

Assessment of patients with functional neurologic disorders

A. CARSON^{1*}, M. HALLETT², AND J. STONE³

¹Departments of Clinical Neurosciences and of Rehabilitation Medicine, NHS Lothian and Centre for Clinical Brain Sciences, University of Edinburgh, Edinburgh, UK

²Human Motor Control Section, National Institute of Neurological Disorders and Stroke, Bethesda, MD, USA

³Department of Clinical Neurosciences, Centre for Clinical Brain Sciences, University of Edinburgh, Edinburgh, UK

Abstract

We describe an overall approach and structure to the clinical assessment of the patient with a functional neurologic disorder. Whilst the primary purpose of the assessment is to make a diagnosis and develop a treatment plan, we believe the assessment also plays a key role in treatment in its own right, as it sets a tone and context for future clinical interactions. We aim to set up an atmosphere of collaboration based on taking the patient's problems seriously, and emphasizing that all facets of the patient's presentation – physical, psychological, and social – are of importance. Patients with functional disorders can be perceived as difficult to help and yet with the correct approaches we believe the consultation can be much more satisfying for both patient and doctor. Finally, we discuss and list some of the common diagnostic pitfalls in the assessment of functional neurologic disorders, looking at features that lead to erroneous diagnosis of neurologic disease (such as old age, *la belle indifférence*, and lack of psychiatric comorbidity) and an erroneous diagnosis of a functional disorder (such as “bizarre” gait in stiff-person syndrome).

INTRODUCTION

In this chapter we describe our general approach to clinical assessment; individual symptoms and signs in specific functional presentations are described in other chapters. Here we shall concentrate on an overall approach and structure to the assessment. This is a topic we have written on at length in other review papers and the material presented here synthesizes many of these thoughts; in particular it either duplicates or draws heavily on material described in previous papers (Stone, 2009; Carson and Stone, 2013; Stone et al., 2013), and is reproduced with permission.

Whilst the primary purpose of the assessment is to make a diagnosis and develop a treatment plan, we believe the assessment also plays a key role in treatment in its own right as it sets a tone and context for future clinical interactions (Stone, 2014). In this regard we aim to set up an atmosphere of collaboration based on taking the patient's problems seriously, and emphasizing that all facets of

the patient's presentation – physical, psychological, and social – are of importance. Patients with functional disorders can be perceived as difficult to help (Carson et al., 2004), and yet with the correct approaches we believe the consultation can be much more satisfying for both patient and doctor, leading more productively to explanation (see Chapter 44) and further treatment.

PREPARATION AND START OF THE CONSULTATION

Setting

The initial contacts with healthcare services should take place in a medical setting where adequate examination facilities are available. Obviously this is the norm where the first contact is with a neurologist, but it is not uncommon in psychiatric facilities for there to be a lack of basic equipment such as examination couches and the standard tools of physical examination. Such facilities are seldom

*Correspondence to: Alan Carson, Robert Fergusson Unit, Royal Edinburgh Hospital, Tipperlin Road, Edinburgh EH10 5HF, UK.
E-mail: a.carson@ed.ac.uk

available in psychotherapeutic settings. Even in situations where the primary purpose of the examination is to assess potential psychopathology, we believe that the willingness of clinicians to engage with physical examination is not just for diagnostic information but sends a clear signal that the physical element of the presentation is being taken seriously. It is sometimes suggested that patients might be referred directly from primary care to specialist psychotherapy. We think this approach is mistaken and that proper medical assessment is essential, partly because primary care diagnoses of functional neurologic disorder are often erroneous (Carson et al., 2000), and also because we think that the physical assessment and diagnosis are the key first steps in multidisciplinary treatment (Healthcare Improvement Scotland, 2012).

One core component of the examination is to allow enough time. Such consultations are always more satisfactory when there is enough time to deal with the patient's problems properly without either patient or doctor feeling rushed. Whilst consultations have to fit into the time available we generally find that 25 minutes or less is a false economy and an hour is preferable. In the UK 30 minutes is a squeeze but most of the key components can be addressed. Indeed, for complex patients longer may be advisable. It is far better to have time to attend to all aspects of the consultation and draw a meaningful conclusion than to have multiple return visits.

Preparation

Where possible review the patient's past medical history from medical records. Patients tend to have inaccurate recall of their own medical records and the information provided by them during history taking can be misleading, possibly especially so in patients with functional disorders (Schrag et al., 2004a,b). A previous diagnosis of a functional disorder might have been made, but the patient may not mention it. A thick pile of records has been considered a possible sign indicating a functional disorder since these patients may well have multiple consultations and a large number of tests. A major predictor of a likely functional disorder is a previous history of functional disorder (Hotopf et al., 2000); this may be present but be given alternate, disease-based labels, by patients in their own report.

In our experience functional motor disorders, such as paralysis, are unusual first presentations of functional disorders. Although this does happen, we would look for evidence of prior problems with functional symptoms that are more on a spectrum with normal experience, such as irritable-bowel syndrome, heavy painful menstrual bleeding, a history of unexplained abdominal pain, or a history of chronic back pain. It should be noted that these disorders are common in the general population

Table 15.1

Functional syndromes presenting to different medical specialties

Medical specialty	Functional symptom
Rheumatology	Fibromyalgia
Orthopedics	Chronic back pain
Neurology	Functional movement disorder
	Dissociative (nonepileptic) seizure
Ear, nose, and throat	Atypical facial pain
	Chronic unexplained dizziness
	Functional dysphonia
	Globus pharyngis
Infectious diseases	Chronic fatigue syndrome
Cardiology	Noncardiac chest pain
	Palpitations with normal investigations
Gastroenterology	Functional dyspepsia
	Irritable-bowel syndrome

anyway and their presence should be viewed only as indicative of an increased risk (Table 15.1).

By contrast, a prior history of psychiatric disorder is often unhelpful or misleading. Whilst a previous history of anxiety disorders or depression increases the risk of having functional disorders (Katon et al., 2001), such disorders are also common in neurologic disease and indeed can increase the risk of many neurologic diseases. In an approximate summary an emotional disorder will be present in two out of three functional cases and one out of three neurologic cases, so as diagnostic markers they should be treated with caution. Interestingly, in our experience the presence of psychotic illnesses such as bipolar illness or schizophrenia is seldom associated with functional disorder. We are unaware of any high-quality epidemiology to support this assertion, but the lack of comorbidity has been notable in some case series (Kranick et al., 2011) and in clinic. We would certainly recommend caution around making the diagnosis of functional disorder in a patient with a psychotic illness; and it should be noted that history taking in this group of patients can on occasions be difficult and the range of tardive movement disorders secondary to antipsychotics is wide and can be bizarre (Owens et al., 1982).

The referral letter itself can provide clues and patients with functional disorder are more likely to have multiple symptoms and sometimes a less clearly identified primary complaint.

There is also an association between previous operative procedures in particular, appendectomy (with normal appendix), hysterectomy and surgical sterilization, and functional disorders for reasons which are poorly understood but have been replicated in a number of studies.

Some clinicians like to use preclinic assessment questionnaires. These can be helpful but the risk is that they

end up as a lesser substitute for the process of listening and recording the patient's difficulties in person. In particular, symptom count measures, despite their recent hype, have little predictive validity (Carson et al., 2014; see Chapter 5 in this volume).

Beginning a consultation

It is often helpful to begin the consultation by simply allowing patients to speak freely about the problem that has brought them to clinic without interrupting. The mean duration of spontaneous talking time without interruption was 92 seconds in a primary care study. By contrast, most doctors interrupt within 20 seconds, following which patients can be inhibited in introducing new issues (Gask and Usherwood, 2002; Langewitz et al., 2002). Patients, however, frequently do not begin with the symptom that is most important to them and therefore this brief period of free-flowing dialogue can often allow the doctor to get a far better idea of what the key issue is more quickly than otherwise might be the case.

Everyone has their own style of assessment but it is noticeable how often patients with functional disorders want to “start from the beginning” of the story. To ensure the consultation and treatment focus on current issues, disability, and obstacles to recovery, it is often helpful to indicate to the patient that you wish to focus initially on how things are now and that you will come back to the story of how it happened later.

