The Diagnosis of Functional Movement Disorder

Functional movement disorder is diagnosed using positive signs, allowing a clinician to "rule-in" the diagnosis.

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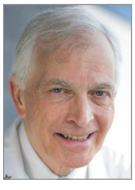

Functional movement disorder (FMD), encompassing abnormal movements and weakness, is a common subtype of functional neurologic symptom disorder (FND), with a motor-dominant presentation seen in 61% of a cohort of people with FND. FMD accounts for 2% to 20% of referrals to movement disorder clinics.[1,2] Women are more frequently affected than men, and the mean age of onset is 37 to 50, although FMD may present at any age. Individuals with FMD experience quality-of-life impairments to a similar, if not greater, degree as those with Parkinson disease (PD).[2] FND has a significant socioeconomic impact, with a mean 35% of untreated patients not working and 26% receiving disability benefits.[1]

Symptoms of FMD are subjectively experienced as involuntary, despite historical and examination findings that demonstrate preserved neurologic function. FMD is diagnosed on the basis of positive signs indicating internal inconsistency, such as tremor distractibility or the Hoover sign in functional weakness and incongruence with known neurologic etiologies (Table 1).[3] When discussing FMD, there is often an emphasis on diagnostic features of individual symptoms, but it is important to note that complex and overlapping phenomenologies are frequent, with multiple movement phenotypes coexisting and evolving over time.[1] Functional movement symptoms are often part of a complex syndrome that includes other functional symptoms of pain, fatigue, and cognitive fog; these nonmotor symptoms are often associated with a greater impact on quality of life.[1] Additionally, FMD frequently coexists with other neurologic disorders, either as risk factors or triggering events, with a large multicenter study finding neurologic disorders in 22% of people with FMD.[4] Thus, a comprehensive assessment of individuals presenting with abnormal movement is necessary for the diagnosis of FMD.

Etiology and Pathophysiology

A single, unifying etiology for FMD is not yet known. Importantly, psychologic factors are no longer considered causative to the development of FMD. Rather, FMD is a complex neuropsychiatric disorder best conceptualized using a biopsychosocial framework in which heterogeneous predisposing factors render the nervous system more vulnerable to the development of FMD, which then emerges in the context of precipitating events, with perpetuating factors strengthening these symptoms over time (Table 2).[5]

Recent research has sought to understand the neural mechanisms by which functional movements may emerge. Candidate processes relevant to the development of FMD include abnormalities in self-directed attention, illness-related expectations, sense of agency for self-generated movements, motor learning and plasticity, and the limbic-motor interface. Given the fundamental features of FMD, it is hypothesized that an underlying disturbance of brain function allows voluntary movement to be perceived by the patient as involuntary, and elucidating this process/mechanism is the key to understanding the development of these symptoms.[2] Based on functional neuroimaging and neurophysiology studies, a model has been proposed with FND resulting from the interaction between higher-order influences (eg, attention to self and expectation) and bottom-up limbic influences (eg, trauma or arousal), which together influence basic motor functions.[6]

Clinical Assessment

The diagnosis of FMD is a process involving a careful history and examination for positive signs to rule in the diagnosis, while considering the differential diagnoses of functional presentations. Positive signs for FMD phenotypes are described in detail in Table 1. There are common general features seen in the history and examination that apply across all FMD phenotypes, and additional points that are relevant for specific phenomenologies.

