The role of the Neurologist in Functional Neurological Disorders

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Outline

• What are functional neurological disorders?
• Neurology or Psychiatry
• Functional disorders in the neurology clinic – cases
• What causes it, how do we diagnose it, treatment?
• Examples of medicolegal cases
What are functional neurological disorders?

‘a common experience [for the patient] was to feel dismissed by the neurologist as having something “all in the mind,” often accompanied by not so subtle suggestions of malingering, and to be sent to the psychiatrist who would respond, equally unhelpfully, “this patient has nothing psychiatric wrong” or even “are you sure the diagnosis is correct?”

[Functional Neurological Disorders in Handbook of Clinical Neurology: Edited by Mark Hallett, Jon Stone, Alan Carson]
Terminology - the progression

- Hysteria – used for centuries
- Conversion disorder
- Psychogenic – 20th Century
- Medically unexplained symptoms
- Functional – 19th / early 20th Century and again now
- Somatisation disorder
- Dissociative neurologic symptoms disorder
Conversion disorder (Functional Neurological Symptom Disorder)

- One or more symptoms of altered voluntary motor or sensory function
- Clinical findings provide evidence of incompatibility between the symptom and recognised neurological or medical conditions
- The symptom or deficit is not better explained by another medical or mental disorder
- The symptom or deficit causes clinically significant distress or impairment in social, occupational, or other important areas of functioning or warrants medical evaluation.
6B60 Dissociative neurological symptom disorder

Parent
Dissociative disorders

Description
Dissociative neurological symptom disorder is characterized by the presentation of motor, sensory, or cognitive symptoms that imply an involuntary discontinuity in the normal integration of motor, sensory, or cognitive functions and are not consistent with a recognized disease of the nervous system, other mental or behavioural disorder, or other health condition. The symptoms do not occur exclusively during another dissociative disorder and are not due to the effects of a substance or medication on the central nervous system, including withdrawal effects, or a Sleep-Wake disorder.

Exclusions
- Diseases of the nervous system (B900-8E72)
- Factitious disorders (8D50-8D5Z)

All Index Terms
There are no index terms associated with this entity
### Psychiatric Disorders
- Mood disorders
- Personality Disorders
- Schizophrenia

### Neurological Disorders
- Mood disorders complicating Neurological disease e.g. Parkinson’s disease
- Tourette’s syndrome

#### Functional Neurological Disorders

#### Disorders that have moved to Neurology
- Writer’s cramp dystonia
- Autoimmune encephalitis
OCCASIONAL PAPER

How psychogenic is dystonia? views from past to present

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‘In the everyday world of the clinic, psychiatrists are distinguished from other medical specialists not because they are concerned with “minds” rather than “bodies”, but because they focus on complaints appearing in people's thoughts, perceptions, moods, and behaviours rather than their skins, bones, muscles and viscera … The diagnostic process may be difficult, but causal explanations are always complex and depend on the physician's capacity to evaluate issues ranging from intermediary metabolism (a “body” issue) to interpersonal misunderstanding (a “mind” issue). Psychiatric concerns thus extend from the ultrastructure of the body to the relationship of groups of minds within a social context.’

McHugh & Slavney in *The Perspectives of Psychiatry*
In the clinic… patient 1

- 29 year old woman
- Last well 8 years earlier
- Soon after delivery of second child, severe left leg tremor, spread to whole body without altered awareness; lasted 5 minutes
- Four years earlier woke with right arm shaking, chest tightness and symptoms of panic, tried to stand and legs gave way. Ambulance to A+E. No abnormality found. Not right since.
- Intermittent episodes of legs giving way (10min in the park with children on one occasion)
- Other symptoms:....
Dizziness, worse looking into light, intermittent flashes and dots in vision, whole body pain in particular face and neck, numbness left arm, face, groin, and sometimes legs, right hand shaking, tremors in arms, legs, tingling both legs.

Described feeling she is not quite there [dissociation], sometimes with chest pain

No difficulty with bladder control

No alcohol excess, giving up cigarettes, family history of multiple sclerosis
In the clinic… patient 1

- Examination:
  - Normal apart from..
  - Give-way weakness left arm
  - Distractable tremor right hand
  - Positive Hoover’s sign on the left
  - Reflexes symmetrical, sensation and cerebellar testing normal
The approach.....

