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Frontiers between neurology and psychiatry

Motor functional neurological disorders: An update



B. Garcin ^{a,b,*}

^aNeurology department, Memory Center, Avicenne University Hospital, AP-HP, 125, route de Stalingrad, 93009 Bobigny, France

^bUPMC UMRS 1127, Inserm U 1127, CNRS UMR 7225, Institut du cerveau et de la moelle épinière (ICM), 75013 Paris, France

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ABSTRACT

Motor functional neurological disorders (FNDs) are motor symptoms not explained by a lesion or related to a known dysfunction of the central nervous system, yet functional imaging studies suggest the presence of a genuine brain dysfunction. With this common disabling condition, there is a particular need for collaboration between neurologists and psychiatrists. Neurologists can search for positive clinical signs to make the diagnosis, which can then be followed by an explanation of the disease, whereas psychiatrists can look for psychological factors and psychiatric comorbidities in order to deliver appropriate treatment. Such a multidisciplinary approach is important, particularly with the participation of neurologists, psychiatrists, physiotherapists and psychologists. If necessary, additional treatments such as transcranial magnetic stimulation (TMS), hypnosis and sedation may be proposed.

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1. Introduction

Functional neurological disorders are neurological symptoms that cannot be explained by a lesion or related to an identified dysfunction of the central nervous system (CNS). The name that should be given to these symptoms has been the subject of debate for decades [1,2]. The term “hysteria”, proposed in the first three editions of the *Diagnostic and Statistical Manual of Mental Disorders* (DSM-I-III), suggests the disorder is the consequence of uterine dysfunction. For this reason, it was abandoned in the fourth edition (DSM-IV) published in 1994 and replaced by “conversion disorder” [3]. However, the term

“conversion” itself reflects Sigmund Freud’s theory that unconscious conflicts are converted into neurological symptoms, an idea that has never been scientifically demonstrated. Other terms frequently used by clinicians include “psychogenic disorders”, “somatization disorders”, “non-organic symptoms” and “medically unexplained symptoms”. The term “functional neurological disorders” (FNDs) is relatively acceptable to patients and is now widely accepted by physicians. It appears in DSM-V and best reflects the current knowledge of the pathophysiology [4,5]. The term has also been proposed for the 11th revision of the International Classification of Diseases (ICD-11), which should be available

* Correspondence. Neurology department, Memory Center, Avicenne University Hospital, AP-HP, 125, route de Stalingrad, 93009 Bobigny, France.

E-mail address : beatrice.garcin@aphp.fr.

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in 2018 [6]. For these reasons, this is the term used in the present review.

The clinical presentation of FNDs is heterogeneous and includes motor deficits, movement disorders, sensory deficits, non-epileptic seizures, language disorders and/or swallowing impairment. The present report mainly focuses on FNDs presenting with motor symptoms (mFNDs), such as motor deficits and movement disorders.

2. Epidemiology and Challenges

Studying the epidemiology of FNDs is especially difficult most notably because of the changes in diagnostic criteria for FNDs over the past several decades [7]. According to epidemiological studies, the incidence of FNDs varies from 5 to 12/100,000 population [8]. Their prevalence, calculated from population registries, is approximately 50/100,000, yet in clinical practice, FNDs are frequently observed. In a consecutive series of 3781 patients seen in Scottish outpatient neurological clinics, 30% had medically unexplained symptoms and 6% met the criteria of conversion disorder according to DSM-IV. These figures are similar to those described in eight smaller patient series [9], thereby demonstrating the high prevalence of FNDs in neurology clinics.

Thus, FNDs represent an important socioeconomic challenge, and there is now clear evidence that their economic burden is considerable. In the above-mentioned Scottish cohort of 3781 patients, those with medically unexplained symptoms were more likely to not be able to work because of their health issues, and they also received more health benefits for their handicap than patients with “organic” neurological diseases [10]. In the present author’s experience at the Salpêtrière University Hospital in Paris, more than half of the 482 patients with mFNDs received disability-related benefits because of their symptoms [11], while a UK study estimated that the annual cost of somatization disorders represents 10% of public health expenses, translating to a total annual sickness cost of £ 14 billion [12]. FNDs also constitute a human challenge: given the similar severe motor problems, the quality of life of an FND patient is as altered as in an “organic” disease such as Parkinson’s [13].

The management of these disorders is also challenging. First, there is no consensus or recommendations on how to manage FNDs. Second, there is a lack of training of neurologists and psychiatrists, partly due the position of FNDs at the boundary between neurology and psychiatry. In a survey to assess how much neurologists like to manage specific neurological diseases, FNDs came out last in a list of 20 of them. This ranking reveals how uncomfortable neurologists feel with a diagnosis of FND [14,15]. Moreover, as there is no identified lesion in the CNS, the prognosis of patients with FNDs is generally poor. A meta-analysis involving 2069 patients with mFNDs found that symptoms persisted or worsened in 50% of cases at the 1-year follow-up. After 7 years of follow-up, only 24% were in remission [16]. In that meta-analysis, the best predictors of a good outcome were short duration of disease, early diagnosis and high patient satisfaction with their care.

3. Pathophysiology considerations

Patients suffering from FNDs have motor symptoms that often mimic organic deficits, making it logical to wonder such symptoms are not real, but being feigned by patients instead. However, if they are considered “real” experiences for patients and not being voluntarily produced, it then becomes reasonable to suspect that a cerebral dysfunction is underlying these symptoms. The emergence of functional imaging techniques has allowed both hypotheses to be tested, and an increasingly large number of such studies have been performed over the past 20 years (for a review, see Aybek and Vuilleumier [17]).

3.1. Functional imaging

Functional imaging studies show that cerebral activation in patients with mFNDs differs from the cerebral activation in patients simulating the same deficit or movement disorder [18–21]. These results suggest that the symptoms are not voluntarily produced, but in fact reflect genuine brain dysfunction. Although functional imaging studies are heterogeneous in terms of their experimental paradigms and patients’ characteristics, some results remain relatively constant. In particular, patients with mFNDs consistently show hypoactivation in both cortical and subcortical motor pathways, with no recruitment of the prefrontal regions usually associated with voluntary motor inhibition; in contrary to what is observed with simulated deficits [19]. In addition, some authors have evidenced hypoactivation of the right temporoparietal junction [18,22], a region often associated with the sense of agency. This is in agreement with theories that place disruption of the sense of agency as a key triggering factor [23]. Furthermore, some authors have noted greater amygdala activity in arousing stimuli [24] and abnormal recruitment of the ventromedial prefrontal cortex (PFC) [25], possibly in relation to emotion dysregulation.

3.2. Theories of pathophysiology

Why FND symptoms arise has been a source of debate since antiquity [26]. Until recently, it was accepted that psychological factors are the main causes of symptoms [27]. However, such a causal relationship between psychological factors and symptoms has never been demonstrated, and psychological factors have recently been removed from the diagnostic criteria of FNDs. However, recent functional magnetic resonance imaging (fMRI) studies have suggested a dysregulation of emotional processing in patients with FNDs [25,28,29]. New cognitive theories have also recently emerged: most notably, Edwards et al. [30] proposed that psychological or physical trauma might induce the formation of an abnormally strong predisposition. In such cases, the conjunction of an individual’s beliefs and expectations about how the brain and body can go wrong, a disrupted sense of agency for movement and an abnormal switching of attention to the body/symptoms could lead to functional symptoms.

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