TABLE 1. POSITIVE SIGNS FOR THE DIAGNOSIS OF FUNCTIONAL MOVEMENT DISORDER

Phenotype	Positive signs	Description
General examination features in functional movement disorder	Variability	Abnormal movement patterns change over time (eg, amplitude, frequency, distribution)
	Distractibility	Abnormal movements resolve during tasks requiring attention to be directed elsewhere
	Enhancement with attention	Abnormal movement emerges or worsens when attention is drawn to it
	Motor inconsistency	Movement impaired at some times, preserved in others; inconsistent performance on examination and times when patient is not being actively examined
	Incongruency	Clinical findings discordant with known neurologic disease
	Suggestibility	Atypical movement elicited or markedly exaggerated with verbal instructions/suggestions or maneuver from examiner (eg, tuning fork test)
Functional weakness	Hoover sign	Weakness of hip extension in paretic leg resolves with testing hip flexion power in other leg
	Hip abductor sign	Weakness of hip abduction in paretic leg improves with testing hip abduction power in other leg
	Finger abduction sign	Weakness of finger abduction improves with testing contralateral finger abduction power
	Give-way weakness	Limb collapses with light touch or appreciated normal strength suddenly collapses
	Drift without pronation	While testing for pronator drift, the arm drifts down without pronation
	Co-contraction	Agonist and antagonist muscles contract simultaneously, resulting in little movement
Functional tremor	Variability	Changes in tremor characteristics (frequency, direction, pattern) over time (including induced by distracting maneuvers) and with position
	Entrainment	Tremor changes to the same frequency of a cued rhythmic movement
	Cocontraction	Tremor caused by simultaneous contraction of agonist and antagonist muscles
	"Whack-a-mole" sign	With restraint of affected body part, involuntary movement emerges or worsens in other body part
	Common features of functional tremor	Rest = posture = action; absent isolated finger tremor; knee flexed and foot plantar flexed (heels lifted from the floor)
Functional cranial dystonia	Functional facial spasms	Unilateral downward lip pulling, ipsilateral jaw deviation, ipsilateral platysma contraction
	Functional blepharospasm	Tonic eye closure, constant bilateral eye closure with lack of prominent muscle activity; contraction of corrugator and procerus muscles without orbicularis oculi involvement
	Functional hemifacial spasm	Lack of spasms in sleep; asynchronous involvement of upper and lower face; bilateral involvement of lower face (often synchronous or alternating); lack of "other Babinski sign" (synchronous contraction of orbicularis oculi and frontalis muscles)
Functional fixed dystonia	Hand posture	Flexion of digits 3-5, sparing pincer function
	Foot posture	Foot inversion and plantar flexion, flexion of toes
	Cervical posture	Torticollis, ipsilateral laterocollis, ipsilateral shoulder elevation, contralateral shoulder depression
	Resistance to passive movement	Increasing muscle contraction opposing attempts to perform passive movements
Functional paroxysmal dystonia	Variability	Variable phenomenology, duration, frequency of attacks; can be induced by suggestion
	Overlap with functional seizures	Prolonged aura and symptoms of dissociation
Functional jerks and myoclonus	Variability	Variability in duration, pattern muscle involvement
	Entrainment	Jerks change to the same frequency of a cued rhythmic movement
	Variability in stimulus-sensitivity	Variable and long latency from stimulus to jerk; jerk triggered by threat of stimulus; habituation with repeated stimulation
Functional tics	Common phenomenologic features of functional tics	Large-amplitude arm movements, self-injurious movements, wide repertoire of complex and unstereotyped "tics" including odd words/phrases, coprolalia without shortening to starting syllable of word considered inappropriate that often occurs in Tourette syndrome (eg, "shi" or "fu") and may be directed and context dependent (swearing vs true coprolalia), interferes with voluntary actions or speech; premonitory urge and ability to suppress not present

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TABLE 1. POSITIVE SIGNS FOR THE DIAGNOSIS OF FUNCTIONAL MOVEMENT DISORDER (*continued*)