- Explain the approach to neurological disorders
- Investigate early and thoroughly
- Here MRI brain, cervical spine, range of blood tests including vitamin B12 level (borderline low) and vitamin D (borderline low)
- Meet again and reassess
- Follow up consultation
  - Demonstrate Hoover’s sign
  - Explain the diagnosis
  - Website / support groups
  - Follow up and refer if needed
In the clinic… patient 2.. The importance of a positive diagnosis

- Background… 2003 letter
- Neurologist number 8
- 70 year old woman with symptoms since late 20s
- Fatigue, dizziness, tripping and difficulty walking
- Deteriorated significantly after a hysterectomy at age 44
- Symptoms: difficulty walking uphill, climbing stairs, loss of balance, tripping, facial pain, pins and needles / sharp stabbing pain generally, cramp, fatigue, inability to lift or carry objects, feeling that she is moving in slow motion, sensation of water thrown at her, vibration in her back, stabbing sensation in her eye.
- Neurological examination normal apart from positive Hoover’s sign bilaterally, give-way arm weakness, and a bizarre gait.
- Several consultations and many normal investigations later we settled on the diagnosis of functional neurological disorder
Videos demonstrating several functional neurological signs
<table>
<thead>
<tr>
<th>Distinguishing semiologic or exam features</th>
<th>Psychogenic nonepileptic seizures</th>
<th>Epileptic seizures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Emergence out of EEG-confirmed sleep</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>Concurrent tongue biting (severe, side of tongue) and urinary incontinence</td>
<td>Rare</td>
<td>Common after GTC</td>
</tr>
<tr>
<td>Ictal dystonic posture with contralateral automatisms</td>
<td>Not present</td>
<td>Occurs in mesial TLE</td>
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<tr>
<td>Ictal figure-of-four sign</td>
<td>Not present</td>
<td>Occurs in TLE</td>
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<tr>
<td>Ictal fencing posture</td>
<td>Not present</td>
<td>Occurs in mesial FLE</td>
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<tr>
<td>Ictal grasping (gripping of an object with one hand or both hands)</td>
<td>Rare</td>
<td>Occurs in FLE and TLE</td>
</tr>
<tr>
<td>Postictal stertorous breathing</td>
<td>Not present</td>
<td>Common after GTC</td>
</tr>
<tr>
<td>Postictal nose rubbing</td>
<td>Not present</td>
<td>Occurs in TLE</td>
</tr>
<tr>
<td>Impaired corneal reflex</td>
<td>Not present</td>
<td>Common after GTC</td>
</tr>
<tr>
<td>Extensor plantar response</td>
<td>Not present</td>
<td>Common after GTC</td>
</tr>
<tr>
<td>Closed eyelid during peak of ictus</td>
<td>Very common</td>
<td>Rare</td>
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<tr>
<td>Gradual onset and prolonged duration</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Undulating motor activity</td>
<td>Common</td>
<td>Very rare</td>
</tr>
<tr>
<td>Asynchronous limb movements</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Side-to-side head shaking</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Ictal or postictal whispering/stuttering</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Ictal signs of emotional distress (e.g., grimacing, weeping)</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Pelvic thrusting</td>
<td>Sometimes</td>
<td>Rare</td>
</tr>
<tr>
<td>Memory recall for period of unresponsiveness</td>
<td>Sometimes</td>
<td>Rare</td>
</tr>
<tr>
<td>Resisted eyelid opening</td>
<td>Common</td>
<td>Very rare</td>
</tr>
<tr>
<td>Guarding of hand dropping over face</td>
<td>Common</td>
<td>Rare</td>
</tr>
</tbody>
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Modified from Benbadis and LaFrance (2010).
EEG, electroencephalogram; GTC, generalized tonic-clonic seizures; TLE, temporal-lobe epilepsy; FLE, frontal-lobe epilepsy.
Approach to a patient with functional neurological disorder

- Often clues in GP letter or early on
- Open ended start
- ‘When were you last 100% well?’
- Drain the symptoms dry
- Examination
- Read old notes and look for clues in the past
- Investigate in detail early on
- Review in a long appointment
Explaining the diagnosis and cause

- Long term outcome determined in large part by the manner and content of the explanation

- My approach:
  - Brief explanation of the nervous system
  - Demonstrate positive signs e.g. Hoover’s sign
  - Software versus hardware problem
  - Point out I’m not saying it is all the head or psychological
  - Explain the complexity of the software.. E.g. normal sensation perception when not focused on a limb
  - Explain what we think may trigger symptoms in some
  - Explain approaches to treatment (individualise)
  - Give information, websites such as [www.neurosymptoms.org](http://www.neurosymptoms.org)
  - See the person again!
Functional and Dissociative Neurological Symptoms: a patient’s guide

How to use this website...

This website is about symptoms which are:  • neurological (such as weakness, numbness or blackouts) • REAL (and not imagined) • and due to a PROBLEM with the FUNCTIONING of the nervous system, and NOT due to neurological disease. These symptoms have many names (including dissociative symptoms and conversion symptoms) but are often described as "functional symptoms" or "functional disorders". Symptoms like these are surprisingly common but can be difficult for patients and health professionals to understand. This website, written by a neurologist with many years of experience in this field, is aimed at providing information for patients and their families.
And what about malingering / factitious disorder?