THE ASSESSMENT OF PHYSICAL SYMPTOMS IN THE HISTORY

Make a list of physical symptoms

After this initial opening we think it is most helpful to get a list of all the symptoms currently being suffered. During this phase the patient can be discouraged from going into minute detail and this can be further signposted by leaving space on clinic notes which will obviously be annotated later. Again, we find this tends to save time, and by getting all the symptoms out into the open, one often realizes that a number of symptoms are actually more or less facets of the same issue but described in different ways. In this context it is important to ask about pain, memory, fatigue, dizziness, and sleep disturbance. It is also worth enquiring briefly about other bodily systems and generally encouraging disclosure: “Is there anything else? I want to make sure I know everything bothering you.”

The sense that everything has been asked about and the assessment is complete will often do more than any other strategy to secure a good collaborative nature to the consult. When a core complaint is widespread bodily pain, the use of pain maps is often particularly helpful in allowing the patient to describe the symptomatology

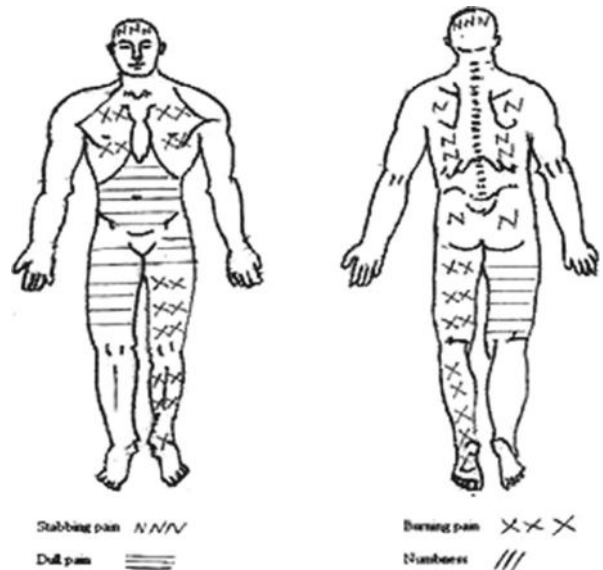


Fig. 15.1. The use of pain maps can help clearly transmit a lot of information about widespread pain quickly.

(Fig. 15.1). On completion of the list, do also ask: “What bothers you the most?”

TYPICAL DAY AND DISABILITY

It is often more informative to ask about what the patient can (rather than can't) do. Patients with functional symptoms have a tendency to report what they can no longer do rather than what they can. Whilst it is helpful to hear about previous function, ask them what they are able to do – do they enjoy it?

Taking a patient through a typical day provides information about levels of activity and social contacts and can give important supplementary information about mood and cognition. For example, if they enjoy a regular drama on TV then their mood may not be too bad and their cognition sufficient to follow the program.

ONSET AND COURSE

The onset in patients with weakness and movement disorders is sudden in around 50% of cases. It is helpful to explore whether there was a trigger. Studies have found that patients with functional neurologic symptoms often report a physical injury, or some pathophysiologic disease or physiologic event at the time of onset. This event may well have a role in shaping the future functional symptoms. Thus, a painful injury to a leg may lead to functional paralysis, shaking from rigors may lead to functional tremor, traveler's diarrhea to irritable-bowel syndrome, or a simple faint to future dissociative seizures (Table 15.2 and Fig. 15.2).

Table 15.2

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

In this model the initially noxious somatic experience, whilst quite benign, is modified by a range of cognitive processes (see Chapter 10 and below) to create the functional symptom but it is that physical experience that dictates the timing of onset and possibly shapes the nature of symptom.

Such physical triggers may also include symptoms experienced as part of psychiatric or emotional states, in particular panic. Dissociation (see below) is also commonly experienced at onset. More gradual-onset symptoms are often associated with fatigue.

By contrast, the typical description of psychologically traumatic life event is less frequently reported (Stone et al., 2009a), although Nicholson et al. (2016) found that, with very detailed examination via the Life Events and Difficulties Scale interview schedule taken over several hours, such events occur more commonly than they are typically reported in clinic. Whatever the correct answer to this controversial topic, we believe that detailed exploration of the question of recent life events can usually be left to one side at first contact unless the patient is obviously keen to explore it.

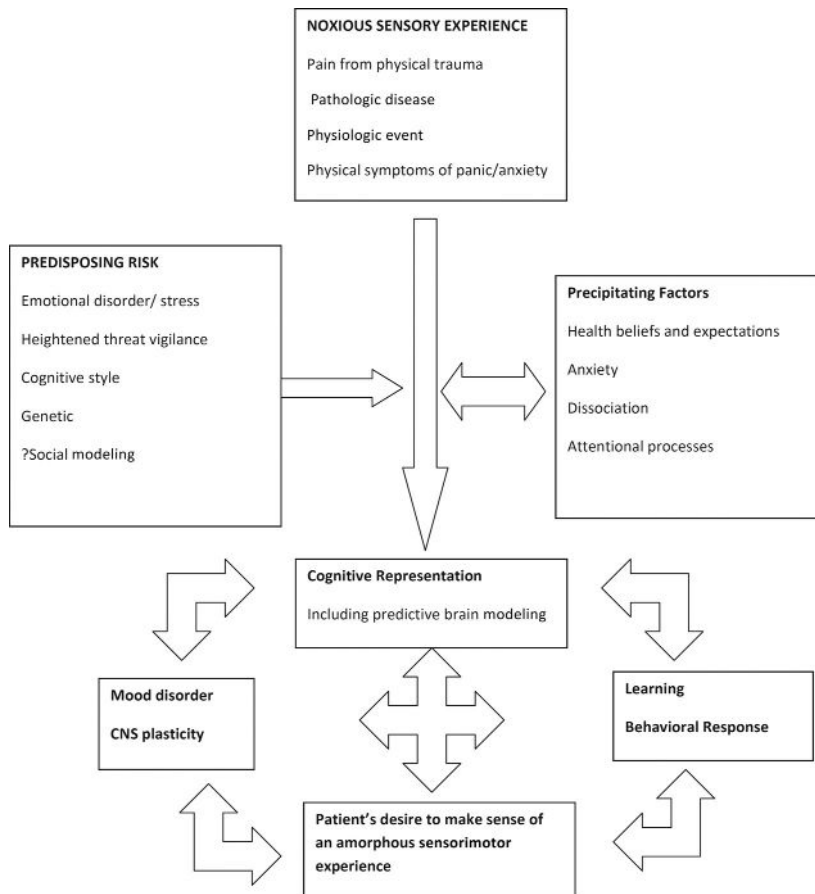


Fig. 15.2. Physical triggers to functional symptoms. CNS, central nervous system.

Trying to make sense of a long and complex history in a short consultation can be a challenge. The question, “When were you last really well, without any of these problems?” can be helpful, as can assessing the impact on work.

Mapping out the severity of the symptoms crudely on a graph of time vs. severity can help to understand the course and the relationship of symptoms to other medical interventions, illnesses, accidents, or life events.

Dissociative symptoms

We always ask specifically about symptoms of dissociation at time of onset. Dissociation is “a somewhat ambiguous collective term describing a range of psychopathological processes altering a person’s level of awareness and/or the integration of sensorimotor function, emotions, thoughts, memories and identity which may be subjectively perceived as a sense of disconnection” (Carson et al., 2012). It can be conceptualized in a range of different ways (Holmes et al., 2005), but at this stage we ask in particular about symptoms of depersonalization and derealization (Stone, 2006; see Chapter 8).

Depersonalization, a sense of disconnection from the body, is commonly described as *I felt strange/weird, I felt as if I was floating away, I felt disembodied/disconnected/detached/far away from myself, apart from everything, in a place of my own/alone, like I was there but not there, I could see and hear everything but couldn’t respond, like I was there but not there, I could see and hear everything but couldn’t respond.* Or less commonly, as “puppet-like,” “robot-like,” “acting a part,” *I couldn’t feel any pain, like I was made of cardboard, I felt like I was just a head stuck on a body, like a spectator looking at myself on TV, an out-of-body experience, my hands or feet felt smaller/bigger, when I touched things it didn’t feel like me touching them.* Sometimes with autoscopic experiences it can be that the patient’s perceived actual movements and the false perception of movement are engaged in different tasks.

