Abstract

Functional movement disorders are relatively common neurological manifestations that are not related to a known neurological or a medical diagnosis. We aim to present an updated overview of this topic and present important diagnostic tips and pitfalls in the management of these potentially puzzling presentations. Particularly, as such psychosomatic disorders could simulate known disorders, including tremor, paralysis, seizures, or ataxia. This often results in excessive, unnecessary and costly investigations, which may in turn further enhance the sick role and complicates the management. Early recognition and prompt intervention are needed to prevent social and academic disruptions, and hence a debilitating outcome.

Keywords: Functional; Movement; Disorders; Update; Review

Introduction

Functional movement disorders (FMD) are relatively common neurological manifestations that are not related to a known neurological or a medical diagnosis including abnormal involuntary movements that can be affected by distraction or prompting [1-3]. Such psychosomatic disorders can take many forms and could simulate known disorders, including tremor, paralysis, seizures, or ataxia. If the proper diagnosis and management are not instituted promptly, they may become debilitating and result in significant social and academic disruptions [4]. It is important to note that these patients are not fabricating the signs and symptoms (malingering), despite of absence of an organic pathology. Functional movement disorders often result in excessive, unnecessary, and costly investigations, which may in turn further enhance the sick role and complicates the management [5].

How common of a problem are FMD?

FMD affect both adults and children. There is a relative shortage in pediatric data mainly the result of diagnostic difficulties [6]. The estimated pediatric prevalence ranges between 1-4 per 100,000 [7,8]. Motor weakness and abnormal movements were the most common symptoms among children [7]. In another study, pseudoseizures constitute 25% of children with FMD [9]. All ages are affected and patients as young as 4 years were identified [10,11]. Such cases often present repeatedly to pediatric neurology. In a study by the National Health Service, one third of 3781 patients who presented to neurology clinics had symptoms that were categorized as ‘somewhat’ or ‘not at all’ explained by organic disease [12]. A recent study from Saudi Arabia found up to 8% of referrals with psychiatric complaints were diagnosed as functional [13]. Such patients often have repeated emergency room visits or hospital admissions [14]. In general, FMD are more common in females with a 3-4:1 ratio among children aged 12-14 years [15-17].

How can we explain the occurrence of FMD?

Some studies suggested functional changes in the brain including reduced regional cerebral blood flow in the thalamus and basal ganglia contralateral to the involved region using single photon emission computerized tomography (SPECT) [18,19]. These changes were transient. Another study using electroencephalography (EEG) and functional magnetic resonance imaging (MRI) also showed abnormal changes [20-
These included increased dorsolateral prefrontal cortex activity, reduced hippocampal and para-hippocampal activity, increased activity in the right supplementary motor area, and temporoparietal junction. Other studies revealed decreased right inferior frontal cortex activity, and increased connectivity between the right supplementary motor area SMA and left amygdala [23,24]. Finally, a study using Voxel-based morphometry in hemiparetic motor conversion, increased cortical thickness in the bilateral premotor cortex and a trend towards increased grey matter volume were found [25]. Such data suggests that there are genuine changes, often transient, that occur in the brain as a result or in association with FMD.

Clinical Spectrum

FMD usually presents as a single movement disorder affecting the dominant extremities, however, complex movements or gait disorders are also common [26-28]. Children with functional disorders are expected to make a full recovery, as the conversion symptoms last no longer than 3 months typically [29]. The majority of them have a good long-term outcome with a rare recurrence of symptoms in general [30-32]. Triggering factors or precipitating events are often identified [33]. These include physical injury, infection, surgery, or other significant stressful life events [34]. Irritability, depressive mood, and anxiety often coexist among these children [35]. Others become anorexic with considerable weight loss [36].

