Welcome to the eleventh in a series of articles in ACNR exploring clinical dilemmas in neuropsychiatry. In this series of articles we have asked neurologists and psychiatrists working at the interface of those two specialties to explain their own personal views on today's case-based clinical dilemmas. We would welcome feedback on these articles, particularly from readers with an alternative viewpoint.

Severe Medically Unexplained Neuro-Disability: Should you investigate (again) and is there a cure?

Case

Dear Doctor

Please could you see this 48 year old man; I have just taken over his care. He has a 12 year history of ‘MS’ which began with vertigo, bilateral leg weakness and collapse while working on a building site. He was thoroughly investigated at the time (he has a fat file containing medical notes) and the results were equivocal. He is mostly confined to a wheelchair and has a catheter in situ. He requires carers to come in twice a day. He complains of blurred vision and has double or even triple vision when you test his eye movements. He has backache and is under the local pain team who have him on a transdermal patch. He still complains of dizziness and chest pain when he tries to stand and the cardiologists are investigating both. He also has irritable bowel syndrome and type 2 diabetes. He is rather disgruntled especially with his former employers but not depressed. I was wondering whether it would be useful to have a fresh look at his problems and perhaps carry out some investigations. He had a normal MRI scan of his brain and spine and lumbar puncture two years ago and a neurological opinion was that it was ‘functional’. Surely a person wouldn’t be as disabled as this purely because of conversion disorder (especially with the incontinence)? Do you think it would be worth repeating these investigations? Is he treatable?

We are now well used to hearing that a large proportion of patients seen in neurology clinics have a ‘functional’ disorder – or put another way, 30% have symptoms that are not all or only somewhat explained by neurological disease.1 This medically unexplained group has higher levels of disability and distress and are in receipt of more disability related state benefits than patients with symptoms explained by neurological disease. A chronic course is not uncommon; for example, Stone and colleagues in Scotland2 reported the 12 year prognosis of 60 patients with unilateral ‘functional’ weakness or motor-conversion disorder. Of those followed up, the vast majority reported continuing symptoms and physical limitation; 20% had taken medical retirement. Patients often had other somatic symptoms and in only one did a true neurological disorder emerge. Buried within these common disorders is a subgroup of patients with very severe disabling conditions. A number of case-series have been reported. For example 25 patients referred to liaison psychiatry or neurological disability services in Oxfordshire3 were selected on the basis of persistent severe disability; over half had a diagnosis of motor-conversion disorders with the rest a diverse group of somatoform disorders and chronic fatigue syndrome. Most were unable to walk, unemployed and receiving disability living allowances. A subgroup of 10 of these patients confined to a wheelchair was studied in further depth.4 All had a diagnosis of conversion or somatoform disorder, six had a previous history of major depression. Duration of illness was very long, with wheelchair use being on average 8.3 years; most were regarded as incurable. An earlier study from a spinal injury centre5 painted a rather similar picture. Patients were identified who following their admission were rediagnosed with ‘hysterical paraplegia’. However, the authors reported that the response to intensive physically based rehabilitation prognosis was good.

Switching to the primary care setting, a postal survey to general practitioners, surveying a catchment area population in Nottingham found 18 with conversion disorder6 six of whom were very disabled, bed or wheelchair bound. It was clear that the burden of care for these individuals fell on the general practitioner. The overall prevalence rate works out at 4N/100,000.

There seems to be no limit to the extremes of converting symptoms of disease. As the estimate from Scotland2 showed, even after 12 years it is possible to experience symptoms which are still not explained and now include the whole body. One of the patients in our series mentioned ‘double or even triple vision’ which is explained by a particular pathology (triple vision is usually a manifestation of ophthalmoplegia which in itself is not fully understood in conversion disorder). The ‘functional’ explanation is therefore not only unsatisfactory but also a potential source of patient distress. It is an area which needs further research and clinical interest in functional symptoms within neurology, especially the symptom of weakness. He writes regularly on this topic in scientific papers and for textbooks of neurology and psychiatry.

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severe medically unexplained pseudo-neurological disability that is attributable to psychological or social rather than neurological dysfunction. Sufferers are often characterised by extreme forms of dependence, convoluted and often deeply unsatisfactory encounters with a whole range of medical specialists and finally very expensive and extensive care packages, often drawing resources from health, physical disabilities and social care. They frequently undergo unnecessary investigation and procedures which are not only costly but also reinforce their medical model of illness, especially when minor abnormalities are found and misinterpreted as aetiologically relevant. Such patients lead engendered lives and place an enormous burden on carers and families, not to mention social and health care systems and society as a whole.