Derealization, a sense of disconnection from the environment, may be described as *My surroundings seemed unreal/far away, I felt spaced out, it was like looking at the world through a veil or glass, I felt cut off or distant from the immediate surroundings, objects appeared diminished in size/flat/dream-like/cartoon-like/artificial/unsolid.*

Patients may also describe other dissociative symptoms involving memory: *I drove the car home/got dressed/had dinner but can’t remember anything about it, I don’t know who I am or how I got here* (fugue state), *I remember things but it doesn’t feel like it was me that was there.* They may describe their identity: *I feel like I’m two separate people/someone else* or distortions in

time: *I felt like time was passing incredibly slowly/quickly,* or personal boundaries: *I get so absorbed in fantasy/a TV program that it seems real,* or a loss of “sense of being,” *I felt an emptiness in my head as if I was not having any thoughts at all.*

We generally ask directly about a few such symptoms as we find patients are seldom willing to freely volunteer them as they seem so odd and they do not want to be considered as “going mad.” After a patient discloses such symptoms it is helpful to offer a few words of reassurance that, despite the odd nature of the experience, such symptoms are commonplace, do not indicate “madness,” are not sinister and in fact may help us to understand the nature of the complaints.

EXPLORING THE PATIENTS BELIEFS

It is vital to understand the patient’s perspective on the cause of the symptoms. It is known that the patient’s beliefs have a significant effect on outcome (Sharpe et al., 2010) and erroneous beliefs, and the sensory and motor distortions they produce in the nervous system, are increasingly believed to play a significant part in the etiology and mechanism of functional neurologic disorders (Edwards et al., 2012).

The assessment of a patient’s views on the illness is best conducted in line with Leventhal’s common-sense model of illness (Cameron and Leventhal, 2003). This is described in more detail in Chapter 10, but is outlined in brief in Table 15.3. The key in assessment is not just to ask about the five elements of illness belief but to be alert to areas of distortions of illness beliefs where the patient has followed a line of reasoning that at one level makes sense but that is ultimately maladaptive. A basic example of this might be a patient who hurts his back digging the garden. Rest helps the pain and when he returns to mobilization it is sore again, so he concludes that he must rest for longer rather than mobilize gradually until the pain eases. Although in the short term this might be reasonable behavior, if it continues day after day layers of avoidance and related anticipatory anxiety will lead to an escalation in pain and disability (Fig. 15.3).

One tip in the exploration of illness beliefs is to remember the effect of conditioning, in particular single-event aversive conditioning, in creating a link between two factors that should otherwise be physiologically unrelated, e.g., *whenever I drink tea I get severe paresthesia in my feet.* This type of linkage should be explored whenever patients report a clear association in their mind between two pathophysiologically implausible factors.

Some people require quite a lot of encouragement to admit their thoughts about what’s wrong and may need encouragement to voice disagreement with previous medical opinion. It is possible and often helpful to allow

Table 15.3

The common-sense model of illness regulation

Element	Cognition	Distortions	Example in functional disorders
Identity	What are these symptoms?	Symptoms cause labels But labels also lead to the self-generation of symptoms	<i>I have limb weakness; I think it is a stroke</i>
Cause	What caused these symptoms?	Beliefs that symptoms are due to damage and therefore irreversible	<i>A stroke is a clot in the brain; I think it happened because I was overdoing it</i>
Consequences	What effects will the symptoms have on my life?	Cog representations guide subsequent behavior	<i>I am scared I could end up in a wheelchair or even die</i>
Time line	How long will the symptoms last?	Behaving and adjusting life in the belief something will go on for ever, i.e., quitting job, can become a self-fulfilling prophecy	<i>I don't think my leg will ever get better. What if I get more disabled?</i>
Cure and control	What will help make the symptoms better?	Change in symptoms provides feedback on coping strategies and may result in reappraisal of symptoms or adoption of maladaptive strategies, i.e., pain on activity leading to increased downtime	<i>I think I need to rest more to make sure this doesn't happen again. It is important I don't do anything to provoke it or cause it to recur</i>

From Stone et al. (2009b), by courtesy of the Guarantors of Brain.

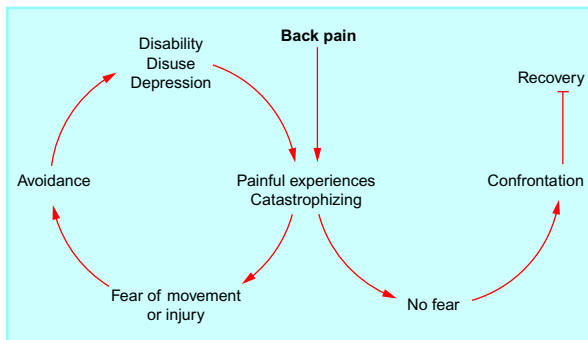


Fig. 15.3. The role of illness beliefs in the development of chronic back pain in the absence of structural disease. (From Main and Williams, 2002 with permission from BMJ Press.)

patients to vent feelings about prior diagnostic opinions without having to take sides. Some patients will “parrot” an explanation they have been given without necessarily believing it. Asking the patient what level of confidence they have in that diagnosis on a scale of one to 10 can be revealing:

“Doctor: What do you think is the cause of your weak leg?”

Patient: They said it was some kind of dysfunction of the brain. They said it was like software, not hardware.

Doctor: Yes, but do you think that's correct?

Patient: Well, you're the doctor.

Doctor: I am, but it's important for me to know what you really think, or even if you just have a hunch about something, so that I can try to help. How confident are

you that the diagnosis of functional disorder is correct? 20% confident? 80% confident?

Patient: About 50%.

Doctor: Are there any other conditions you were still wondering about or had niggling doubt about?

Patient: Well, I was wondering whether they could have missed multiple sclerosis.”

Using some of the basic techniques of cognitive-behavioral therapy assessment can also be useful here. Asking patients not just what they think but what the personal significance of those thoughts are can be helpful. One can then ask them what they actually do when they have these thoughts or concerns and whether viewed objectively they make sense:

“Patient: I am really frightened of the attacks in case they damage me [dissociative seizures].

Doctor: Are you worried about any specific way they might damage you?

Patient: Yes, when I have had the attacks and have fallen and hurt myself, I've had quite a few injuries and had to go to casualty.

Doctor: Do you do anything to try to prevent that happening?

Patient: Yes, whenever I get any sense an attack is coming I run to the bathroom and lock myself in and sit on the toilet.

Doctor: If you fall in the bathroom is there much you can hit? (pause) How will people help if the door is locked?

Patient: Oh, it just sort of felt safe.”

Finally, this part of the consultation is a useful point to establish what the patient was hoping that health professionals in general ought to do to help, and what, in particular s/he was hoping for from the assessment on the day.

Time spent on illness beliefs pays dividends later when helping the patient to understand the diagnosis. For example, if you haven't established that the patient is actually really concerned about a throwaway remark a junior doctor made about a scan showing some "high signal lesions," then progress with treatment may be slow. Alternatively, if you realize that the patient is only in the clinic because she had a new young general practitioner, she had accepted her disability years ago, and she wasn't really looking to engage in rehabilitation, then you could be talking at cross-purposes.

THE PSYCHOLOGIC ASSESSMENT OF PATIENTS WITH FUNCTIONAL DISORDERS

The basic assessment of depression, anxiety, and other psychologic disorders such as posttraumatic disorder or obsessive compulsive disorder can take place during a neurologic assessment and should be part of a psychiatric assessment. It is however not always essential to spend a lot of time on this. If you have discussed a typical day, fatigue, sleep, and concentration you will already have most of the relevant information. In addition, paying attention to some of the basic features of a mental state examination, such as eye contact, reactivity of mood, or agitation, can be revealing.

Asking about depression, anxiety, and other common psychologic symptoms

At the first assessment, overly blunt questioning in the domain of depression and anxiety can be counterproductive to a therapeutic relationship. The question of how detailed to make this should be led by cues from the patient, who is usually expecting an assessment of his weak leg or blackouts, not his mental state.

The typical patient with depression feels down, tearful, and lethargic. This is accompanied by a cognitive triad of distorted thoughts with a sense of hopelessness and futility about the future, a sense of worthlessness about the present, and a sense of guilt about the past. The symptom of anhedonia, the inability to experience pleasure, is central. There is usually a range of somatic symptoms, including disturbed sleep with early-morning wakening and lack of refreshment, loss of appetite, poor concentration, loss of libido, and a sense of general malaise.

Rarely, patients may be frankly suicidal and this represents a medical emergency to be dealt with immediately.

