EDITORIAL

Dissociative psychopathology, non-epileptic seizures, and neurology

The latest edition of the International Classification of Diseases (ICD-10) defines dissociation as “...a partial or complete loss of the normal integration between memories of the past, awareness of identity and immediate sensations, and control of body movements” (p 151). Thus defined, the concept of dissociation captures a heterogeneous group of psychiatric conditions previously subsumed within the broader construct of hysteria. The dissociative (conversion) disorders category in ICD-10 comprises subcategories of dissociative amnesia, dissociative fugue, trance and possession disorders, dissociative anesthesi and sensory loss, dissociative motor disorders and dissociative convulsions, and waste basket categories included to capture uncommon, impure, or less severe instances of dissociation. Other symptoms previously viewed as hysterical are placed within the somatoform disorders category of ICD-10, on the grounds that their primary characteristic is an apparent disturbance of bodily rather than cerebral function. By contrast, DSM-IV2 classifies convulsions, motor dysfunction, and sensory loss as somatoform disorders, despite adopting an otherwise similar definition of dissociation and categorization of dissociative complaints. In the context of this editorial we embrace the more inclusive scheme offered by ICD-10.

The basis for their inclusion within current psychiatric nosology is the assumption that dissociative disorders are primarily psychological in nature. Nevertheless, the concept of dissociation has important implications for practice and research within the neurological sphere also. Dissociative psychopathology was witnessed almost exclusively within the neurological setting until the late 19th and early 20th centuries, and many such patients present to neurologists even now. What is striking about dissociative symptoms is their apparent similarity to those found in many common neurological conditions, and it is often extremely difficult to distinguish between the two; the case of dissociative convulsions provides an instructive example.

Dissociative convulsions (non-epileptic seizures)

Recent estimates suggest that between 9% and 50% of patients referred to specialist epilepsy centres have paroxysmal events that, despite resembling true epileptic episodes, are actually non-epileptic. In some instances, patients with reliably established epilepsy exhibit additional seizures of apparently unknown origin, although many patients may present exclusively with these so-called non-epileptic attacks. Although some non-epileptic seizures may be attributable to physical causes other than epilepsy, a demonstrable organic basis is absent in many such patients, suggesting that psychological factors are instrumental in their pathogenesis. Of these cases, a proportion may be associated with an identifiable psychiatric illness such as psychosis or depression that may account for the seizure-like symptoms (for example, depressive fugue). A significant number, however, present with so called “pseudoseizures” or dissociative convulsions, as an isolated psychiatric condition in its own right; it is the recognition of these cases that represents a particular challenge to neurologists working within this domain.

There is no completely reliable set of procedures for identifying dissociative attacks and distinguishing them from epileptic events can be an extremely difficult task. As a result, many patients with dissociative attacks have received an erroneous diagnosis of epilepsy, a misjudgment that can have far reaching implications. Anticonvulsant medication misprescribed to non-epileptic patients with so-called “pseudoseizures” or dissociative convulsions, as an isolated psychiatric condition in its own right; it is the recognition of these cases that represents a particular challenge to neurologists working within this domain.

The diagnosis of dissociative attacks requires the exclusion of epilepsy and other physical disorders; indeed, DSM-IV places dissociative convulsions in the somatoform rather than the dissociative disorders category to emphasize the importance of excluding physical illness in the differential diagnosis of these episodes. Several factors should be considered in the diagnostic process and judgments should always be based on converging lines of evidence. A three pronged approach comprising adequate EEG, a description of preictal, ictal, and postictal events, and careful clinical history taking is the best strategy in the first instance.

The absence of ictal EEG discharges characteristic of epilepsy is a good sign that an attack is non-epileptic in nature although this may not be true for some partial
Although there has been little agreement across researchers, experience a typical episode during the monitoring period, diagnosis of seizures, it is an expensive procedure, and is zure activity with brain electrophysiology. In addition, recording (video-EEG telemetry) is one of the most useful the patient in everyday life. Simultaneous video and EEG recordings in particular are useful for determining the congruence between an individual attack occurring in the hospital or laboratory and those typically displayed by the patient in everyday life. Simultaneous video and EEG recording (video-EEG telemetry) is one of the most useful techniques as it allows for the correlation of observable seizure activity with brain electrophysiology. In addition, telemetry is extremely useful for assessing whether ictal EEG changes are cerebrally derived or artifactualy produced by episode related movements.