Phenotype	Positive signs	Description
Functional gait disorder	Variability	Inconsistent abnormalities in gait and stance
	Monoplegic leg dragging gait	Leg is externally rotated and dragged behind on the medial forefoot
	Excessive slowness	Slow gait despite normal toe-tapping performance
	"Walking on ice" gait	Reduced step height and stride length
	"Tight-rope" gait	Narrow base, arms abducted, oscillations of center of gravity, demonstrating intact balance
	Uneconomical postures	Walking pattern requires considerable effort and balance to maintain posture
	Knee-buckling	Falls are only occasionally present
Functional parkinsonism	Hypokinesia without decrement	Excessive slowness of movement without decrement of speed or amplitude
	Paratonia	Variable resistance to passive manipulation of a joint; may decrease with distraction (lacks typical enhancement with contralateral limb movement of true rigidity)
	"Huffing and puffing"	Apparent great effort in performing simple movements
	Normal speed during "automatic" tasks	May be noted when not formally examining patient

Adapted with permission from Lidstone SC. Functional movement disorders. In: Eichler AF, ed. UpToDate. 2022. https://www.uptodate.com/contents/functional-movement-disorders

General Features on History

Patients often experience a sudden onset of symptoms, frequently occurring at the time or shortly after a physical injury (often quite minor), illness or other neurologic symptom.[5] Symptoms usually progress to maximal severity rapidly or are maximal at onset, in contrast to the typical symptom progression seen in many other movement disorders.[2] If more than 1 functional symptom is present, it is common that multiple symptoms accumulate then wax and wane over time, including pain, fatigue, and cognitive symptoms. A careful history will differentiate between symptom accumulation over time vs progression of an individual symptom. It is common for those with FMD to notice variability in symptom severity, often described as "good days and bad days," and occasionally have spontaneous remissions. Others describe a static course, with symptoms constantly present, although within that, variability is present to some degree, which may not be initially evident to the patient or clinicians. Some with FMD experience exclusively paroxysmal abnormal movements, whereas others may complain of paroxysmal worsening of more persistent movements; both groups may experience unusual triggers for the paroxysms. Most experience their symptoms worsening with pain, fatigue, stress/anxiety, or after exertion.

General Features on Examination

Clues to the diagnosis should be assessed throughout the entire consultation, from observations made in the waiting room to the end of the clinical interaction. The key features of most FMD phenotypes include variable abnormal movements throughout the assessment that are distractible and enhance when attention is drawn to them.[7] Mixed movement disorder phenomenologies are common, and paroxysmal attacks may occur in the clinic, occasionally triggered with suggestion.

Functional Tremor

Tremor is the most frequent FMD presentation, occurring in 40% to 50% of persons diagnosed with FMD.[8] Functional tremor can occur in any body part, often with hands and arms most affected. Several clinical examination maneuvers are helpful for diagnosing functional tremor, particularly when assessed in combination.[2,9,10] Unlike in other causes of tremor, distraction maneuvers including contralateral limb ballistic movements, mental tasks, patterned contralateral finger tapping and the cranial nerve examination, result in temporary improvement or abatement of functional tremor. Importantly, longstanding functional tremor can be resistant to distraction, requiring unexpected or complex tasks for adequate distraction. Tremor entrainment is a key examination finding with high specificity for functional tremor. When a patient is asked to follow an examiner's set frequency of finger tapping with the nontremoring hand, pure entrainment of tremor frequency, a shift in tremor frequency, or relative maintenance of the frequency at the expense of poor distracting task performance (eg, simple rapid side-to-side tongue movements) may be seen. Head tremor may demonstrate entrainment with a set frequency of pronation/supination movements of the arms or tongue, and functional palatal tremor often entrains with finger tapping.

Functional Dystonia

Functional dystonia is the second most common presentation of FMD.[8] There are 3 distinct presentations that have been described: functional cranial dystonia, functional fixed dystonia, and functional paroxysmal dystonia.