In many patients with functional symptoms detection is less straightforward. Patients may emphasize the somatic element of the presentation and view mood symptoms as a rational response to intolerable physical symptoms rather than an illness in their own right. The presence of low mood may be denied in response to direct questions, partly because the patient is aware that the doctor is "angling" for a psychiatric diagnosis. Exploring mood in this situation requires considerable tact. When suspicion is raised due to the presence of typical somatic symptoms (Fig. 15.3), sympathetic, leading questions can be more fruitful:

"It must be difficult living with all that pain ... Have you cut down on your range of activities? ... Do you find you stopped enjoying things that you can still manage to do physically? What about watching your favorite program on TV? Do you still enjoy it?"

When assessing inpatients a critical question is often: *When friends or relatives come to visit do you look forward to their company as a break from the monotony? or do you just want to hide away and wish they would go?*

In patients who report mood symptoms a further diagnostic challenge is to separate out those with new symptoms from those who have dysthymic personalities by asking, *When did this first start? Have you always been like this since you were a teenager? Is this a change from your normal self?*

The core of an anxiety disorder is disproportionate, persistent, and unwelcome worry. Anxiety disorders present with a range of somatic symptoms such as muscle tension/pain, fatigue, tingling, nausea, and poor concentration, and symptoms associated with excessive, shallow, or disordered breathing. Abdominal bloating and borborygmi, from aerophagy are common. Peripheral paresthesiae affecting fingertips, toes, and perioral regions is common but tetany is rare. Patients will often report sensory symptoms as unilateral, but on questioning will usually disclose very mild symptoms on the opposite side. Patients often complain of fluid sensations under their scalp or tightly localized, transient headaches which they "can put a finger on." Commonly, anxiety tends to exacerbate existing primary headache disorders such as migraine.

Where anxiety disorders are suspected the key distinction is to separate generalized anxiety, which presents with ruminative worry about a wide range of topics with no consistency or theme, from phobic anxiety, in which anxiety presents in response to a given stimulus.

In patients with functional disorders a phobic component of anxiety may be obscured by misattribution to physical disease. This can follow an agoraphobic pattern.

For example, “attacks” attributed to effort occur on leaving the house: *My heart beats like crazy, my legs turn to jelly, I feel I am going to collapse, I just have to sit down, I can only manage to walk 200 yards before it happens.* Alternatively, the fear may be of a symptom – “bringing on pain” or “falling” both being common. This leads to cycles of decreased activity which can in turn lead to physiologic complications through disuse (see [Chapter 10](#)).

As with depression, be careful asking questions about anxiety and panic in patients with functional symptoms – there is a risk they will see you as criticizing them personally or labeling them a hypochondriac. Useful questions include:

“Do you often find yourself feeling worried about your symptoms? Do you often feel on edge or tense about things? Do you ever feel like you can’t keep a lid on that worry? Do you ever get lots of physical symptoms all at once? Is it frightening when that happens?”

Occasionally the unexpected *You’re getting all these severe leg pains, you’ve been off work for 6 weeks and yet you are not worried – I would be!* pays dividends.

Many psychologic symptoms require specific questioning to elicit, sometimes because patients are embarrassed by them, sometimes because patients just don’t realize they may be relevant or even pathologic, and sometimes because patients fear that a discussion of their psychologic state will detract from a proper physical assessment. The core of a posttraumatic stress state includes intrusive symptoms (such as nightmares and episodes where patients ‘relive’ traumatic experiences – so-called flashbacks), avoidance (e.g., avoiding driving after a car accident), negative feelings (e.g., feeling empty or having difficulty thinking about the future), and alterations in arousal (e.g., being hypervigilant or excessively “jumpy”).

Obsessive compulsive symptoms are also not uncommon in patients with functional disorders but underreported by embarrassed patients. Obsessions are repetitive and intrusive thoughts or images that cause distress which the person tries to suppress. Compulsions are repetitive and excessive behaviors sometimes performed in response to obsessional thoughts which the person recognizes are excessive and which cause distress, take more than an hour a day or interfere with normal functioning, and tend to be underreported by patients who may be embarrassed by them. Full-blown obsessive compulsive disorder does not appear to be particularly common in patients with functional symptoms but by contrast obsessive traits do seem to be common. One can often spot circumlocution whilst taking the history, with a need to fill in every back clause in detail, including lots of extraneous material, without ever really coming to the point.

“Doctor: If I was looking at your cupboards at home, would I find everything was kept neatly in order, each thing with its own place?”

Patient: Yes, my wife is always teasing me about it.

Doctor: What if I moved something – could you just leave it or would you be unable to relax until it was back in the proper place?”

It is conceptually helpful to think of pain communication behavior ([Waddell et al., 1984](#)) as part of the mental state examination. Pain itself is conceptually difficult ([Perl, 2013](#)), but one aspect of the consideration of pain, pain communication behavior, is best considered within the context of consideration of emotions. It is the interactive process between the patient and the clinician that surrounds the communication of pain. In essence the more the pain is communicated via sighing, grimacing, groaning, inappropriate response during examination, and so on, the higher the likelihood of a significant psychogenic component. This is separate from the simple hyperreactivity of pain response that can occur in a pure allodynia which should not be associated with the communicative element of pain but simply a reported response to soft or blunt touch on examination.

Family history, childhood, and recent stress

Functional symptoms are multifactorial in etiology. Genes may play a part (see [Chapter 14](#)), so remember to consider a family history from that perspective. Of course, one must be careful in ascribing familial clustering to a genetic cause and in some reports this has been explained by intrafamilial suggestibility and mimicry ([Stamelou et al., 2013](#)).

Childhood adverse experiences predispose to functional disorders in adult life. It is however important to remember that, whilst such aversive experiences increase the risk of a functional symptom, a significant proportion of patients, 30–60%, will have had no such experiences, with events such as sexual abuse being rarer still. Additionally, such experiences are unfortunately far from rare in the general population so, whilst they may be one of many relevant etiologic factors, when present they are not diagnostically helpful. Enquiry should be tactful and may be best left for a subsequent occasion if there are signs that it is a “difficult” first encounter. Even treatment of functional symptoms does not need patients to disclose every traumatic and abusive experience – indeed, in many circumstances that may be actively unhelpful.

What a psychiatrist, and sometimes the neurologist, may wish to gain is some general overview of childhood. If the patient discloses, or hints strongly at, significant physical or sexually abusive experiences it is often more helpful to let the patient set the pace of any disclosure rather than to push the issue: *Is that something you would*

be able to tell me a bit more about or is it something you would prefer to pass over for now?

More commonly, however, the aversive experiences are milder; questions such as the following will help build a picture.

“Did you feel secure and cared for as a child? Did you feel a burden to your parents? Did you get bullied at school? What was the atmosphere like at home? Did your parents argue a lot? Did they ever hit each other? Did either of your parents drink too much?”

Recent life events and stressors may also be important in some patients but, again, it’s important to avoid assumptions. Studies of patients presenting to primary care with functional disorder show that they may volunteer explanations based around stress for their physical symptoms but doctors close down such enquiries too early in a rush to exclude biomedical causes of disease (Ring et al., 2005).

“Patient: The pain is just kind of all over.

Doctor: And when does it come on?

Patient: It started shortly after my divorce.

Doctor: And are you OK generally, weight steady, no night sweats?”

In patients with functional neurologic disorders presenting to secondary care, however, there is some evidence that they are often less forthcoming. For example, patients with functional disorders are often less likely to attribute their symptoms to stress than patients with disease (Crimlisk et al., 1998; Stone et al., 2010) and will flatly deny any problems in their life even though you sense that they may be distressed by their personal circumstances. This can be difficult to deal with; challenging them usually just makes the patient defensive. Patience is usually the key, so keep a mental note that it is a subject to return to. Conversely, there are patients in whom stress and life events are really not a factor in the development of their symptoms. A recent study that evaluated a series of patients with functional movement disorders with diurnal cortisol levels did not find any difference from controls (Maurer et al., 2015). The take-home message is that it is important not to insist that the patient must be stressed.

Dealing with anger and excessive praise

Dealing with anger can be a problem when assessing patients with functional problems, but if the steps outlined in this chapter are followed, we hope it will be an unusual occurrence. However, no matter how well a consultation is conducted one will be faced with an angry patient on occasions. The first rule in dealing with this is not to get angry yourself. The patient’s anger is often a sense of frustration secondary to a feeling that he or

she is not being understood. It is important to remember that this may not be the result of your consultation but a legacy of previous contacts or other factors within the patient. Depending on the level of anger shown, acknowledging this either indirectly or, if required, directly is usually a helpful starting point.

“It can be very frustrating living with these symptoms every day – do you feel that we as doctors really understand how difficult it is?”

