

Functional neurological disorders: acute presentations and management

Authors: Hannah R Cock^{A,B} and Mark J Edwards^{C,D}

ABSTRACT

Functional neurological disorders (FND) are common and associated with significant morbidity and healthcare costs. Patients with FND often present acutely, particularly with dissociative seizures (resembling epilepsy) or persistent weakness resembling a stroke. History and careful observation and examination are critical to diagnosis, as investigations will often be normal or non-contributory. The nature of convulsive movements in dissociative seizures often differs from that in epilepsy, and long duration of individual events, waxing and waning, closed eyes and high reported frequency in an apparently well individual are all suggestive. In those with stroke-like episodes, demonstration of normal power even briefly (eg Hoover's sign, 'give way' weakness) together with distractibility are positive physical features indicating a functional disorder. A positive diagnosis and clear non-judgemental explanation, backed up by reliable information sources associated with prompt onward referral to a neurologist can greatly reduce distress and ultimately improve outcomes.

Background

Functional neurological disorder (conversion disorder, FND) is one of the commonest diagnoses made in neurology clinics.¹ It is far from a benign disorder; it causes disability and impaired quality of life in the long term similar to that seen in people with multiple sclerosis² or Parkinson's disease³. Misdiagnosis and inappropriate treatment carries a significant risk of iatrogenic injury, morbidity and cost to patients and healthcare systems.^{4–6} Despite this, services specifically designed to help people with FND are very limited. The experience of many patients is an endless round of referrals, re-referrals and investigations, and if a diagnosis is made, it is often explained in a

Authors: ^Aprofessor of epilepsy and medical education, Institute of Medical and Biomedical Education, St George's University of London, London, UK; ^Bconsultant neurologist, Atkinson Morley Regional Neuroscience Centre, St George's University Hospitals NHS Trust, London, UK; ^Cprofessor of neurology, Institute of Molecular and Clinical Sciences, St George's University of London, London, UK; ^Dconsultant neurologist, Atkinson Morley Regional Neuroscience Centre, St George's University Hospitals NHS Trust, London, UK

manner which implies the symptoms are not real, not significant or are feigned. However, there has been a recent resurgence of clinical and research interest in FND among neurologists, neuropsychiatrists and allied health professionals. New approaches to diagnosis, diagnostic explanation and treatment backed up by epidemiological and pathophysiological work have begun to improve the evidence base for management of FND, bringing with it the possibility of improved outcomes for patients.

FND in A&E

FND commonly presents acutely and so patients often attend A&E at the onset of symptoms. Others, particularly those

Key points

Functional neurological disorders are common and associated with significant costs and risks for patients and healthcare services

Frequent emergency presentations include dissociative seizures and stroke-like episodes (functional weakness) and should be considered as differentials from the outset

Long duration of individual events, fluctuating course, closed eyes, later recall of items during the event and a high reported attack frequency in an apparently well individual are among features suggesting dissociative seizures rather than epilepsy

Hoover's sign, 'give way' weakness and distractibility, demonstrating a discrepancy between volitional and non-volitional motor function and recurrent episodes with normal imaging suggest functional weakness rather than stroke or other neurological disorders

A positive diagnosis based on the clinical features, clear explanation together with providing reliable sources of information and prompt onward referral improves patient experience and outcomes

KEYWORDS: functional neurological disorders, dissociative seizures, clinical features, diagnosis, management ■

with recurrent attacks of symptoms that resemble epilepsy (dissociative seizures [DS]), may be frequent attenders, and often inappropriately loaded with antiepileptic or even anaesthetic drugs if misdiagnosed. With the development of hyperacute stroke services, patients with stroke-like presentations of FND commonly enter the acute stroke pathway: about 9% of admissions to hyperacute stroke units have FND.

Making a positive diagnosis

The two commonest presentations to acute settings are with seizure-like episodes or with persistent motor symptoms that resemble a stroke. Diagnosis can be difficult in some situations, and there may be diagnostic uncertainty that can only be resolved by specialist review and/or performing investigations, but it is essential to consider the diagnosis of FND in the differential diagnosis and to look out for clinical indicators of FND during the patient's initial assessment, rather than as something that can only be entertained after all other conditions have been excluded.