TABLE 2. POSSIBLE ETIOLOGIC FACTORS IN THE DEVELOPMENT OF FUNCTIONAL MOVEMENT DISORDER

Predisposing factors	Precipitating factors	Perpetuating factors
• Female sex • Low socioeconomic status; financial strain • Comorbid neurologic conditions • Comorbid psychiatric disorders • Health anxiety and somatic vigilance • Physical, sexual or emotional trauma; neglect • Major losses such as bereavement or divorce • Maladaptive personality traits	• Physical injury or surgery • Preceding illness • Accidents (eg, motor vehicle accident) • Autonomic hyperarousal event (eg, panic attack) • Dissociation event • Interpersonal conflict • Job loss or employment-related stressors • Significant acute loss event	• Chronic pain and/or fatigue • Chronic medical conditions • Physical deconditioning • Entrenched abnormal motor programs[a] • Invalidation by healthcare system; stigma • Maladaptive illness beliefs; lack of diagnostic agreement • Anxiety and hypervigilance around symptoms • Avoidance patterns • Interpersonal or work-related stressors • Pending litigation • Unconscious secondary gains

[a]**Longstanding functional movements (especially tremor) may develop refractory motor programs that show little or no change with distracting maneuvers on examination. Further, these entrenched programs can be resistant to distraction-based treatments and may require additional therapeutic techniques.** Adapted with permission from Lidstone SC, MacGillivray L. The biopsychosocial formulation for functional movement disorder. In: LaFaver K, Maurer CW, Nicholson TR, Perez DL, eds. *Functional Movement Disorder: An Interdisciplinary Case-Based Approach*. Humana Press; 2022

Functional Cranial Dystonia. When functional dystonia affects the cranial region, the typical pattern is lower face involvement with unilateral downward lip pulling, ipsilateral jaw deviation, and ipsilateral platysma contraction (Figure).[11,12] Symptoms are often paroxysmal, with tonic spasms that may be painful, but sustained posturing also occurs. Other common presentations may resemble blepharospasm and hemifacial spasm.[11]

Functional Fixed Dystonia. Uncommon in other causes of dystonia, functional fixed dystonia with reduced joint mobility is a well described presentation of FMD, particularly when the fixed posture is present from onset. Functional fixed dystonia often develops rapidly after minor peripheral injury. Typical postures observed when functional fixed dystonia involves the hand, foot, or neck are shown in the Figure and described in Table 1.[12] Pain is present in most cases, with some patients meeting diagnostic criteria for complex regional pain syndrome type I. Typical features of nonfunctional dystonia are not present, including overflow dystonia and response to sensory tricks.[13] Functional fixed dystonia is challenging to distract, although certain examination maneuvers such as asking a patient to propel a swivel chair may briefly normalize the posture of a dystonic foot. Observation during nonexamination movements is invaluable for noting variability of posture (eg, when patients are removing or putting on socks and shoes).

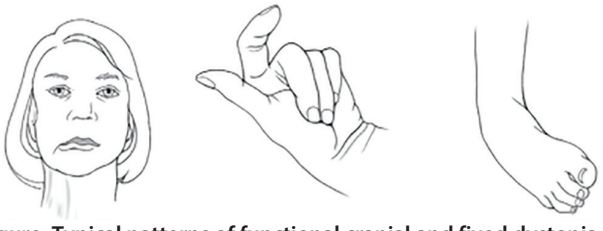

Figure. Typical patterns of functional cranial and fixed dystonia. Reproduced with permission from Espay AJ, Aybek S, Carson A, et al. Current concepts in diagnosis and treatment of functional neurological disorders. *JAMA Neurol*. 2018;75(9):1132-1141.

Functional Paroxysmal Dystonia. Paroxysmal attacks of functional dystonic posturing affecting the extremities and/or trunk, often combined with other hyperkinetic movements including tremor and jerks, are a common presentation of functional paroxysmal dystonia. In contrast to primary (ie, genetic) forms of the paroxysmal dyskinesias, which present in childhood or adolescence, the onset of functional paroxysmal dystonia is in midlife.[14]

Functional Jerks and Myoclonus

Functional jerks and myoclonus account for 13% to 20% of FMD.[8] Propriospinal myoclonus is a well-defined presentation of functional myoclonus characterized by flexor arrhythmic jerks involving the trunk, hips, and knees, occasionally with associated vocalization. These jerks are often stimulus sensitive and worsen in a supine position.[15]