I am sorry, I am obviously frustrating you, I didn’t mean to. What is it that you really want me to understand most?”

The problems however may not be secondary to the patient’s symptoms but secondary to some other problem. It may not be something you are able to help with but simply feeling you have understood the difficulty can often diffuse matters for the patient.

Try to understand what the patient thinks you are doing or thinking. This can often be done by summarizing the symptoms so far, and emphasizing the associated disability as you go – some of the technical information we wish to gather as clinicians to make a diagnosis can seem rather irrelevant to the patient – and asking, *Am I getting it right so far?*

Patients’ anger can often relate to erroneous beliefs or fears. This may be about things you have never thought of or considered; try to understand what they are.

A proportion of patients who have had functional symptoms have suffered highly aversive and abusive experiences throughout their lives and have as a result distorted styles of attachment. Attachment style describes the way in which someone habitually approaches interpersonal relationships. Try to remember that experience has taught the patient that even if a person is nice today s/he may well horrendously assault the patient tomorrow. Trust is a commodity in scant supply – don’t take it personally. Equally, remember the same processes can lead to overly idolizing attachments after minimal contact. It may be that you are *the best doctor ever* and *the only one who cares and understands*, but whilst such comments are always pleasing for the ego, they may relate more to an abnormal attachment style on the patient’s part rather than your own brilliance. That doesn’t mean one cannot politely accept a compliment but equally be careful if the consultation becomes overly familiar of if you are invited into conspiratorial conversations about the skills of a colleague, for example.

Sometimes patients become angry because they detect hostility, boredom, or anger in the clinician. There are several reasons why clinicians find patients with functional disorders difficult to help. These include the complex mixture of multiple problems, lack of training, lack of time, concerns about exaggeration (see below)

and negative attitudes that clinicians often bring to the consultation (see [Chapter 44](#)). There are probably others, though, which are more on the patient's side of the equation. These include the presentation of distress and apparent request to help correct not just a neurologic symptom, but a whole set of symptoms and life circumstances. In addition some patients, most definitely the minority, appear to have a reduced sense of awareness of the time constraints of the clinic, reduced ability to take turns during conversations, and such a compulsive need to describe their problems at great length that they unwittingly reduce the time available to receive the help they crave. The clinician making the assessment should not be critical of the patient in this situation. The reasons for this behavior may relate again to attachment styles discussed earlier. The clinician can however strive to help direct the consultation so that the patient does feel listened to but still keep time to explain the diagnosis and move forward with treatment. And, as a final rule, don't get angry!

Exaggeration

During assessment, especially during the examination, the clinician may become aware of behavior that appears exaggerated. It is worth considering the various explanations for this, only one of which is that the patient is deliberately exaggerating.

An example of verbal exaggeration may be that the patient may report pain as 11 out of 10 in severity, even when you suggest that 10 out of 10 would be the worst pain imaginable, but is able to converse normally during the assessment. Putting a numeric value on an abstract sensation like pain is hard for anyone, but especially when measured against someone else's experience. The phrase "11 out of 10" should usually just be interpreted as meaning "it's really bad," although, paradoxically, these apparent verbal exaggerations often lead to clinicians devaluing the patient's complaints.

For some patients the more dismissive the clinician appears, the more likely they are to have pain communication behavior in a misguided, and usually nonwillful response to convince the clinician that there really is a problem. Again, paradoxically, this may make the clinician even more dismissive and the outcome is poor for both parties. One helpful question to ask yourself when you see something that any layperson would regard as exaggeration is 'is it "exaggeration to convince" or "exaggeration to deceive.?"' Something similar has been described in functional gait disorder as the "huffing and puffing sign," where a gait is associated with signs of effort ([Laub et al., 2015](#)). A blinded study of video material concluded that 44% of 131 patients had at least mild signs of this whereas none of the 37 neurologic controls did.

The diagnosis of functional motor disorders is usually made on the basis of internal inconsistency. Most of the diagnostic maneuvers, for example, Hoover's sign or tremor entrainment, rely on the principle that the more the patient thinks about the movement, the worse it gets. Therefore, if a patient's gait is much worse during formal examination than it was when he walked in to the room, this is really just in keeping with a functional disorder relating to an abnormal attentional state and is not clearcut evidence of willful exaggeration, however much it may look like it. Clinicians should not be naïve either, but exaggeration can only be recorded with more confidence if there is a marked discrepancy between recorded and observed function. Even this can be problematic. A study of actigraphy in functional tremor showed that even patients who know they are being monitored are hopeless at guessing how bad their symptoms are. In this study, the 10 patients with functional tremor thought, on average, that their symptoms were present 83% of the time when in fact they were only present 4% of the time (compared to 58% reported vs. 24% observed in organic tremor) ([Pareés et al., 2012](#)). Factitious disorders and malingering are discussed in more depth in [Chapter 42](#).

Mental state examination

The mental state examination in patients with functional symptoms is often relatively uninformative compared to gaining an understanding of the patient's illness beliefs and behaviors. Only in a minority of patients does the traditional picture of anxiety or depression presenting with physical symptoms apply. A significant proportion of patients will have relatively normal mental states on examination. Perhaps the single most important feature to be aware of is that this is perfectly compatible with a diagnosis of a functional disorder.

The most commonly encountered abnormal mental state is of a largely anxiety-driven hyperarousal accompanied by a slightly obsessive speech structure, anxiously driven, unfocused, and so full of subclauses that it is difficult to control the interview and "separate the wood from the trees." The patient is often in an egocentric state and relatively oblivious to normal social cues from the doctor ([Stone and Carson, 2015a](#)). There is a hypervigilance, often directed to perceived verbal slights surrounding the reality of the symptoms. This is often accompanied by an attentional bias towards the affected body area that shows itself by repeated checking and monitoring behavior, as well as eye gaze deviation. This can often change quite dramatically into a friendly and appreciative state if the consultation has gone well and, on occasion, anger if it has not.

The true anhedonic state of significant depressive illness, which is of emptiness rather than emotional upset, is, by contrast, relatively rare but is occasionally encountered in cases of depression presenting predominantly with physical symptoms. Here there is a monotonous, monosyllabic speech with little in the way of elaboration of answers. Eyes are cast downward, and the whole interview feels slow and lugubrious.

Pure “somatized” anxiety as opposed to anxiety comorbid to a functional disorder usually shows itself as a general health anxiety or a nonlocalized physical symptom but also tends to be accompanied by a lack of selective attentional bias to a specific body area.

Pseudohallucinations in which the patient recognizes the false sensory experience comes from her own mind are occasionally encountered along with occasional paranoid phenomena (the seeing of clear illusory image when gazing at an ill-defined stimulus which intensifies with attention, e.g., seeing a face in a cloud), especially in patients with borderline/emotionally unstable personality types, but true hallucinosis with associated searching behavior, such as seeking the source of the voice or vision, are very rare indeed and should be a red flag for misdiagnosis.

Patients with functional symptoms often show high levels of selective attention, albeit towards their own bodies. A display of poor selective attention such as being distracted by every extraneous noise should suggest some alternate diagnosis.

Patients will often describe disruption of concentration and memory. The features of this are described in detail in [Chapter 35](#) on functional cognitive disorders.

La belle indifférence

La belle indifférence (smiling indifference to disability) has appeared as a key diagnostic feature of conversion disorder for over a century and originated in the works of Freud and Janet. It epitomizes the “hydraulic” theory of conversion in which intrapsychic distress from a conflict is converted into a physical symptom, thus reducing distress, so-called primary gain. It is a difficult clinical sign to quantify and therefore study. However, data from 11 studies found that *la belle indifférence* occurs in a similar frequency in patients with functional disorders as those with neurologic disease (21% vs. 29%) ([Fig. 15.4](#)). There is also a differential diagnosis of indifference which the clinician should consider. The patient may just happen to have a stoic attitude to disability, whether caused by disease or not. Others are good at putting a “brave face” on for a clinician. Sometimes, perhaps especially in patients with functional disorders, this tendency is amplified by an awareness that the clinician is angling to find a psychiatric disorder. This perhaps is the commonest scenario in functional disorders in our experience. When the patient has a factitious disorder, this may be associated with indifference for obvious reasons. Neurologic diseases affecting frontal/executive function are particularly likely to lead to apathetic or indifferent states. One patient referred to us (JS) thought to have typical *la belle indifférence* turned out to have Wilson’s disease. Finally, there are some patients who even on further assessment may be said to have true “indifference,” but our own experience is that this is rare ([Stone et al., 2006](#)).

