Functional Neurological Disorder in Pediatrics: Diagnostic Considerations

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ABSTRACT

Functional neurological disorder (FND) is a common diagnosis of varied neuropsychiatric symptoms presenting to pediatric healthcare settings, including primary, urgent and subspecialty care. A key diagnostic shift appearing in the DSM-V is that FND is no longer a diagnosis of exclusion; rather, a rule-in diagnosis based on suggestive elements of symptom presentation. This article reviews diagnostic criteria, clarifying features, risks, and prognostic factors. This is the first in a series of six articles on FND and will introduce an FND case that will be examined in each subsequent article in the context of their more specific subject matter.

KEYWORDS: functional neurological disorder, FND, functional neurological symptom disorder, FNSD, conversion disorder, pediatrics, non-epileptic seizures

CASE

BH is a 12-year-old female with a history of an orthopedic procedure two years ago, following a poorly healed arm fracture, who presented to an outside hospital with nausea, poor oral intake and dehydration leading to admission. In addition to the orthopedic history, BH had a negative endocrine work-up for short stature. Pertinent family health history includes anxiety in mother and younger sister. BH lives with her parents, twin brother, older sister, and younger brother. She is in 8th grade, an A student, and described as a "perfectionist" and "the caretaker of the family" by her parents. She experienced the loss of a grandparent recently.

On Review of Systems (ROS), BH reported frequent stomach/headaches and "mild" social anxiety. Parents report BH has recently been more isolated, spending less time with family or playing with siblings. The physical exam was noncontributory on initial presentation. During her hospitalization, BH developed acute onset of arm and leg paralysis, intermittent memory loss, regressed speech, and periodic staring.

- What are the diagnostic considerations?
- What would help clarify the diagnosis?

INTRODUCTION

Functional neurological disorder (FND), also known as conversion disorder, is a diagnosis describing motor, sensory, or cognitive symptoms resulting from impaired nervous system functioning in the absence of structural pathology. FND has been described in both developed and developing countries,[1-6] with an estimated incidence of 1.3/100,000 pediatric patients, appearing more commonly in females.[7] Motor dysfunction is the most common presenting symptom in children (weakness, abnormal movement), followed by functional (non-epileptic) seizures.[8] FND is the second most common diagnosis presenting to outpatient neurology clinics,[9] comprising 16% of outpatient neurology visits[10] and 23% of new patients.[11] Additionally, functional (non-epileptic) seizures may account for up to 30% of admissions to an Epilepsy Monitoring Unit (EMU).[12] Another study found an average of three patients per week presented to a large children's hospital emergency department with a diagnosis of FND over a three-month period.[13]

DIAGNOSIS

In 2013, the Diagnostic and Statistical Manual of Mental Disorders 5th Edition, Text Revision (DSM-5-TR) updated its classification of the somatic symptom predominant disorders group, of which FND is a subcategory, for clarity and ease of use by providers at initial encounters. Key changes include paired nomenclature of "Conversion Disorder" with "Functional Neurological Symptom Disorder" (FNSD or FND), FND no longer being considered a diagnosis of exclusion, clarification that stress or trauma may be present but are not required for diagnosis, and destigmatizing language.[14] The specific DSM-5-TR diagnostic criteria now include the following[15]:

A. One or more symptoms of altered voluntary motor or sensory function.

B. Clinical findings provide evidence of incompatibility between the symptom and recognized neurological or medical conditions.

C. The symptom or deficit is not better explained by another medical or mental disorder.

D. The symptom or deficit causes clinically significant distress or impairment in social, occupational, or other important areas of functioning or warrants medical evaluation.

Broad symptom categories include motor, sensory, functional seizures, and mixed symptoms, with motor symptoms and functional seizures being the most common in pediatric patients.[15] Providers may include diagnostic modifiers from one of these categories specifying symptoms present at the time of encounter. Available modifiers are weakness or paralysis, abnormal movements, swallowing symptoms, speech symptoms, attacks or seizures, sensory loss, special sensory symptoms (i.e., visual, olfactory, or hearing), or mixed symptoms.[15]

As with any childhood illness, a comprehensive clinical history and physical exam are important foundational elements in diagnosing functional illness. In the physical exam, a child may exhibit clinical features or positive signs that reinforce FND as a rule-in diagnosis. These key clinical features are contrasted between direct versus functional exam, incongruence of symptom presentation and anatomic distribution, and presentation elements with low specificity for known illness. For example, weakness on direct exam of ankle plantar flexion despite being able to walk on toes, numbness not following a specific dermatome, or fixed posturing versus mobile positioning in functional dystonia.[16] In the case of functional seizures, duration longer than two minutes, forcefully closed eyes, eyelid fluttering, side-to-side movements, fluctuating course, ictal crying, and memory of the event are associated with greater than 90% specificity.[17]

Some positive signs have a defined name and are utilized to rule in a particular functional symptom. The entrainment test can be a useful aid in diagnosing functional tremors. This test involves asking the child to perform a rhythmic task with the unaffected extremity and noting decreased tremors in the affected side. For children presenting with a unilateral lower extremity weakness, a positive Hoover's sign is suggestive of FND.[10] A positive Hoover's sign in unilateral functional weakness involves the examiner holding the child's heels while supine and eliciting no response on the affected side when asked to press heels into the exam table (or extending at the hip). The next step in Hoover's sign is holding the heel of the affected leg, asking the child to raise (or flex at the hip) the unaffected side while the examiner pushes down. A positive sign will result in the examiner experiencing the heel of the affected side pushing down into their hand.