Although video-EEG telemetry has revolutionised the diagnosis of seizures, it is an expensive procedure, and is not widely available. Moreover, many patients will not experience a typical episode during the monitoring period, particularly if the interval between seizures is long. There are techniques to provoke seizures that could address this problem although, in certain cases, their use remains highly controversial. Sleep deprivation and photic stimulation are generally regarded as ethical but may not be effective. Other techniques involve “suggesting” seizures using, for example, a placebo injection or hypnosis, which the patient is told will automatically provoke an attack. Seizures triggered using this and similar techniques are then compared with the patient’s typical attacks and, assuming a match is found, the video-EEG record can be used for diagnostic purposes. Although suggestive techniques can successfully generate seizures in many cases, the use of deception of this sort is considered unethical by many clinicians and it is not widely used in the United Kingdom for this reason. It is also important to note that the suggestibility of a seizure is not, in itself, a reliable guide to diagnosis, as both epileptic and non-epileptic attacks can be provoked using these techniques. Moreover, contrary to established clinical lore, there is little systematic evidence demonstrating that those with dissociative attacks are any more suggestible than those with epilepsy.

Other clinical features associated with the ictus have also been offered as potentially useful diagnostic indicators, although there has been little agreement across researchers and empirical findings have proved inconsistent. Moreover, there is no such thing as a “typical” non-epileptic seizure; whereas some ictal features may be suggestive of a particular diagnosis, the practical value of these features should be viewed with at least a degree of scepticism. It is also necessary to be alert to the possible unreliability of third party descriptions of seizure related events. Such descriptions are typically given by friends, relatives, or other lay persons whose reports are easily tailored to the enquiring physician’s expectations.

Careful clinical history taking is also a central element in the assessment of seizures. It is important to appreciate, however, that alteration of consciousness is not uncommon in patients with non-epileptic seizures and such a history cannot therefore provide an adequate foundation for a diagnosis of epilepsy. Age of seizure onset may be a useful diagnostic indicator, with late onset being suggestive of a non-epileptic basis, especially if an MRI is normal. Sexual or physical abuse may be common in patients with dissociative attacks and should be sensitively enquired about; given the potential for underreporting, repres- sion and so called false memories one should, however, be judicious in accepting the face validity of any such reports. Practitioners should also bear in mind that epilepsy itself may arise from cerebral pathology sustained through childhood physical abuse and should exercise caution accordingly.

Other clinical features have been associated with dissociative seizures, although such features cannot by themselves be regarded as sufficient for an inclusive diagnosis of non-organic pathology. Potentially suggestive of a dissociative diagnosis are a personal and family history of psychiatric illness and the presence of comorbid psychopathology, a family history of epilepsy, and evidence of clear psychosocial seizure precipitants.

Rhythms are often paroxysmal and may acquire the much unilateral or bilateral, sometimes with sharp components. Such records can provide information on the neurological plausibility of ictal events and, in certain cases, indicate whether dissociative attacks are likely. Video recordings in particular are useful for determining the congruence between an individual attack occurring in the hospital or laboratory and those typically displayed by the patient in everyday life. Simultaneous video and EEG recording (video-EEG telemetry) is one of the most useful techniques as it allows for the correlation of observable seizure activity with brain electrophysiology. In addition, telemetry is extremely useful for assessing whether ictal EEG changes are cerebrally derived or artifactualy produced by episode related movements.

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Raised postictal serum prolactin concentrations (greater than 1000 IU/l) in the presence of normal baseline values are found after epileptic seizures, although this is limited to certain seizure types; moreover, false positives can occur (for example, with syncope). Self injury is common in both epileptic and dissociative seizures, although the site and severity of lesions often differ between the two. Carpet burns, for example, are almost always indicative of non-epileptic attacks. The presence of other dissociative “stigmata”, such as unexplained sensory loss, may also support a non-epileptic diagnosis. On the other hand, incontinence is typically a poor guide to seizure type.