History

In those with dissociative seizures, history from the patient and witnesses is invaluable and clear documentation of this (including video recording with appropriate consent) can significantly aid clinicians who might see the patient at a future date (eg in a first fit clinic). Key features of the event which can be helpful in distinguishing dissociative from epileptic seizures^{7–9} (although none are completely fool-proof) are summarised in Table 1. Clusters of features are more helpful than individual signs, and context is also relevant. Event frequency is higher in patients with DS, and recurrent admissions with apparent seizures, or daily

convulsive events suggest DS, especially if reported by a well and alert patient. A reported history of epilepsy should not stop consideration that the current attack was dissociative in nature: a prior misdiagnosis of epilepsy is not uncommon (with the time from seizure onset to diagnosis of dissociative seizures typically ranging from 3–8 years¹⁰), and a low proportion of people with epilepsy also have comorbid dissociative seizures. There is evidence that the manner in which people with dissociative seizures talk about their seizures is different from people with epileptic seizures, meaning that linguistic analysis can distinguish between dissociative and epileptic seizures with a reasonable degree of accuracy.¹¹

In those with functional motor symptoms, history of previous events of a similar nature can be helpful, along with information on previous investigations. A history of recurrent episodes of stroke-like symptoms with repeated normal imaging is typical of some patients with FND. Waxing and waning of symptoms is common in FND including (sometimes brief) episodes of complete resolution of symptoms. Patients may notice that these happen during times when they are distracted or movement occurs more automatically.

Examination

Examination in-between DS is typically normal, although some patients will have additional functional motor symptoms interictally. Examination during an attack may reveal resistance to gentle attempts to open the eyes, or shaking movements may alter if the limb is gently held or moved (for example, gentle restraint of arm shaking may lead to a dramatic increase in shaking or switch of shaking to another body part). Active alternating flexion and extension movements suggest DS, in contrast to clonic movements which typically have a brief active flexion phase and either tonic stiffness or relaxation in between. Sternal rubs, nail bed pressure and other noxious stimuli are not advisable – patients can be injured by such procedures, and for those patients who retain awareness during attacks but cannot respond, these procedures can be very distressing indeed.

In people with functional weakness, physical examination often reveals positive physical signs. The most well-known is Hoover's sign, where hip extension in a patient with unilateral functional weakness is weak when tested directly, but briefly returns to normal when triggered by contralateral hip flexion. This is a clear demonstration of 'normal wiring' of the nervous system despite the person's inability to access movement when attempting to carry out willed movements. Such signs can be usefully demonstrated to patients as part of the diagnostic explanation.¹² Functional weakness often has a collapsing or 'give way' quality where full power is generated for a second, but then disappears. Of note, people with acute pain often have a similar pattern of weakness. Functional facial 'weakness' is often caused by active contraction of muscles of one side of the lower face resulting in an apparent facial droop that is in fact caused by excessive muscle activity. Functional tremor can often be stopped temporarily by externally paced tapping movements of another limb.¹³ People with functional gait disturbance often have a 'walking on ice' pattern where the upper body lurches from side to side without falling – something which indicates an intact balance system. All these signs rely on the demonstration of a capacity for normal function within a body that the patient cannot make function when they are trying. This ties in with the common

Table 1. Clinical features helpful in distinguishing epileptic from dissociative seizures.^{7–9}

Favour dissociative seizures	Not useful discriminators
Long (>5 min) duration of individual events	Tongue biting (except significant lateral tongue biting)
Fluctuating course (waxing and waning)	Incontinence
Asynchronous rhythmic movements ^a	Gradual onset
Pelvic thrusting ^a	Non-stereotyped ^c
Side-to-side head/body movements during a convulsion	Flailing/thrashing movements
Closed eyes	Opisthotonus
Ictal crying	History of associated Injuries
Recall of items during event ^b	

^aCan be seen in frontal lobe focal seizures.

^bPatients often report being able to hear what is going on around them but not being able to respond.

^cClinical experience suggests that dissociative seizures are less stereotyped than epileptic seizures, but stereotyped attacks of itself does not argue strongly in favour of epilepsy. Features favouring epileptic seizures include prolonged post-event confusion and stertorous breathing.

finding of symptoms in general being better with distraction and dramatically worse during direct examination.

These features are all positive diagnostic features of FND because they demonstrate discrepancy between the presentation and the expected findings in other neurological disease processes, and normal basic function within the nervous system.¹⁴ They can help reduce dependence on normal investigations to make the diagnosis by exclusion. FND is not a diagnosis that should be made just because one can't think of another diagnosis or because tests are normal.

