34 Functional (Psychogenic) Movement Disorders: Phenomenology, Diagnosis and Treatment

A. Macerollo and M.J. Edwards

34.1 Introduction

Functional (psychogenic) movement disorders (FMD) are part of the spectrum of functional neurological disorders [1], one of the commonest reasons for a neurological outpatient consultation [2, 3].

There is limited consensus on what to call functional/psychogenic disorders. 'Psychogenic' is the most commonly used phrase. However, this presupposes a psychological formulation can always be made which is not supported by current evidence [4]. It also suggests that biological factors are not relevant, in contrast to the biopsychosocial model of all other illnesses. The term psychogenic has poor acceptance amongst patients [5]. We prefer the term 'functional', for reasons that are outlined in detail here [6], although there are opposing views, summarised here [7].

FMD occupy a grey area between neurology and psychiatry characterized by complex pathophysiology, difficulties in making and communicating the diagnosis to the patients and management for which there is a limited evidence base and which often needs the cooperation of different specialists. This difficult situation has been termed a 'crisis for neurology' [8].

The commonest FMD are tremor and dystonia that together account for about 70 % of patients, followed by myoclonus and gait disturbance. Functional parkinsonism, tics and chorea are rare [9].

Women are more often affected than men, and the mean age at onset ranges from 37 to 50 years [9, 10]. FMD are characterized by level of disability and impairment in quality of life equivalent to that reported by patients affected by Parkinson's disease [11]. Consequentially, considerable health and social care costs are associated with this group of patients [12].

In this chapter, we provide an update on the current knowledge on clinical phenomenology, diagnosis and management of FMD.

34.2 Phenomenology

In broad terms, FMD are disorders of movement which, commonly, implies additional movements but in some cases also a reduction of movement associated with abnormal postures.

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Gait disturbance is a common accompaniment to FMD. A variety of gait patterns are described; the most common is the *astasia/abasia* known as 'tightrope walker gait' or 'walking on ice gait'. The patient shifts their centre of gravity quickly from side to side when walking without falling, thus showing excellent balance despite their subjective complaint. Furthermore, in contrast to other patients with poor balance, patients with functional gait disorder tend to walk with a *narrow base* and may show a *dramatic response to Romberg's test*. Other features of functional gait disturbance are *hesitation*, where small forward and backward movements of the leg may be observed while the feet seem to stick on the ground, *fluctuations* in gait impairment and *excessive slowness of movements*, resembling walking in slow motion. A monoplegic dragging gait is commonly seen in patients with functional leg weakness [13–15].

Functional tremor (FT) most frequently involves *hands and arms*, but functional tremor of the head, legs and palate can also occur. Patients typically direct visual attention towards their affected limb, and tremor often worsens significantly *during examination* [16]. FT is typically present *at rest*, *on posture and on action*. It is characterized by *variability in amplitude and frequency*, clear *distractibility and entrainment* with clinical manoeuvres [17] and *pauses with ballistic movement* [18, 19]. Furthermore it may paradoxically *worsen with loading and restraint* [20].

Functional dystonia (FD) is the second most common presentation in patients with FMD [9]. The typical presentation is with *fixed abnormal postures*, rather than the typical mobile postures of organic dystonia. These commonly have an *unusual distribution* given the age at onset (e.g. leg dystonia in an adult) [21, 22]. *Severe pain*, similar to that present in chronic regional pain syndrome type 1 (CRPS1), is commonly associated [23]. The usual *precipitating factor is a minor peripheral trauma*, and the term 'causalgia-dystonia' was previously coined [24]. The spread of abnormal postures to other body parts without further injury is seen. Important features of FD are the *absence of task or position specificity*, the *absence of a sensory geste* and *poor response to botulinum toxin* [21]. *Limbs* are usually involved, but FD affecting the neck and face is also commonly reported and has specific patterns of muscle involvement [22, 25].

Functional myoclonus (FM) is characterized by *marked daily variability in distribution* and frequency of jerks. It is typically exacerbated by movement [26]. Jerks may be *dramatically stimulus sensitive*, exaggerated compared to the stimulus sensitivity seen in organic myoclonus and associated with a *lack of habituation* [27]. If the limb involved is restrained by the examiner, the myoclonus may worsen and spread to other parts of the body [26].

A small group of patients present with *functional parkinsonism* (FP). The average age at onset is 47 years, *younger* than typical Parkinson's disease (PD). *Symptoms are typically symmetrical*, and tremor, showing features typical of FT, is prominent; rigidity is similar to *voluntary stiffness*, and the resistance may paradoxically decrease with performance of synkinetic movements of an opposite limb. The progressive fatiguing and slowing of repetitive movements seen in true bradykinesia is absent. Postural stability assessment using the *pull test typically leads to dramatic loss of balance and falls* [28, 29].