Functional Gait Disorder

Functional gait disorder may be isolated but more commonly presents as FMD that interferes with gait. In 1 study, 36.6% of persons with FMD had an abnormal gait, whereas only 5.7% had an isolated functional gait without other functional movement abnormalities.[16] In mixed FMD with a gait disorder, the abnormal gait often is incongruent with what is expected from the seated exam. For example, a distractible tremor without ataxia may be present while seated, and the gait exam reveals an unsteady, wide-based gait (ie, ataxic) without tremor. Variability is a key feature in diagnosing a functional gait disorder, so careful observation of gait cycle abnormalities with walking, tandem walking, heel/toe walking, jogging, and going up and down stairs is useful, as is employing distractions such as conversing while walking.

There are several challenges when it comes to diagnosing a functional gait disorder, considering the wide spectrum of presentations. A common pitfall is to assume a "bizarre" gait pattern is functional, which is often not the case.[17] Despite this, some recognizable functional gait patterns and additional helpful signs have been described (Table 1).[3,18]

Functional Tics

Functional tics have generally been considered a less common presentation of FMD,[8,19] which may reflect that making a diagnosis of functional tics is challenging considering the overlapping features with nonfunctional tics, such as suggestibility and distractibility. Individuals with nonfunctional tics will regularly appreciate some or all of their movements (and sounds) as a volitional response/capitulation to an involuntary urge, in contrast to the lack of self-agency typical of FMD. Interest in functional tics has increased since the beginning of the COVID-19 pandemic because of the explosive onset of functional tics linked to social media use (See *Functional Tic Disorder: a Pandemic in a Pandemic* in this issue).[19] The presence of isolated motor and vocal tics with onset in childhood or adolescence and comorbid psychiatric symptoms, including obsessive-compulsive behaviors or attention-deficit disorders, is generally more supportive of tic disorders. Conversely, historical features suggesting functional tics include an abrupt or explosive onset, an identifiable triggering event, onset in adulthood, comorbid anxiety and/or depressive disorders, lack of family history of tic disorders, and presence of other FND symptoms.[19]

Functional Parkinsonism

Functional parkinsonism is rarely reported, with a recent systematic review identifying 120 reported cases of functional parkinsonism without comorbid PD.[20] Functional tremor is a common coexisting symptom, occurring in 72% of those with functional parkinsonism. The diagnosis of functional parkinsonism can be challenging, particularly in elders, given the increased incidence of PD. Further confounding the situation, there is some evidence that patients with an underlying synucleinopathy may have a higher than expected incidence of FND.[20] The presence of a prodrome suggestive of a synucleinopathy (ie, constipation, rapid eye movement [REM]-sleep behavior disorder, anosmia) may aid in differentiating functional parkinsonism from PD.

Investigations

FMD is a clinical diagnosis and investigations should be carried out as appropriate. Brain or spinal imaging may be necessary to rule out comorbid neurologic diseases.[17] In challenging cases, electrophysiology can be a useful adjunct in making a laboratory-supported diagnosis, particularly in functional tremor and functional myoclonus.

Electrophysiology

An electrophysiologic test battery using surface EMG and accelerometry has been validated for the diagnosis of functional tremor, with 89.5% sensitivity and 95.9% specificity.[9] Positive signs for diagnosis of functional tremor include the following: brief pause in the tremor during contralateral ballistic movements, incorrect tapping performance at various frequencies, tremor suppression/entrainment/frequency shift with tapping task, coherence in bilateral arm tremor, tonic muscle coactivation before onset of tremor, increased tremor amplitude with loading, and demonstration of suggestibility.[9,21]

Surface EMG is useful in the diagnosis of functional jerks and myoclonus, which demonstrate long and variable burst duration, variability in order of muscle recruitment, variable and long latency of onset when stimulus-sensitive, and habituation with repeated stimulation.[21] EMG-EEG back-averaging may demonstrate a premovement potential, known as the *Bereitschaftspotential*, supporting a diagnosis of functional jerks and propriospinal myoclonus.[3]