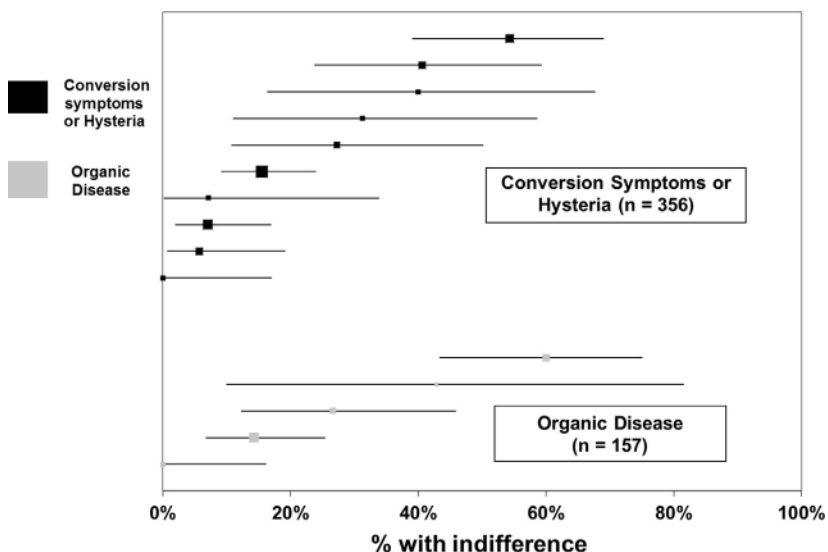


Fig. 15.4. Frequency of *la belle indifférence*. Each point represents an individual study, the size of the point represents the number of patients in the study, and the associated lines are 95% binomial confidence intervals. (From [Stone et al., 2006](#) with permission from the Royal College of Psychiatrists.)

PHYSICAL EXAMINATION

The use of the physical examination, so essential to making a diagnosis of functional neurologic disorder, is discussed in each individual chapter in this book. Some common diagnostic pitfalls in relation to dissociative (nonepileptic) seizures and functional motor disorders are discussed below. Articles summarizing these physical signs are also available (Stone, 2009; Stone and Carson, 2015b). One aspect we would highlight here though is that we recommend explaining the features of particular positive signs during or at the end of the examination rather than keeping them as secrets (see Chapter 44).

PRINCIPLES OF DIAGNOSIS AND DIAGNOSTIC PITFALLS

Reaching a diagnosis

The guiding principle of diagnosis of most functional symptoms is that there should be inconsistency during the physical examination (so-called internal inconsistency) or incongruity with recognized neurologic disease. That is, there should be positive signs. Like much of neurology, there are gray areas, and patients for whom there is considerable diagnostic uncertainty. Clinicians perhaps tend to have a feeling that the important thing is not to diagnose a functional disorder and have it later transpire that there was an underlying disease explanation. However, if the consultation has been conducted in a collaborative fashion we generally find that patients are accepting of this. By contrast, clinicians seem to seldom worry about mislabeling functional symptoms as a neurologic disease, although in our experience this can often be more damaging and can lead to very difficult consultations when one tries to correct it (Coebergh et al., 2014).

Eight shades of diagnostic change

Even when the diagnosis does appear to change over time, it is rarely as simple as *I thought it was functional, but actually it is multiple sclerosis*. There are different kinds of diagnostic change with different degrees of error. As well as the best-known type of misdiagnosis – when you look back and think, *I got that wrong* – there are other types of change that could be construed as error when in fact they aren't (Table 15.4).

For example, someone presenting with functional hemiparesis who later goes on to develop motor neurone disease, may genuinely have had a functional hemiparesis; it's just that you didn't detect (and weren't able to detect) the comorbid neurologic disease predisposing to it at the time. Alternatively, a patient presenting with a functional movement disorder may 1 year later have

a stroke, but it still doesn't account for the functional movement disorder. Diagnostic disagreements and patients where the diagnosis of functional symptoms is initially in the differential but then drops out also form part of the list of ways in which diagnoses may change over time without there necessarily being a "howler."

Diagnostic pitfalls – general considerations

Table 15.5 lists some factors that we often come across in patients who have been erroneously labeled as having a disease when they actually have a functional disorder, and vice versa.

"THIS PATIENT IS ANXIOUS/RECENTLY STRESSED/HAS A PERSONALITY DISORDER," SO MUST HAVE FUNCTIONAL SYMPTOMS

Probably the commonest source of diagnostic error is when the clinician pays too much attention to the patient's psychosocial history and not enough to the presenting symptom. A generation and more of doctors have been taught via psychiatric diagnostic criteria that functional neurologic symptoms are a form of conversion disorder and as such represent the conversion of recent stress into a physical symptom. As discussed earlier, recent and remote adverse experience as well as comorbid psychiatric disorder (such as anxiety, panic, and depression) and maladaptive personality traits (such as avoidant or borderline personality traits) are more common in patients with functional neurologic disorders in most studies. However, many patients with functional symptoms are psychiatrically normal and around a third of patients with defined neurologic disease have comorbid psychiatric symptoms.

The upshot of this is that it's dangerous to base your diagnosis on the psychosocial history, however tempting the narrative may appear. Just as you wouldn't make a diagnosis of stroke because someone smokes and has high blood pressure, these features should be regarded as supporting risk factors but not diagnostic in their own right. Be particularly careful of the patient who thinks the symptoms are stress-related, as patients with disease are more likely to present with psychosocial attributions than patients with functional symptoms (Stone et al., 2010).

"THE PATIENT IS TOO NORMAL/NICE/STOIC/MALE/YOUNG/OLD/INTELLIGENT/MUCH LIKE ME," SO MUST HAVE A NEUROLOGIC DISEASE

The converse and quite common pitfall we have observed is the patient with functional symptoms who has the misfortune to share the same social and demographic features of the doctor attempting to make the

Table 15.4

Change in diagnosis doesn't necessarily mean you got it wrong first time round

Type of diagnostic revision	Example	Degree of clinician error
Diagnostic error	Patient presented with symptoms that were plausibly all due to multiple sclerosis but was diagnosed with functional symptoms. The diagnosis of multiple sclerosis had not been considered and was unexpected at follow-up	Major
Differential diagnostic change	Patient presented with multiple symptoms. Doctor suggested chronic fatigue syndrome as most likely but considered multiple sclerosis as a possible diagnosis. Appropriate investigations and follow-up confirmed multiple sclerosis	None to minor
Diagnostic refinement	Doctor diagnosed epilepsy but at follow-up the diagnosis is refined to juvenile myoclonic epilepsy	Minor
Comorbid diagnostic change	Doctor correctly identified the presence of both epilepsy and nonepileptic seizures in the same patient. At follow-up, one of the disorders has remitted	None
Prodromal diagnostic change	Patient presented with an anxiety state. At 1-year follow-up the patient has developed Alzheimer's disease. With hindsight, anxiety was a prodromal symptom of dementia but the diagnosis could not have been made at the initial assessment as the dementia symptoms (or findings on examination or investigation) had not developed sufficiently	None
<i>De novo</i> development of organic disease	Patient is correctly diagnosed with chronic fatigue syndrome. During the period of follow-up, the patient develops subarachnoid hemorrhage as a completely new and unrelated condition	None
Disagreement between doctors, without new information at follow-up	Patient is diagnosed at baseline with chronic fatigue syndrome and at follow-up with chronic Lyme disease by a different doctor even though there is no new information. However, if the two doctors had both met the patient at follow-up, they would still have arrived at the different diagnoses. This would be reflected in similar divided opinion among their peers	None
Disagreement between doctors, with new information at follow-up	Patient is diagnosed at baseline with chronic fatigue syndrome and at follow-up with fatigue due to a Chiari malformation by a different doctor because of new information at follow-up (in this case a magnetic resonance imaging scan ordered at the time of the first appointment). However, the first doctor seeing the patient again at follow-up continues to diagnose chronic fatigue syndrome, believing the Chiari malformation to be an incidental finding. This would be reflected in divided opinion among their peers	None

Adapted from Stone et al. (2009b) with permission from the Guarantors of Brain.

diagnosis. In line with the discussion above, middle-aged males, people who are “normal,” “nice,” or “seem genuine enough” can all develop functional symptoms, even dramatic ones (Carson et al., 2011). Studies on older patients with dissociative (nonepileptic) attacks show that they have an equal gender ratio and often suffer from potentially life-threatening disease (such as ischemic heart disease or severe asthma), triggering health anxiety which links to the attacks themselves (Duncan et al., 2006).