Aids in diagnosis

Suggestion and placebo challenge can aid in distinguishing psychogenic from organic disease. Its use is controversial because it may affect the trust and doctor-patients relationship. However, when it is used as part of comprehensive treatment, it can play a role in the diagnosis [37]. Electrodiagnostic testing can provide additional supportive evidence for the diagnosis of FMD. Surface electromyography (EMG) for tremor analysis and accelerometry can identify features associated with functional tremor which include entrainment of tremor, simultaneous activity in agonists and antagonists muscles, increased tremor amplitude and frequency with weight loading of the involved limb and variability in tremor frequency [38,39]. However, diagnoses should not be made solely on frequency variability, as there is often an overlap between organic and functional tremor [40,41]. In contrast, a tremor occurring in different muscle groups at different frequencies or tremors faster than 11Hz usually indicates an organic etiology [42]. Electrodiagnostic techniques can also be helpful in differentiating functional myoclonus from true myoclonus. Features associated with functional myoclonus on surface EMG include abnormally long and variable latency between the stimulus and the onset of movement; prolonged myoclonic burst duration with a well-organized triphasic pattern of agonist and antagonist muscle activation. In organic myoclonus, the myoclonic jerk has a short latency of less than 70 ms. A pattern consistent with voluntary movement is suggestive of functional myoclonus [43].

Diagnosis

A careful understanding of the nature of FMDs is recommended for diagnostic accuracy. Due to an attributed similarity, it has been documented that some organic disorders have been mistaken for functional disorders in the past, examples of which are both Tourette's syndrome and task-specific focal dystonias. Thereby, an acknowledged proficiency of neuroanatomy and neurophysiology is in order and must be kept in mind to correctly interpret abnormal presentations of recognized diseases. For instance, organic disorders may present with some controlled movements such as tics, tardive dyskinesias, parkinsonian rest tremors and chorea. Others are characterized by an abrupt onset followed by remission [44]. Organic paroxysmal dyskinesias may be passed for a psychogenic etiology; however, the clinical features of functional paroxysmal movement disorders allow a differentiation [45]. On the other hand, it should also be noted that some diseases exhibiting movement disorders might initially present with psychiatric dysfunction, such as Huntington's disease and Wilson's disease. Although they are separate diseases, organic and functional movement disorders may coexist in some patients. This is also rarely seen in cases of epilepsy. In order to allow for further advancement in the accuracy of diagnosis, it has been suggested to follow the guidance of clinically based and phenotype-specific FMD diagnostic criteria. This will include the signs and symptoms from a clinical examination in addition to the findings from history taking. Many times an experienced neurologist with a specialized background on movement disorders is needed in order to make the diagnosis of FMD's clinically and not as a diagnosis of exclusion [46]. One incorrect notion, however popular, is that functional movement disorders are a diagnosis of exclusion when in fact obtaining a thorough personal, neurological, and family history as well as detailed examination is necessary to make the diagnosis (Table 1). In some cases, the need for diagnostic testing is needed [47].

Clues to the diagnosis in specific types of FMD

FMD can take the shape of known movement disorders, tremor being a most common presentation followed by dystonia, myoclonus, and ataxia [48]. Tics have been reported, along with other possible but less common movement disorders such as psychogenic Parkinsonism, psychogenic facial movements, some ocular manifestations, and palatal tremor. The following specific discussion will help differentiate the functional types:

<table>
<thead>
<tr>
<th>History</th>
<th>Sudden onset of symptoms, followed by a plateau</th>
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<tbody>
<tr>
<td></td>
<td>Subside if patient was unknowingly watched</td>
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<td>Presence of secondary gain</td>
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<td>Spontaneous remission</td>
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<tr>
<th>Examination</th>
<th>Variable nature of the movement</th>
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<tr>
<td></td>
<td>Increase by watching and if not, they decrease</td>
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<tr>
<td></td>
<td>Lack of anatomic or physiologic correlations</td>
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<td></td>
<td>Continuous unremitting or out of proportion</td>
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<td></td>
<td>Intervention to initiate or stop the movements</td>
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<td></td>
<td>Entrainment</td>
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<td>Deliberate slowness of the movement</td>
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<th>Therapeutic outcomes</th>
<th>Improvement upon the use of placebo</th>
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<tr>
<td></td>
<td>Positive recovery after sessions of physiotherapy</td>
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<td></td>
<td>Pertinent medications show no significant impact</td>
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This article is available in: http://pediatrics.imedpub.com/archive.php
**Tremor:** Variability in the tremor character was greater than 90%, while amplitude and frequency variance was observed in all subjects, frequently males [49,50]. There may be some tonic co-activation at tremor onset. The tremors usually consisted of resting, postural, and kinetic components along with a selective but not task specific disabilities. It changes by distraction often pause during contralateral ballistic movements. Neurological examination was indecisive, and drug treatment was ineffective. A nice trick is to elicit an increase in tremor amplitude with weight loading. Children with psychogenic tremor may be more likely to be diagnosed with an FMD based on clinical grounds and without extensive laboratory or imaging investigations [51].