The ease with which epileptic and dissociative events are confused, coupled with the serious implications of misdiagnosis, demonstrates that physicians must be aware of the various clinical phenomena that may present as epileptic and non-epileptic seizures, and the converging lines of evidence that are required before a firm diagnosis can be made in either direction. Making a diagnosis is clearly a complex and difficult process that requires an experienced clinician, with both neurological and psychiatric knowledge, and the support of adequate EEG and neuroimaging facilities. As in so many areas of medicine, the more quickly a firm diagnosis can be established, the better the prognosis.

### Presenting the diagnosis of dissociative seizures

Once a firm diagnosis has been reached, presenting that diagnosis to the individual patient is the first, and one of the most important, parts of treatment. Considerable care needs to be taken at this point, especially when a previous, incorrect diagnosis has been in place for some time. Contrary to what many neurologists think, a patient is unlikely to be jubilant to discover that their seizures, previously identified as epileptic, are actually dissociative in nature. In the first instance, patients are likely to be upset that they have been given an erroneous diagnosis in the first place, particularly as they may have encountered prejudice as a result of the epileptic label. Secondly, most patients will be angry if they have been given unnecessary medication, especially if unpleasant drug side effects have had a large impact on their quality of life. Thirdly, coming to terms with the psychiatric diagnosis itself is difficult for many patients, who often think that their suffering is being dismissed as “all in their mind” or, worse still, deliberately faked for secondary gain. Although there are cases where seizures have been consciously simulated (“malingering”),
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such cases are rare except in certain settings. Unless there is good reason to suspect simulation (for example, impending legal claim coupled with an evasive attitude to assessment and an inconsistent seizure presentation/clinical history etc.), it should be assumed that the events are subjectively genuine and treated accordingly.

The diagnosis of dissociative seizures is ideally presented by a psychiatrist or neuro psychiatrist who is a member of the team involved in the patient’s care. To begin with, the patient should be informed of the positive news that their attacks are not the result of cerebral damage and abnormal brain activity and are not, therefore, epileptic in nature. At this point, they should be told that anticonvulsant medication will be of no benefit to them and, if they are taking any such medication, that it should be withdrawn slowly but as soon as possible. In some cases, showing the patient the video-EEG record on which their diagnosis was based may help them to accept the non-epileptic nature of their attacks.

Patients should then be gently introduced to the notion that their seizures are caused by psychological rather than neurological factors. Resistance to this idea may be reduced if it is emphasised that such a diagnosis does not mean that their seizures are “faked” or that they are any less real or distressing than “genuine” epileptic attacks; patients should also be reassured that the diagnosis does not mean that they are “insane” or “crazy” (assuming the absence of other significant psychiatric problems). Many patients will raise the legitimate question of how it is possible for their attacks to be psychological when they are unaware of any emotional upset or psychological problems, or any link between such problems and their seizures. The clinician should use such an opportunity to introduce the idea that dissociative seizures are normally caused by unconscious psychological processes, emphasising the role of such processes in normal behaviour and giving everyday examples (see Devinsky and Thacker) where possible. Examples demonstrating the close link between mind and body (for example, palms sweating when nervous) may be particularly useful in this respect.

For many patients, coming to terms with a psychological explanation for their attacks will be far from easy. As such, presenting the diagnosis of dissociative seizures in a confrontational manner is unlikely to be of use and will increase resistance to the diagnosis in many cases. Rather, the clinician should be as supportive and reassuring as possible, emphasising that psychological attacks often have a better prognosis than those associated with epilepsy. Careful use of terminology is extremely important and certain expressions should be avoided. The term “psuedo seizure”, for example, often meets with resistance as it seems to imply that the patient’s seizures are faked. Also to be avoided are the terms “hysteric” and “hysterical”, which are pejorative and have acquired lay connotations that are entirely misleading. In our experience, patients also strongly object to being informed that their seizures are “all in the mind” or “imaginary”. We think that the term “non-epileptic attacks” is probably the most appropriate in this context as it is accurate, theoretically neutral, and acknowledges the authenticity of the patient’s complaint.