Diagnostic explanation

FND is often explained to patients as a 'psychological reaction' or as 'symptoms due to stress'. These explanations usually fail and result in patients feeling alienated, stigmatised and not-believed. The main reason for the failure of such explanations is that they take a risk factor for the development of FND (adverse life events) and turn it into the cause of the problem. Life event studies do show an excess of both distant and more recent adverse life events in people with FND,¹⁵ but many people have not experienced such phenomena, or they were events that happened years, sometimes decades previously. It is not unreasonable that someone might have trouble understanding how such an event could be linked to the recent emergence of symptoms. It is also the case that in common parlance (and indeed among many health professionals) the description of symptoms as 'psychological' suggests that they are trivial and not genuine. It also suggests that the diagnosis has been made on the presence of psychological factors, whereas it has been made on the presence of positive symptoms and physical signs. Indeed, making the diagnosis because the patient has a history of an affective disorder or trauma or conversely not making the diagnosis because the patient appears 'psychologically normal' are two very common sources of diagnostic error. Many people develop functional symptoms triggered by other health events such as infections, injuries and operations – it is useful to acknowledge the reality of such trigger factors while also explaining that the functional symptoms develop independently after being triggered and not because of nervous system damage due to the trigger.

Diagnostic explanation should instead follow the process of normal practice in other disorders.¹⁶

- 1 Tell patients what they have wrong with them: 'You have dissociative seizures'; 'You have a functional weakness'.
- 2 Tell them that this is something you recognise and believe in: 'This is a really common reason for people to get neurological symptoms'; 'I believe your symptoms – I don't think they are made up or put on'.
- 3 Tell them something about how it happens: 'The basic wiring of the nervous system is ok, but you can't get access to it in a normal way'; 'This is like a software problem not a hardware problem'. Showing people their physical signs such as Hoover's sign can be useful here.
- 4 Explain that, like many illnesses, we don't know exactly why it happens, but there are risk factors which include recent illnesses, recent stressors and past stressors.
- 5 Explain that it is a problem that can improve and get better, but that it can take time and treatment to achieve this.
- 6 Direct people towards reputable sources of information eg www.neurosymptoms.org, www.FNDHope.org, www.FNDAction.org.uk.

These simple things can make an enormous difference to people's experience of and trust in healthcare and are in their own right a treatment for FND. For example, in a cohort of people with dissociative seizures, 50% were regularly attending A&E. Diagnostic explanation resulted in a 74% drop in attendance in A&E.¹⁷

Further management

For a patient in dissociative status (ongoing/recurrent attacks without recovery), protecting them from injury (eg nursing on a mattress on the floor) is all that is needed, together with calm reassurance even if they appear unconscious.

Referral for neurological assessment, either acutely or in outpatients, is an essential next step. Depending on the patient and their comorbidities, additional assessment from neuropsychiatry can be helpful. Current best evidence for treatment of dissociative seizures is with specific psychological therapy such as cognitive behavioural therapy with a therapist who is familiar with dissociative seizures.^{18,19} People with functional motor symptoms commonly benefit from specialist physiotherapy,^{20,21} or, in more complex patients, specialist multidisciplinary rehabilitation.^{22,23} With treatment, many patients experience significant improvement in symptoms. A positive experience of care in emergency settings can greatly reduce distress and improve the likelihood of engaging with treatment at a later stage. ■

Conflicts of interests

Professor Hannah Cock reports in the last 3 years non-financial support from European Academy of Neurology; personal fees from Sage Pharmaceuticals Ltd, Eisai Europe Ltd, UCB Pharma Ltd, European Medicines Agency, from UK Epilepsy Nurse Specialist Association, non-financial support from Special Products Ltd. Outside the submitted work, Professor Edwards reports in the last 3 years honoraria from Merz Pharma and UCB, and the International Parkinson's Disease and Movement Disorders Society.

Author contributions

ME wrote the first draft. HC wrote the DS content, take-home messages and SAQs. Both authors edited the final manuscript.

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Address for correspondence: Professor Mark Edwards, St George's University of London, Institute of Molecular and Clinical Sciences, Cranmer Terrace, London SW17 0RE, UK. Email: medwards@sgul.ac.uk