**Dystonia:** Features of organic dystonia are summarized in Table 2. Functional dystonia is often painful with associated non-anatomic sensory changes and false weakness [52]. Most patients have movements that are inconsistent with those of dystonia. Four distinctive functional dystonia phenotypes have been identified including; 1) fixed wrist and finger flexion often with relative sparing of the thumb and index fingers, 2) fixed foot plantar flexion and inversion, 3) unilateral lip and jaw deviation, and 4) torticolis with ipsilateral shoulder elevation and contralateral shoulder depression [41]. On the other hand, organic dystonia can be misdiagnosed as psychogenic due to the intermittent nature and variation in the presentation.

**Myoclonus:** The movements are typically segmental rather than focal or generalized in nature, occurring at rest and often exacerbated by voluntary movement [53]. It tends to be more common in females and when axial it is often flexion in character. The jerks occur in a non-stereotyped multifocal manner with no clear pattern.

**Tics:** These are trickier as tics are often triggered by emotional or psychological stress [54]. Tics in the setting of a psychogenic illness have a sudden dramatic onset of jerking movements usually involving one or both arms, head, and neck [55,56]. These tics are not preceded by premonitory urges, were not suppressible, and were not stereotyped. In addition, older age of onset and lack of family history should raise the suspicion even further. Thus, the movements differed from true tics in several important diagnostic features.

**Ataxia:** The presence of knee buckling or astasia-abasia alone suffices to confirm a diagnosis of primary functional gait disorder [57]. An alternative presentation includes an atypical or bizarre gait such as, tightrope walking, trembling, stiff-legged gait, dragging, scissors gait, gradual fatigue, truncal myoclonic gait, and walking-on-ice gait [58,59]. The need to include at least one associated FMD in gait patterns other than knee buckling and astasia-abasia should protect against the incorrect diagnosis of functional gait in patients with bizarre but organic gait disorders.

**Chorea:** The diagnosis of isolated functional chorea, the rarest of functional phenotypes, cannot be made with clinically definite certainty because the key features of variability in speed and amplitude, entrainment, suppressibility, and changes in distribution can be seen in organic chorea [60]. In modern series of FMD, psychogenic chorea and athetosis have a relatively low prevalence of less than 12% [61].

**Psychogenic Parkinsonism:** Psychogenic Parkinsonism has been reported in several studies [62,63]. Tremor and bradykinesia are the two most common features observed in patients with psychogenic Parkinsonism. Tremor usually has the same features as psychogenic tremor without Parkinsonism. Bradykinesia in psychogenic Parkinsonism patients is usually unaccompanied by amplitude decrement or interruptions in repetitive movements as observed in Parkinson disease (Table 3). Features of psychogenic gait such as buckling of the knees and astasia–abasia may be observed [64]. Arm swing can be reduced or abolished in psychogenic Parkinsonism, sometimes with one arm held stiffly extended and adducted to the side. This feature can persist while running.

**Psychogenic facial movements:** Psychogenic facial movements can present as lip protrusion, hemifacial spasm, or blepharospasm. The most common pattern of movements was lateral or downward lip protrusion with an ipsilateral deviation of the jaw. In contrast to organic oromandibular dystonia, patients had a unilateral involvement, and no sensory tricks or speech [65]. In psychogenic hemifacial spasm, an earlier age of onset was noted with no simultaneous compensatory contractions of the frontalis muscle and a normal blink reflex [66-68].