Such an approach allows the patient to accept the diagnosis while saving face, something that is particularly important in the context of the patient’s interaction with their family and wider social circle. Indeed, in many cases it is desirable, where permission has been given, for the clinician to inform members of the patient’s family of the non-epileptic diagnosis also.

Treatment issues
Once the diagnosis of dissociative convulsions has been presented, treatment can begin. Depending on the patient, some form of psychological therapy is typically the approach of choice, and there is limited evidence indicating the efficacy of such a treatment strategy. However, there are few, if any, adequately controlled studies addressing this issue empirically. Ideally, treatment will be conducted by a psychiatrist, psychologist, or psychotherapist who is part of the multidisciplinary team involved in the patient’s care from the outset. Where this is not possible, the referring physician should remain involved to some degree so that the patient does not feel “abandoned in the hands of a psychiatrist.”

To begin with, the presence of comorbid clinical anxiety or depression should be addressed, often with pharmacotherapy. Psychological therapy should then be considered, with the specific approach depending largely on the nature of the individual patient and the presenting problem. In some cases, psychotherapy addressing the emotional issues surrounding the occurrence of dissociative events may be of some use. However, such an approach requires a certain degree of insight and psychological mindedness on behalf of the patient and not all patients have the capacity to face the emotional demands made by this sort of treatment. Cognitive and behavioural approaches aimed at identifying and controlling the factors precipitating and maintaining dissociative seizures are less demanding than psychotherapy and seem to work in many cases. Behavioural approaches seem particularly well suited to the treatment of those with a learning disability, and/or where secondary gain seems to be an important maintaining factor. In many cases, family therapy may also be an important aspect of treatment, particularly where a dysfunctional family dynamic seems to be contributing to the occurrence of the dissociative attacks. Some clinicians also advocate the use of hypnosis as an adjunct to the psychological therapy of these patients. Such an approach may be of some use, although the patient’s beliefs about hypnosis and their attitude towards it should be carefully explored before proceeding.

The few studies considering treatment outcome in patients with dissociative attacks suggest that the prognosis of these patients is actually fairly good, given careful psychological management and early diagnosis. However, there is a desperate need for well controlled, systematic studies comparing the various treatment approaches to dissociative attacks and assessing the factors predicting positive outcome.

Understanding and operationalising dissociation
The confusion surrounding the differential diagnosis of non-epileptic seizures provides just one example of the challenge that dissociation represents within neurological practice. As the dissociative disorders categories of ICD-10 and DSM-IV indicate, the symptoms of many neurological conditions have a dissociative counterpart. Thus, both neurological and psychiatric examples of amnesia, fugue states, motor problems, sensory loss, stupor, and coma have been described, with the list apparently only limited by the number of neurological symptoms currently subject to classification. In many such cases, practitioners attempting to separate dissociative from organic phenomena encounter the same diagnostic dilemma as that discussed here for epilepsy and non-epileptic seizures.

The principle problem in this context is that the diagnosis of dissociation is primarily based on the exclusion of organic pathology rather than the preferred set of psychiatric inclusion criteria. Identifying such criteria promises to be a laborious task requiring a degree of conceptual and
methodological sophistication at present lacking in this domain. Current theory concerning the dissociative disorder continues to be dominated by the ideas of Janet25 and his conceptual successor Hilgard,26 and many Freudian concepts also remain popular. Such accounts are, however, vague and do not lend themselves well to empirical evaluation,27 leading to a descriptive rather than a mechanistic understanding of the term “dissociation”.28 The shortcomings of existing theory and research are well illustrated by a recent study29 attempting to distinguish organic and non-organic epileptic events using the dissociative experiences scale (DES),30 a putative operationalisation of dissociation. Although dissociative (non-epileptic) patients would be expected to report more dissociation than their non-dissociative (epileptic) counterparts, total DES scores were not significantly different between these groups,30 despite being relatively high in both cases. Moreover, any modest differences on DES factor or item scores appeared attributable to discrepancies in sexual and physical abuse reported by epileptic and non-epileptic patients. Thus, whereas patients with so called dissociative psychopathology have “dissociative” experiences as part of their clinical presentation, this study clearly demonstrated that patients with non-dissociative, organic pathology also have such experiences, at least as measured by the DES. Indeed, a recent review by Good32 advocates that dissociative experiences may be common in other physical conditions, demonstrating the existence of so called “organic dissociation” akin to the concept of “organic repression” suggested by Schilder33 many years ago.