Presynaptic Dopamine Imaging

Imaging of the nigrostriatal dopamine system (ie, dopamine transporter [DaT] single-photon emission computed tomography [SPECT] or positron emission tomography [PET] with several different ligands) may be helpful if there is diagnostic uncertainty during evaluation of suspected functional parkinsonism. Dopamine imaging is expected to be normal in the case of functional parkinsonism, and thus an abnormal scan may be useful in differentiating PD, with the caveat that an abnormal scan does not rule out the presence of coexisting functional parkinsonism.[3,5]

Diagnostic Criteria

Although diagnostic criteria for FMD have evolved over the last few decades, reflecting a growing understanding of the underlying neurobiology, criteria remain limited in scope and applicability. In everyday clinical practice, a history suggestive of FMD, the presence of positive signs on examination, and appropriate investigations rule-in the diagnosis of FMD. The *Diagnostic and Statistical Manual of Mental Disorders, 5th edition* (DSM-5) criteria for FND are the most often used by clinicians, emphasizing "clinical findings that show evidence of incompatibility between the symptoms and recognized neurologic or medical conditions."[22] The removal of the requirement for an identifiable psychologic stressor was a major shift from the DSM-IV, with the relevant clinical implication that psychologic factors are no longer considered etiologic for the development of FND. This change is in opposition to the earlier, formal diagnostic criteria specific to FMD, which emphasized the role of psychologic factors.[7,23,24] The Gupta-Lang criteria added "laboratory-supported definite," based on electrophysiologic findings, shifting the diagnosis toward clinical findings.[7] This

reflects an unresolved and substantial gap in the field: DSM-5 criteria are the most widely used by neurologists, but are not within the traditional scope of expertise of mental health professionals because of the requirement for comprehensive knowledge of the differential diagnoses of neurologic presentations. Developing updated diagnostic criteria based on the clinical assessment and a holistic understanding of the underlying neuropsychiatric features of the disorder is essential.

Providing the Diagnosis

The clinical assessment allows a clinician to diagnose FMD and also serves as a unique and important opportunity to provide patients with an understanding of how FMD causes their symptoms.[25] After a comprehensive assessment of the range of symptoms present, including features beyond the motor symptoms (eg, pain, fatigue, cognitive fog, and sleep disturbance), a clinician should follow a typical structure for explaining a diagnosis. This begins with naming the problem, by explaining "you have a functional movement disorder," and further explaining that this is a common and real cause of neurologic symptoms. It is helpful next to explain how functional symptoms arise, emphasizing malfunction or disconnection within the nervous system, as opposed to damage. Various analogies can be helpful, including that this is a software rather than hardware malfunction, disconnection between intact wiring of the nervous system and intact motivation, or abnormal motor programs being learned and overriding normal/automatic motor programs. It is extremely useful to show patients and family members positive signs, for example demonstrating the Hoover sign or tremor distractibility, because this not only helps patients understand how the diagnosis was made, but demonstrates how symptoms arise and are potentially reversible.[25] In addition, some positive signs lend themselves well to treatment suggestions that can be provided by the neurologist at the bedside or in the clinic; for example, patients can be encouraged to develop their own distraction techniques to control their tremor similar to the maneuvers shown to them by the neurologist in the exam. Discussion around why functional symptoms arise is best done from a standpoint of risk factors for the development of FMD, as opposed to direct causation. Finally, providing patients with reliable educational resources, such as www.neurosymptoms.org, is essential.

Conclusion

FMD is a common cause of abnormal movements seen in neurology clinics, often part of a complex syndrome including chronic pain, fatigue, and cognitive symptoms, with key positive signs supporting a rule-in diagnosis. Ultimately, a careful assessment of patients presenting with abnormal movement, including examination for positive signs, is necessary for the diagnosis of FMD. ■

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