34.3 Diagnosis

The diagnosis of FMD should not be regarded as a diagnosis of exclusion, nor a diagnosis made on the basis of co-existence of a movement disorder with psychological disorder. Ideally, the diagnosis should be a positive diagnosis, based on positive features from history, examination and appropriate investigations.

Formal *diagnostic criteria* have been developed by Fahn and Williams (the most widely used) [30], recently revised by Gupta and Lang (Table 34.1) [31]. These criteria combined, when appropriate, with additional electrophysiological and imaging testing provide different levels of certainty of diagnosis.

Table 34.1 Fahn-Williams and Gupta-Lang criteria for diagnosis of functional movement disorders [30, 31]

Category	Criteria
Fahn-Williams criteria	
Documented	Persistent relief by psychotherapy, suggestion of placebo response, which may be helped by physiotherapy, or absence of abnormal postures when patient is unobserved
Clinically established	The posture is inconsistent over time, or it is incongruent with classical dystonia (e.g. impossibility to move limbs on request and great resistance during passive movements), plus one of the following manifestations: other psychogenic signs, multiple somatizations, or an obvious psychiatric disorder
Probable	The dystonia is incongruent or inconsistent with typical organic dystonia or there are psychogenic signs or multiple somatizations
Possible	Evidence of an emotional disturbance
Gupta-Lang criteria	
Clinically definite	Documented category
	Clinically established category
	Incongruent and/or inconsistent movement disorders (even without the additional presence of psychogenic signs, multiple somatisations or psychiatric disturbance)
Laboratory-supported definite	Electrophysiological tests proving a FMD (primarily evidence of premovement potentials before jerks or data from tremor studies)

The key clues in the *medical history* that suggest the diagnosis of FMD are an *abrupt onset* with rapid progression to *maximum severity*, previous episodes of movement disorders in the same or different body part with complete or partial *remissions* associated with *paroxysmal exacerbations*. The phenotype is often *inconsistent over time*, with complete change in the pattern of abnormal movements. Previous *somatisations* are common. Physical precipitating events (illness, injury) are common before the onset of functional symptoms [32].

Positive findings on physical examination commonly relate to the normalisation of symptoms with the distraction of attention, for example, the change in tremor with entrainment manoeuvres (see above). Other signs also represent normalisation of movement by distraction of attention, for example, *Hoover's sign* [33–35]. *Fluctuation* in the severity of symptoms during examination is typical.

Positive diagnosis in FMD may be helped by appropriate investigations. *Electromyography* with *accelerometry* can be used to assess tremor frequency and to evaluate the effect on tremor frequency of tapping at a different frequency or during ballistic movements with another limb [36, 37]. It is useful also in the presence of jerks to exclude an organic myoclonus, which is characterized by brief bursts (<50 ms) of muscle activity [38]. *EEG/EMG back averaging* is useful to detect a Bereitschaftspotential (BP), which is a slow-rising wave in the EEG before voluntary movements [39]. This is often present before functional myoclonic jerks. *Nuclear imaging*, DAT-scan (dopamine transporter imaging) [29] and 123I-Ioflupane SPECT [40] are helpful in patients with suspected FT or FP. Such tests need to be considered in association with clinical examination since such scans are normal in many organic tremor (e.g. essential and dystonic tremor) and parkinsonian (e.g. dopa-responsive dystonia, drug-induced parkinsonism) conditions.

34.4 Treatment

The best start to the treatment of FMD is the clear and effective communication of the diagnosis. Failure to do so often results in multiple referrals, repeated unnecessary diagnostic tests and harmful treatment including surgical interventions [21, 41].

There is no good evidence to support the use of drugs in FMD. Intrathecal baclofen has been proposed as helpful for patients with fixed dystonia, but this evidence is uncertain, and treatment-related complications are common [42, 43]. Antidepressants may be helpful and should be

used when functional symptoms are associated with anxiety or depression [44].

Psychological therapy, in the form of psychodynamic psychotherapy [45] or cognitive behavioural therapy [46], has some limited evidence for benefit. Physical rehabilitation has face validity as a treatment for motor symptoms, and there is some evidence to support this approach from two previous case-control studies [47, 48] and one randomised trial [49]. Transcranial magnetic stimulation has been reported to have positive impact in patients with FMD, but it sees most likely given the protocols used that this operates via a placebo effect [44, 47]. For severely affected patients, a multidisciplinary inpatient treatment approach may be of benefit, but evidence on which patients are most likely to benefit is limited [50].

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