Although measures such as the DES allow us to assess the degree to which patients experience apparent disruptions in consciousness, memory, identity, or control, they do not provide a means for evaluating which processes, psychiatric or neurological, actually underlie any supposed disturbance. Current theory concerning the dissociative disorder continues to be dominated by the ideas of Janet25 and his conceptual successor Hilgard,26 and many Freudian concepts also remain popular. Such accounts are, however, vague and do not lend themselves well to empirical evaluation,27 leading to a descriptive rather than a mechanistic understanding of the term “dissociation”.28 The shortcomings of existing theory and research are well illustrated by a recent study29 attempting to distinguish organic and non-organic epileptic events using the dissociative experiences scale (DES),30 a putative operationalisation of dissociation. Although dissociative (non-epileptic) patients would be expected to report more dissociation than their non-dissociative (epileptic) counterparts, total DES scores were not significantly different between these groups,30 despite being relatively high in both cases. Moreover, any modest differences on DES factor or item scores appeared attributable to discrepancies in sexual and physical abuse reported by epileptic and non-epileptic patients. Thus, whereas patients with so called dissociative psychopathology have “dissociative” experiences as part of their clinical presentation, this study clearly demonstrated that patients with non-dissociative, organic pathology also have such experiences, at least as measured by the DES. Indeed, a recent review by Good32 advocates that dissociative experiences may be common in other physical conditions, demonstrating the existence of so called “organic dissociation” akin to the concept of “organic repression” suggested by Schilder33 many years ago.

Although measures such as the DES allow us to assess the degree to which patients experience apparent disruptions in consciousness, memory, identity, or control, they do not provide a means for evaluating which processes, psychiatric or neurological, actually underlie any supposed episode of “dissociation”. Clearly, we need to move beyond the descriptive approach to dissociation inspired by existing theoretical models,29–32 advocated by current classificatory schemes32 and embodied by measures such as the DES. To do this, the first goal must be to provide a precise definition of dissociation based on a conceptually coherent and empirically justified account of the processes underlying these phenomena. Armed with such a definition, we may turn to the critical task of constructing a satisfactory operationalisation of dissociation that serves to define the criteria for an inclusive diagnosis of dissociative psychopathology. Whether such a process is likely is that many of the phenomena previously identified as dissociative, such as epilepsy related memory loss, will no longer be viewed as dissociative at all!

In our view, neurology will have an important part to play in the development of any satisfactory theoretical account of the mechanisms underlying dissociative psychopathology. To this end, Whitlock4 and Ludwig41 have suggested that the primary pathophysiological mechanism involved in the creation and maintenance of dissociative (or “hysterical”) symptoms is an attentional dysfunction resulting from an increase in the corticofugal inhibition of afferent stimulation. As a result of this inhibition, partially processed stimulus information fails to be integrated into ongoing awareness, generating dissociative symptoms as a consequence. Some studies have provided empirical support for this account of dissociation.30 Part of the appeal of the approach advocated by investigators such as Ludwig is their emphasis on integrating research and theory from neurology, psychiatry, and psychology in a bid to understand the mechanisms of dissociation.

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Chronic fatigue syndrome: is it physical?