“I’VE MADE A DIAGNOSIS; THERE IS NO NEED FOR ANOTHER ONE”: THE PROBLEM OF COMORBIDITY

The presence of any disease, however small, tends to “trump” the presence of functional symptoms. But the reality is that the experience of bodily dysfunction caused by neurologic disease is one of the most powerful risk factors for developing functional symptoms. Many patients have two diagnoses, for example: epilepsy and dissociative (nonepileptic) attacks; multiple sclerosis and functional limb weakness; idiopathic intracranial

Table 15.5

Functional disorders in neurology; general factors relevant to diagnostic error

Features of neurologic disease presentations that can lead erroneously to a diagnosis of a functional disorder	Diagnostic clues/how to avoid error
The presence of psychiatric disorder, especially personality disorder	Detecting psychiatric comorbidity may be useful in treating the patient but should be ignored in making the diagnosis. Focus on the nature of the attack / the physical examination. Are the physical features typical of functional symptoms?
Presence of schizophrenia or other psychotic illness The patient's presenting complaint is of new-onset mood or behavioral disturbance.	Such patients seldom have functional symptoms Patients with functional symptoms rarely complain of significant psychiatric or behavioral symptoms, e.g., panic, as their primary subjective complaint, even if it is clearly present
The presence of an obvious life event or stressor	Ignore the presence of recent stress in making the diagnosis even if this may be relevant for treatment
Failure to consider that the patient may have a functional disorder and a neurologic disease	Remember that neurologic disease is one of the most powerful risk factors for developing a functional disorder (e.g., epilepsy/dissociative (nonepileptic) attacks, MS/functional limb weakness)
Failure to consider that the patient may have functional disorder and a progressive neurologic disease which may be too early for you to diagnose (yet)	As above, but in some cases, especially where neuroimaging doesn't help, the disease may only become apparent on follow-up (e.g. motor neurone disease, Wilson's disease, Alzheimer's, myopathy)
<i>La belle indifférence</i> – apparent indifference to disability	This concept is wedded to conversion disorder and is of no diagnostic value, apparently occurring as frequently in neurologic disease, especially with frontal-lobe involvement (Stone et al., 2006)
Normal neuroimaging	Many neurologic diseases, e.g., epilepsy, amyotrophic lateral sclerosis, myopathy, spinocerebellar ataxia, have normal CNS imaging. Don't rely on it alone to exclude disease
<hr/> Features of patients with functional symptoms that can wrongly put you off the diagnosis	
The patient is normal/nice/stoic/like me	Normal people get functional disorders too
The patient has no "form," i.e., previous functional symptoms	Patients can present with dramatic neurologic functional symptoms with no prior history
The patient has not been stressed	Between 1 in 3 and 1 in 4 patients have no evidence of recent stress
The patient is not tired/only has one symptom	Lack of fatigue or other symptoms should make you think twice about a diagnosis of functional symptoms but monosymptomatic presentations do occur
The symptoms came on after injury, minor pathologic disease The patient suggests a psychologic causation	Commonplace in functional symptoms (Stone et al., 2012). Around 1 in 4 patients with functional symptoms do think that psychologic factors are relevant
The patient has an established diagnosis of "known epilepsy, "known MS"	Always question other people's diagnoses (and your own!)
The patient is too old	Older patients with functional symptoms often have health anxiety and comorbid disease (Duncan et al., 2006)
Incidental abnormalities on MRI (e.g. enlarged perivascular space, Chiari malformation, disc protrusion), EEG, serology, or other tests	Don't assume that all structural abnormalities are relevant

MS, multiple sclerosis; CNS, central nervous system; MRI, magnetic resonance imaging; EEG, electroencephalogram.

hypertension and functional visual symptoms. It is easy for the presence of disease to obscure the presence of functional symptoms. Conversely, recognizing the functional symptom diagnosis can assist in the patient's treatment as it will often have more potential for reversibility than the underlying disease. In our own Scottish study of 2467 outpatients with neurologic disease, around 12% also had a diagnosis of a functional symptom. In these 12% of patients, no one disease category was more common than another. In other words, patients with, for example, multiple sclerosis or Parkinson's disease do not appear to be more prone to functional symptoms than people with epilepsy or muscle disease (Stone et al., 2012).

Comorbidity can present concurrently; for example, a patient who presents acutely with a mild episode of demyelination in which the clinical features of the weakness are predominantly functional. Or functional symptoms can develop as a later complication of neurologic disease, with health anxiety often having a key etiologic role.

More problematically, some patients, especially those with degenerative and slowly progressive conditions, may present with functional symptoms years before the clear onset of their neurologic disease. In some cases, a definite functional diagnosis such as functional paralysis can present as part of the commonly encountered "psychiatric prodrome" in dementias. In others, it appears as if the experience of having a very mild ataxia, for example, in the very early stages of spinocerebellar ataxia, is enough to trigger the functional symptom. This has also been demonstrated for Parkinson's disease (Onofij et al., 2010).

So always look for comorbid disease, even if the diagnosis of functional symptoms is clearcut, and make two diagnoses if necessary. If there is no disease consider whether there are features that deserve longer-term follow-up that might indicate the patient is in the early stage of a slowly progressive disease. Finally, accept that you will sometimes get it wrong or fail to anticipate the development of a disease, however careful you are. Studies of functional symptoms from the 1970s onwards coalesce around frequencies of misdiagnosis of about 5% after 5 years (Stone et al., 2005). This is the same rate of misdiagnosis for most neurologic and psychiatric disorders, and probably at least as common as misdiagnosis of functional symptoms as disease.

Overreliance on and poor interpretation of neuroimaging

There is a tendency among many physicians to forget that normal neuroimaging does not exclude neurologic disease. That list is very long indeed and includes

amyotrophic lateral sclerosis, Parkinson's disease, epilepsy, and migraine.

Conversely, imaging frequently throws up incidental abnormalities which are of no relevance to the presentation. Many patients with functional disorders suffer iatrogenic damage from the failure of health professionals to place the results of spinal or brain imaging in the correct context.

As a general rule of thumb, your chance of seeing an incidental structural abnormality like a cavernoma or arachnoid cyst on neuroimaging is around 1 in 37 – the same as a roulette wheel. The chance of seeing any kind of "abnormality" such as white dot is around 1 in 6 for a 40-year-old – this time, Russian roulette (Morris et al., 2009).

For spinal magnetic resonance imaging, the frequency of disc degeneration, signal loss, and bulge is around 10% greater than numeric age in years. The frequency of disc protrusion is 30% by age 20 and climbs to 45% by age 80 (Brinjikji et al., 2014).

Diagnostic pitfalls in dissociative (nonepileptic) attacks

Several features can lead to confusion, both in terms of mistakenly calling attacks nonepileptic when they aren't (Smith, 2012), or missing the diagnosis of dissociative (nonepileptic) attacks (Table 15.6).

The diagnosis of dissociative attacks should be made on the basis of objective signs, such as eyes closed, resistance to eye opening, ictal or postictal weeping, and prolonged attacks (Avbersek and Sisodiya, 2010). The problem is that the evidence for many of these signs comes from videotelemetry studies and, in the real world, witnesses, including medical ones, can be very unreliable. For example, reports of eye closure from witnesses may be close to useless when compared with video-electroencephalogram evidence (Syed et al., 2008). Therefore the patient's subjective seizure experience is also important in giving additional clues. Simple questions such as whether the patient "remembers the shaking" can be helpful (Avbersek and Sisodiya, 2010) but there is also evidence that there are conversational features of seizure description typical of dissociative attacks, including reluctance to describe symptoms or giving a poorly detailed description (Plug and Reuber, 2009).