- Rare in clinical practice but doctors not good at detecting
- Finding patients tampering with tests, or clearly functioning in a way that is incompatible with their clinical presentation
- Confession
- Tests of inadequate effort e.g. on cognitive testing
- I would involve psychiatry for advice before making a diagnosis of malingering or factitious disorder
And the cause? The importance of why me, why now?
The etiology of functional symptoms (functional neurologic disorder: FND)

Precipitating
FND is a disorder of sensorimotor processing in which erroneous health beliefs or expectations distort an, often noxious, somatosensory experience. This process is facilitated by misdirected and overly precise attention, anxiety, and dissociation. The symptom formation helps “make sense” of the amorphous somatic experience. The patient can be either consciously or preconsciously complicit in it.

Perpetuating
Once present, FND can be perpetuated by maladaptive behavioral responses, both operant and classic learning, mood disorder, and central nervous system plasticity.

Predisposing
Patients who have pre-existent mood/anxiety problems, excessive threat vigilance, or certain obsessive or rigid cognitive styles are more vulnerable; some of these risks may relate to the experience of abusive or aversive events currently, the recent past, or childhood. There is also a mild genetic risk and almost certainly other risk factors as yet unknown.
Fig. 6.3. Model of how involuntary movements might be generated and how they might not acquire a sense of agency. The model speculates that excessive limbic activity leads to movement generation, but does not produce a normal feedforward signal. Although movement is generated in a normal fashion and feedback occurs, there is a mismatch between feedforward and feedback and agency is not generated.
Fig. 6. Functional connectivity of primary motor cortices. Regions showing an increase in correlated activity with the right primary motor cortex (M1) compared to the left M1 (depicted in red) and conversely (left M1 > right M1, depicted in green) in the normal controls, simulation condition, and conversion patient (threshold \( p < 0.001 \) uncorrected, \( k = 10 \) voxels). Motor connectivity was symmetrical and restricted to sensorimotor areas in normal and simulation conditions; but in the conversion patient, selective increases were found for right M1 connectivity with the precuneus and vmPFC, as well as with the right superior gyrus. Plots represent the parameter estimates (betas) for the rM1 > IM1 connectivity in the normal, simulation, and conversion conditions for precuneus (6 mm sphere centered on xyz = −3, −48, 63) and vmPFC (6 mm sphere centered on xyz = 3, 48, −12).
Treatment

- Explanation
- Physical / physiotherapy
- Perhaps psychological treatment
- Cognitive behavioural therapy
- (Transcranial magnetic stimulation)
- (Sedation)
The role of the Neurologist in clinical care

- FND makes up around 6% of neurology outpatient contacts
- Neurologists role:
  - Make the diagnosis
  - Explain the diagnosis to the patient in a collaborative and constructive manner
  - Initiate treatment (this starts from the point of taking the history)
  - Refer to patient information and support groups
  - Refer to Neuropsychiatry as appropriate
Prognosis

- Studies suggest generally unfavourable but this is dependent on multiple factors
- Young patients diagnosed early have a better prognosis
- Psychiatric comorbidities impact variably on prognosis
- Litigation has been found to be a negative predictor in some studies
The Neurologist in medicolegal ‘functional neurology’

- Similar to clinical, as an adjunct to a psychiatric review
- The neurologist to exclude other causes or more often separate non-functional and functional neurology
- Some examples from my experience:
  - 40 year old woman, developed loss of bladder sensation and inability to urinate spontaneously following a laparoscopy for abdominal pain. Then developed loss of lower abdomen and genital sensation. Normal investigations. One episode of post-operative urinary retention which was thought to be responsible for overdistention and bladder damage.
  - On review several features compatible with Fowler’s syndrome (functional bladder syndrome) and functional neurological disorder.
The Neurologist in medicolegal ‘functional neurology’

- A 25 year old male with multiple symptoms after a trivial head injury.
- More complex when the question arises when an injury has triggered a deterioration in someone with pre-existing functional neurological disorder.
- A 25 year old woman diagnosed with functional weakness or seizures who on examination did not have features of either, but rather a neurological explanation for symptoms that sometimes result in false positive signs of functional neurological disorder.
In conclusion

- Functional neurological disorders are common
- The understanding of the underlying cause and best management is evolving
- Neurologists are essential to clinical care in terms of diagnosis, investigation and management
- Long term management is frequently neuropsychiatric
- In medicolegal terms, neurologists identify functional neurological disorders and symptoms, but psychiatrists essential given the overlap with neurology, for providing a psychiatric perspective and excluding malingering / factious disorder