It is increasingly recognised that chronic fatigue syndrome (CFS) is heterogeneous. A significant proportion of patients fulfilling operative criteria for a diagnosis of CFS will also fulfill criteria for a psychiatric disorder, such as depression or somatisation. Failure to recognise this heterogeneity prejudices attempts to understand CFS in cross sectional studies. In this issue (pp 302–307) Fulcher et al report a study of muscle strength, aerobic exercise capacity, and functional incapacity in a group of patients with CFS without concurrent psychiatric disorder, compared with patients with major depression and a group of normal but sedentary subjects. In an incremental treadmill exercise test, patients with CFS and depressed patients had lower peak oxygen consumption rates, maximal heart rates, and plasma lactate concentrations than the sedentary controls; but this reflected the shorter duration of exercise tolerated by these patients. At submaximal work rates, patients with CFS and depressed patients experienced greater perception of effort than sedentary controls at the same level of work. This is in keeping with the finding that such patients show greater sensitivity to bodily sensations than normal subjects. Overall, there was little difference between the patients with CFS and the depressed patients in exercise characteristics, yet the patients with CFS reported significantly greater degrees of physical fatigue and physical incapacity.

The authors did find one important difference, however. The patients with CFS were significantly weaker than either the depressed patients or the sedentary controls as judged by measurement of quadriceps strength. This is the first study to demonstrate such physical weakness in patients with CFS and the authors suggest that this is because they studied a more coherent group of patients than others have previously.

Fatigue is a complex symptom. In its strictest sense the word means “inability to sustain force”, indicating dysfunction in the neuraxis or neuromuscular apparatus, whether physiological (after strenuous exercise) or due to disease processes. In its colloquial context (fatigare-to tire), it is the sensation experienced when the effort to perform work, whether physical, mental or both, seems disproportionate for the task involved. Physiological fatigue recovers with rest. Chronic fatigue typically does not. Although chronic fatigue is a very common complication of a wide range of medical and neurological diseases, such as multiple sclerosis and Parkinson’s disease, it can also occur in the absence of readily definable pathology, notably in CFS.

What is the cause of this physical weakness in CFS? The authors think that it is related to physical deconditioning due to inactivity and this view is supported by improvement in wellbeing, strength, and exercise capacity after a graded aerobic exercise programme. However, graded exercise therapy improves tolerance and exercise capacity in many disorders, including muscle diseases such as mitochondrial myopathies.

Some patients with CFS show abnormal increases in plasma lactate after exercise at low work rates. Heart rate responses to exercise in that study did not suggest that these patients with CFS were more “deconditioned” than those with normal lactate responses. They also proved less likely to have psychiatric disorder. Furthermore, recent spectroscopic studies of the forearm muscles, which are not usually subject to the effects of deconditioning, showed abnormal muscle energy metabolism in such cases. Deconditioning may well be an important factor in the pathogenesis of CFS, but these findings raise the possibility that some patients with CFS have a form of metabolic myopathy.

Whatever the mechanisms underlying “fatigue”, exercise therapy is likely to become an increasingly important therapeutic modality in various fields and particularly in the management of chronic fatigue syndromes.

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What contributes to quality of life in patients with Parkinson’s disease?

The question posed by Schrag et al (this issue, pp 308–312) is fundamental, but can it be answered? Quality of life belongs to a family of terms which has an improbably wide range of applications, from “quality” as an object of philosophical contemplation to “QoL” as an economic variable. It must be borne in mind that measures such as the PDQ-39 and the SF-36 differ considerably in content. More detail on the parallelism between the PDQ-39 and two other measures would be helpful but the data seem to support the assumption, which is gaining ground in the literature, that such measures reflect a single underlying dimension. Scales specific to Parkinson’s disease and to inflammatory bowel disease have been found to be congruent, again suggesting that disease specific quality of life is a unitary construct. However, we must not lose sight of the fact that at the deepest level, quality of life is inherently unquantifiable.
Philosophical considerations aside, there is practical merit in the more precise question: “What are the associations between this particular measure and other variables?” We need to understand which aspects of Parkinson’s disease weigh most heavily on patients. Although the authors recognise that associations do not indicate causality their bias is revealed when they write of the “influence” of factors such as mood on quality of life. The converse is equally likely. The two dimensions, mood and quality of life, are logically related, and the extent to which the Beck depression inventory correlates with the PDQ-39 tells us as much about the nature of the two measures as about the patient population under study. Much of the variance was not shared, suggesting that a fraction of the putative quality of life construct did not influence mood (why not?); or that aspects of the measure of mood did not influence quality of life: which sounds paradoxical.

