently due to peripheral trauma, are compared with those in patients with a presumed central origin of dystonic symptoms. These results are also compared with those obtained in a group of healthy controls, who were asked to mimic the dystonic posture. Chapter 6 shows the results of soleus H-reflex tests obtained in patients in whom dystonic features were temporarily relieved by electroacupuncture. In chapters 7, 8 and 9 various clinical features in dystonia
patients are described. In chapter 7 a clinical feature helpful in diagnosing patients with writer's cramp is presented. In chapters 8 and 9, based on some patient histories, some problems are discussed that may occur in the diagnostic procedure of a psychogenic dystonic movement disorder.

References


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