**Ocular manifestations:** Convergence spasm consisting of episodes of lens accommodation with ocular convergence usually causing diplopia is evoked mainly by horizontal ocular pursuit [69,70]. Rarely, functional upward gaze paralysis accompanies such convergence spasm.

**Palatal tremor (palatal myoclonus):** Palatal tremor (palatal myoclonus) is characterized by abnormal rhythmic contractions of palatal muscles (tensor veli palatini muscle). Voluntary control of palatal muscles seems necessary for the development of psychogenic palatal tremor [71]. Entrainment and distractibility of palatal movements in psychogenic patients are key features [72]. Patients with psychogenic palatal tremor were younger than patients with symptomatic palatal tremor, more frequently female, had a history of a precipitating event, and frequently complain of ear clicking and multiple somatizations [73].

**Misdiagnosis**

In general, physicians are hesitant to make the diagnosis of FMD resulting in significant diagnostic and management delays. Pediatric patients have reportedly undergone uncalled for investigations and treatments without benefit. Precise
diagnosis is of crucial significance to avoid such unnecessary delay. Identifying pitfalls associated with each diagnostic criterion may be one step forward towards achieving a definitive diagnosis. Any movement disorder that does not pose irregular organic manifestations, may be falsely interpreted, as a clinically definitive FMD should the presence of pain, exposure to a disease model, potential for secondary gain, and multiple somatizations is verified [74].

**Management and Outcome**

These patients need a multidisciplinary treatment plan that includes the cooperation of both the neurologist and the patient to get a better outcome. Since the patients have difficulty in accepting the idea of psychogenic origin disorder, they prefer the term "Functional" movement disorders [75]. The diagnosis is followed by the treatment of the disorder, with no further testing. Evaluations by different specialties are recommended to determine the precipitating factors important in designing the treatment plan. Supportive treatments include physical therapy, psychotherapy, physical exercise, and self-relaxation techniques [76-80]. Occasionally, antidepressants are used in patients with depressive symptoms [81,82]. Despite all interventions, some patients continue to have symptoms [83]. Better outcome is noted in those with more acute problems, with symptoms of shorter duration, with an identifiable stressor, and in those who lack a coexisting organic psychopathology [84-86].

### Table 3 Difference between psychogenic Parkinsonism and Parkinson’s disease.

<table>
<thead>
<tr>
<th></th>
<th>Psychogenic Parkinsonism</th>
<th>Parkinson’s disease</th>
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<tbody>
<tr>
<td>Onset</td>
<td>Sudden onset, maximal deficit soon after</td>
<td>Insidious onset</td>
</tr>
<tr>
<td>Course</td>
<td>Non-progressive course with fluctuating, transient or permanent remission</td>
<td>Progressive evolution</td>
</tr>
<tr>
<td>Rigidity</td>
<td>Active resistance to passive movements on examination (paratonia). Absent cogwheel phenomena</td>
<td>Cogwheel phenomena</td>
</tr>
<tr>
<td>Effect of distraction</td>
<td>Decrease in amplitude or disappear</td>
<td>Increase in amplitude</td>
</tr>
<tr>
<td>Effect of reinforcement</td>
<td>Rigidity diminishes</td>
<td>Rigidity diminishes</td>
</tr>
<tr>
<td>Freezing</td>
<td>Absent</td>
<td>Common</td>
</tr>
<tr>
<td>Arm posture while walking</td>
<td>Extended in adduction, held stiffly at side (this posture may persist while running)</td>
<td>Partially flexed</td>
</tr>
<tr>
<td>Tremor</td>
<td>Rest, postural, kinetic</td>
<td>Rest, postural, kinetic</td>
</tr>
<tr>
<td>Bradykinesia (repetitive movement)</td>
<td>Slowness without amplitude decrement</td>
<td>Progressive slowness with amplitude decrement</td>
</tr>
</tbody>
</table>
References


3. Charcot JM, Goetz CG (1887) Charcot the Clinician. The Tuesday lessons. Excerpts from nine case presentations on general neurology delivered at the Salpêtrière Hospital in 2: 881-989.