The authors found that severity of disease (or rather severity of physical impairment, as this is what the Hoehn and Yahr scale measures) was more predictive than disability of the PDQ-39 score. The relation between physical impairment and quality of life therefore seems to have been indirect, and not mediated through the obvious effects of impairment on activities of daily life. This is consistent with several other studies showing that the association between disability and quality of life is weak.2 Severity of impairment might have been a marker for diverse experiences which were not individually predictive of PDQ-39 score. It is not clear whether the association between postural instability and PDQ-39 was direct or indirect.

Similar considerations apply to the association between the mini mental state examination (MMSE) score and PDQ-39. We should not accept uncritically the implication that dementia is experienced negatively. It could be an indirect marker of some other negative influence, such as carer stress. One problem which the authors do not consider is that quality of life and also depression are notoriously difficult to measure validly in people with dementia. In this sort of study it is virtually impossible to avoid the influence of carers’ opinions on questionnaire responses, especially at the lower end of the MMSE scale, and the authors apparently made no attempt to do so. Carers add a further layer of complexity to the interpretation of these results.

Can a neuropsychological follow up contribute to the diagnosis of parkinsonian syndromes?

Although an akinetic rigid syndrome is the hallmark of idiopathic Parkinson’s disease (PD), striatonigral degeneration (SND), and progressive nuclear palsy (PSP), cognitive and behavioural dysfunctions are often found in these parkinsonian syndromes. If the neuropsychological deficits directly reflect underlying brain neuronal lesions, we may expect that the cognitive and behavioural changes associated with these syndromes depend on the specific pattern of subcortical or cortical lesions that characterise each disease. The earliest and major cognitive deficit in all three diseases is impairment of executive functions as shown by performance on tests sensitive to frontal lobe function. Is it possible to distinguish, among the deficits contributing to this subcorticofrontal syndrome, patterns that are more specific to one or another of the three diseases? This might be useful for clinical diagnosis, especially in early stages of the diseases when the signs often overlap, accurate diagnosis being important for devising investigational and therapeutic strategies. In the paper by Soliveri et al4 in this issue (pp 313–318) patients with PSP performed worse than patients with PD and those with SND on most tests during their initial evaluation, whereas patients with PD and those with SND performed similarly. These results are in agreement with other recently published studies.5,6 Patients with SND had, however, reduced phonemic fluency compared with patients with PD. This difference has to be confirmed by other studies, but might be clinically relevant. Dementia was found only in PSP, and corresponds to a severe subcorticofrontal syndrome caused by lesions in the the striothalamocortical pathways resulting in deafferentation of premotor and prefrontal areas.

Because the natural history of these disorders also varies widely, Soliveri et al attempted to enlarge the differential diagnosis by a longitudinal assessment. Unfortunately, after 2 years, significant proportions of patients with SND or PSP were too disabled or had died, confirming that these two diseases progress more rapidly than PD. Among the patients who could still be tested, those with PSP declined significantly more than the two other groups on the Wisconsin card sorting test, which evaluates conceptualisation and set shifting. Patients with PSP or SND, however, had worsened significantly more than patients with PD on the visual search test assessing attention and visual scanning. There was no correlation between motor and cognitive decline. The proportion of demented patients increased only in PSP. This is not surprising given the more diffuse neuronal lesions in this disease.

In conclusion, clinical studies indicate that a parkinsonian syndrome is the presenting feature of several different diseases. Although most patients with parkinsonism have an idiopathic Parkinson’s disease, others have multiple system atrophy, progressive supranuclear palsy, corticobasal degeneration or Lewy body dementia. Neuropsychological assessment in the early stages of the disease can contribute to the clinical diagnosis. The time course and pattern of progression of cognitive and behavioural decline can also
improve the diagnosis, as shown by the study of Soliveri et al., particularly when postmortem confirmation can be anticipated.

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