WORK-UP

No biomarker or laboratory test exists to diagnose FND. However, in the setting of functional seizures, EEG is often used, and remains the gold standard for diagnosis. Capturing a functional seizure during EEG monitoring adds clarity to diagnosis and is particularly helpful in the setting of co-morbid epileptic seizures.

Prior to criteria revision and influenced by concern of misdiagnosis, patients would undergo extensive work-up before receiving a diagnosis of FND. With updated diagnostic criteria and data showing misdiagnosis around 4%,[18] if other testing is pursued, a judicious work-up is recommended. It is critical to prepare the child and family that further testing is expected to be normal; however, incidental findings may occur and may not correlate with presented symptoms. If concerns are unnecessarily heightened, potentially harmful interventions may be pursued.[19] Because co-morbidities can exist, suspected work-up for possible concurrent diagnoses should be completed with clear communication about the purpose of the evaluation. Likewise, if a patient already carries a neurologic diagnosis but is exhibiting an unexpected degree of disability, there should be consideration for an overlapping diagnosis of FND.

ETIOLOGY

Continuing research on FND has not shown a single causal element that leads to this disorder. However, a variety of biopsychosocial factors may predispose an individual to developing FND (see below). Research is emerging showing the influence of the following factors on a given individual's likelihood of developing FND: subjective distress tolerance and response, the impact of stress on the body's stress response system (HPA axis), and complex imbalance in neurocircuitry.[7,20]

PREDISPOSING FACTORS

While not known to be causal, there are multiple pre-disposing factors linked to diagnosis of FND. These include adverse or stressful life events, comorbid medical or psychiatric illness, and interpretation and experience of somatic symptoms. Estimates of comorbidity prevalence vary widely between studies: findings of co-occurring depression or anxiety range from 14% to 80%, while one systematic review found that 22% of patients with epilepsy carry a dual diagnosis of FND.[21,22] Additionally, existing psychological comorbidities may be worsened by the distress caused by FND symptoms. Understanding predisposing factors enhances our appreciation of risks inherent in vulnerable populations, such as gender non-conforming or neurodivergent individuals.[23] Transgender and gender non-conforming youth are particularly vulnerable to FND given potential struggles with bullying, varying degrees of family, social, and community supports, and higher rates of depression than cisgender youth.[24]

PROGNOSIS

Although remission rates for pediatric FND have varied widely, several factors have been identified as positive prognostic signs.[7] Positive prognostic factors, per the DSM-5-TR, are "short duration of symptoms, and acceptance of the

diagnosis," which includes parental acceptance of diagnosis. Other positive indicators include younger age, with pediatric patients having more favorable outcomes than adults with FND, and symptom presentation with functional motor symptoms or seizures are linked to better outcomes compared to sensory symptoms.[7] Potential negative prognostic factors, as referenced in the DSM-5-TR, include maladaptive personality traits, presence of comorbid physical disease, and receipt of disability benefits.

MANAGEMENT

Key components in managing FND include clarity in diagnosis, psychoeducation, and rehabilitative services in the context of multidisciplinary care (often involving neurology, psychology, psychiatry, and primary care).[7] When communicating the diagnosis to patients and families, it should be done in positive terms of what it is, instead of what it is not. Conveying a positive diagnosis of FND with details of the disorder, hope for treatment and potential for remission can have therapeutic benefits. Acknowledgement that symptoms are both real and treatable is foundational in joining with patients/families, and for successful treatment. For example, while results have varied between studies, as many as half of patients have resolution or improvement in the number of functional seizures after receiving an FND diagnosis and treatment.[25]

CONCLUSION

FND is a complex illness involving one or more neuropsychiatric symptoms and is often encountered in pediatric medical settings. Updated diagnostic criteria redefine FND as a rule-in diagnosis versus a diagnosis of exclusion and incorporate key features on physical exam or positive signs. Prompt diagnosis, psychoeducation and multidisciplinary treatment are effective in improving outcomes. Collaboration between neurologists, psychiatrists, and other specialists is essential for effective management.

CASE UPDATE

Due to the acute onset and severity of symptoms, BH underwent an extensive normal work-up including labs, lumbar puncture, brain MRI, with and without contrast, and a consult with neurology. Additional testing included a normal video EEG as BH began having intermittent episodes of "passing out" with variable jerking of her upper and lower extremities. In the context of a reassuring workup and normal EEG with episode capture, BH was given a diagnosis of functional neurological disorder with mixed episodes. As her functioning was significantly impaired, she was admitted to a medical/psychiatric inpatient program where she began to work with a multidisciplinary team.

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Disclosure

Authors have no conflicts of interest relevant to this article to disclose.

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