Frontal-lobe seizures can be associated with retained awareness or pelvic movements that can lead to assumptions that the patient may be "acting out" abuse (Geyer et al., 2000). It is particularly important to remember that in temporal-lobe epilepsy there can be quite a long prodrome lasting minutes in which the patient may have fear and dissociative symptoms similar to a patient having a dissociative (nonepileptic) attack (Goldstein and

Table 15.6

Dissociative (nonepileptic) attacks: mimics and chameleons

Feature of epilepsy and other neurologic disorders that can look like dissociative (nonepileptic) attacks	Diagnostic clues/how to avoid error
Generalized tonic-clonic seizure	Include: ictal guttural cry (not weeping) typically at onset, stertorous breathing, eyes open
Frontal-lobe seizures	Short duration (less than 30 seconds) Retained awareness during seizures Shouting, truncal, or cycling leg movements Onset often/mostly from sleep
Temporal-lobe seizures with ictal fear	Progression to generalized seizure. Structural cause. Many temporal-lobe features (e.g., olfactory hallucinations, macropsia) can appear in a dissociative nonepileptic attack
Self-induced epilepsy	Some patients with epilepsy can induce their own seizures, or may manipulate their medication to do so
Autoimmune limbic encephalitis (e.g., NMDA, anti-VGKC)	Patient may present with psychiatric symptoms, unusual behavior, and focal seizures
Stress-induced seizures or syncope	Some epileptic seizures and cardiac syncope (e.g., long QT-related) can be triggered by emotional stress
<hr/>	
Features of dissociative (nonepileptic) attacks that can wrongly put you off the diagnosis	
Olfactory hallucinations	Reports of “burning rubber”/“feces”/“chemical smell” appear quite commonly in dissociative attacks
Dissociative experiences	Depersonalization, visual and perceptual changes in dissociative attacks can sound like temporal-lobe epilepsy
Eyes open	Although “eyes closed” is a good clue, some patients with dissociative attacks do open their eyes (with rolling) during attacks
Cyanosis/breath holding	Including low oxygen saturations
Injury	Bitten tongue (sometimes visibly), broken teeth, (recurrent) shoulder dislocation, and falls on stairs all occur in dissociative attacks. Reports of injury may be more common than actual injury
Incontinence	Urinary incontinence is common and fecal incontinence does happen in dissociative attacks
Seizures arising from sleep/when alone	Occurs in dissociative attacks
Response to trial of anticonvulsants/relapse of attacks when anticonvulsants withdrawn	Patients with dissociative attacks may experience both strong placebo effect when anticonvulsants are started and nocebo effect when they are stopped
The patient in ITU who several nonneurologist physicians and anesthetists are convinced is in status	Prolonged events are a risk factor for dissociative (nonepileptic) attacks. Up to 50% of patients attending hospital in apparent status have this diagnosis

NMDA, *N*-methyl-D-aspartate; VGKC, voltage-gated potassium channel; ITU, intensive therapy unit.

Mellers, 2006). Although ictal fear is usually distinguishable from a panic attack by the shorter duration, associated temporal-lobe features, and impaired awareness (Beyenburg et al., 2005), this is not such an easy distinction to make with dissociative nonepileptic attacks, which may have all of these features.

Features of dissociative attacks can easily put clinicians off the diagnosis, such as injury (Peguero et al., 1995) (and especially report of injury), olfactory hallucinations, and going blue.

In clinical practice it is not always possible to be sure what a patient’s attack disorder is due to, even with all this information. For this reason, the careful neurologist strikes a balance between making confident diagnoses where possible, but saying “not sure” where appropriate. In any patient it is important not to completely close the book on the diagnosis, checking seizure descriptions each visit and watching out for the combination of both epilepsy and dissociative (nonepileptic) attacks.

Diagnostic pitfalls in functional motor disorders

The diagnosis of functional motor disorder should always be based on positive evidence on the examination of internal inconsistency (e.g., Hoover's sign for paralysis or a tremor that stops or entrains during contralateral cued rhythmic movement) (Stone, 2009). However, there can be difficulties in overinterpretation of these positive signs and it would be unreasonable to expect them to always perform, especially in isolation. The presence of pain in a limb, inattention or neglect, or simple failure to understand the examiner's instructions are all reasons why these signs may be false positive.

As with epilepsy, things that look bizarre, like stiff-person syndrome or generalized dystonia, particularly if they are inherently somewhat variable, can fool the

unwary into a diagnosis of functional symptoms. The list in Table 15.7 is obviously not comprehensive. Orthostatic tremor (a movement disorder only present on standing), alien-limb phenomena in corticobasal degeneration, and the aura of paroxysmal kinesogenic dyskinesia are just some of the reasons why the diagnosis of functional neurologic disorders should usually be made by a neurologist familiar with the breadth of unusual presentation neurologic disease has to offer.

Conversely, in patients who do have functional disorder there can be surprising findings in some patients. Just as reflexes can be brisk in patients who are anxious, we have seen patients with unilaterally increased reflexes as a transient phenomenon. Such reflex asymmetry was well reported in the older literature (Allen, 1935). Occasionally patients with unilateral motor symptoms also develop something that looks very similar to ankle

Table 15.7

Diagnostic pitfalls in functional motor disorders

Neurologic diseases that can look like functional motor disorders	Diagnostic clues/how to avoid error
Higher cortical gait disturbance/bizarre gait	Don't rely on an odd gait to make the diagnosis
Acute parietal stroke/pathology	May have Hoover's sign/MRI brain
Stiff-person syndrome	Anti-GAD antibodies
Dystonia (<i>geste antagoniste</i> , better walking backwards or running)	Familiarity with clinical presentation of organic movement disorders
Myasthenia (variability, give-way weakness)	Avoid diagnostic weight on Tensilon tests, which can be false positive (even when blinded) in patients with chronic fatigue syndrome. Strong placebo response to steroids may also occur in patients with functional symptoms
Pain with weakness in limbs	Place less reliability on positive signs of functional weakness in presence of pain. Ask patient if s/he thinks pain is the reason the limb is weak
Paroxysmal dyskinesia, especially with aura and urge to move	Familiarity with clinical presentation of organic movement disorders
Tics/Tourette's	Ability to suppress with rebound movements (may be distractible)
Features of functional motor disorders that can wrongly put you off the diagnosis	
Variable ankle clonus	Happens
Facial symptoms	Common
Slightly asymmetric reflexes/mute plantar	Happens
Contractures in fixed dystonia	Happens
Migraine at onset	Separate trigger from current cause
Tremor unaffected by distraction	In chronic functional tremor motor distraction tasks sometimes no longer visibly affect the tremor. Tremor analysis or video recording may be helpful
Urinary retention	Appears to be quite common in patients with acute back pain and leg weakness in the absence of structural changes. Also occurs with opiate use (Hoeritzauer et al., 2015)
Axial propriospinal myoclonus	Usually functional (van der Salm et al., 2014)
Convergent spasm leading to apparent sixth-nerve palsy	Look for variability over the assessment or resolution with a more distant target

clonus but is variable between assessments. It is not unusual for plantar responses to be mute on the same side as functional hemisensory loss.

Facial symptoms, typically with contraction of orbicularis oculis, oris, and platysma, and sometimes with jaw deviation are clinically quite common. They were well described in the older literature and have recently been described again in more detail (Fasano et al., 2012). Since these facial symptoms lead to an appearance of weakness (even though they are due to muscle over-activity), this can result in erroneous diagnosis of stroke if the presentation is acute.

Slightly better known, although still missed commonly, is the phenomenon of convergence spasm which is relatively common in patients with functional motor symptoms (Fekete et al., 2012). You can usually bring this out by asking the patient to converge on a near target for 10 seconds. In convergent spasm the convergence persists long enough to produce the appearance of impaired abduction, which can be mistaken for a sixth-nerve palsy. One way round this is to go back and test eye movements without convergence and using a more distant target at a different point in the assessment or just observe eye movement during the consultation to show the inconsistency.

Contractures can cause concern about a diagnosis of functional or fixed dystonia, but these do occur in patients who have been immobile for a long time, albeit they are rare and should at least prompt some reconsideration of the diagnosis. They can be demonstrated under anaesthesia.

CONCLUSION

The assessment of patients with functional symptoms can be viewed as difficult, but we believe that, with an appropriate structure and technique, such consultations can be conducted in a much more collaborative fashion that is much more satisfactory for both doctor and patient.

Attention paid to all the physical symptoms in the presentation, exploration of illness beliefs, and a potential mechanism of onset can all pay dividends at the time of explanation (discussed separately in Chapter 44). The assessment of comorbid psychological symptoms is not essential for diagnosis at the first assessment and is often best done at the patient's own pace. Psychological comorbidity can lead to diagnostic pitfalls both when present and absent, leading to overdiagnosis and underdiagnosis of functional disorders respectively. The assessment of the neurologic symptoms themselves has many pitfalls, notably in the interpretation of investigations and in remembering that functional disorders and neurologic disease may coexist or the former may precede the latter.

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