Why does this happen? First, there is a lack of a clear diagnostic label – patients with similar clinical presentations acquire such radically different diagnostic labels such as ‘MS’, catatonia, fibromyalgia, chronic fatigue syndrome/ME and so on. Second, there is no clear professional ownership, leading to patients bouncing from doctor to doctor and incoherent care planning; symptoms and disability cross care pathways leading to ‘falling between the stools’ of health and social care; acute care, chronic care and rehabilitation. Finally, such patients engender poor therapeutic relationships – added to which are feelings of helplessness among practitioners (‘heart sink’) and in some instances, strident patient advocacy.

Other diagnostic labels
The case referred to above represents another common scenario where patients have an apparent diagnosis of, for example, multiple sclerosis, stroke or Parkinson’s disease but where the evidence that they are suffering from that condition is at best tenuous or at worst non-existent. Nevertheless the label persists, perhaps with the collusion of professionals with the view to trying to contain and constrain help-seeking medical presentations and providing a mechanism for the affected individuals to receive some kind of acceptable (i.e. physical) support.

While conversion disorder probably accounts for the majority of these cases there are other diagnostic labels under which such patients may be hiding. The diagnosis of factitious disorder tends to be labelled as hysteria/conversion when there is a predominantly neurological flavour to the presentation. The label of factitious tends to be given where there is other evidence apart from the key symptom that the person has been creating symptoms or perhaps misleading the professionals as to the history of previous treatment or extent of disabling symptoms. Follow-up can be subtle and are by no means absent in cases of conversion disorder. Factitious disorder in turn blends hysteria/conversion when there is a predominant neurological flavour to the presentation. The label of factitious tends to be given where there is other evidence apart from the key symptom that the person has been creating symptoms or perhaps misleading the professionals as to the history of previous treatment or extent of disabling symptoms. Follow-up can be subtle and are by no means absent in cases of conversion disorder. Factitious disorder in turn blends hysteria/conversion when there is a predominant neurological flavour to the presentation.

Some dilemmas of management
The case presents two familiar conundrums. The first is, not so much whether to investigate or refer for specialist opinions, but rather when to stop. Reading such a history, albeit brief, it is tempting and indeed justifiable to be thinking that the patient has somatisation disorder – he has a number of medically unexplained symptoms in various systems. He has unexplained severe disability and chronic pain. He also has diabetes, and other conditions of middle age; there is a suspicion of leigned symptoms such as triplopia, perhaps some ongoing dispute or litigation with an employer and possible atypical depression or other psychopathology which may be resistant to treatment. No doubt he is on several different medications so there is also a strong possibility of iatrogenic symptoms. We will not offer specific advice here. A careful review of symptoms, investigations and treatment is essential to obtain a clear picture of what to do in the patient’s best interests but this is likely to be time-consuming and to raise as many questions as answers.

However, a degree of restraint is to be commended and this is supported by evidence. In the mid 1960s, Eliot Slater, neuropsychiatrist at Queen Square published a famous paper of a cohort of patients admitted to that hospital and diagnosed with hysteria. Follow-up after several years revealed, according to Slater’s controversial interpretation, an exceedingly high incidence of neurological disease, which was at times fatal. This tapped into the universal anxiety in clinicians, especially psychiatrists, that they were missing ‘organic’ diagnoses. A follow-up of the same kind of patients three decades later provided considerable reassurance on this matter and a meta-analysis of similar follow-up studies confirmed it. ‘The likelihood of new neurological disease emerging which might explain a hysterical or conversion disorder is very small and has been declining for decades, long before the introduction of MRI and CT.’ Presumably this is due to generally better standards of medicine and diagnostic tools and correspondingly more rigorous reporting of follow-up studies. However, trying to derive a simple ‘take home message’ from this is dangerous. Clearly it would be wrong to say that clinicians needn’t worry about making a proper diagnosis. Refusing to follow up the usual steps to make a diagnosis (including taking account of psychiatric and psychosocial factors) trust your judgement and don’t feel you have to do ‘just one more test’ despite the pressure to do so. Patients may contribute to this pressure and may appear to be reassured by a negative result. So we stress the importance of not getting caught up in the usual steps to make a diagnosis (including taking account of psychiatric and psychosocial factors) trust your judgement and don’t feel you have to do ‘just one more test’ despite the pressure to do so.

The second conundrum raised in the referral is whether the degree of disability goes beyond conversion disorder. There is nothing in this case that might not be explained by conversion disorder. For example, incontinence is often thought to be ‘very rare’ in conversion disorder since surely there can be no net secondary gain for such a distressing symptom? This is wrong in our experience. Another which goes back to Freud and Charcot is the rarity of ‘hysteria’ in men. In fact, Davie from Dagenham is as familiar a figure in 2012 as was Anna O from de siècle Vienna. The combination of chronic somatoform pain (and its treatment) with motor symptoms is also all too familiar.

Is it treatable?

The importance of this problem is not just its cost but the fact that there is a solution. While the prognosis of untreated conversion disorder is poor, there have been several studies which have shown dramatic responses to treatment in these patients once the nature of the condition has been recognised and an appropriate management plan put in place. Much has been written about the principles of treatment which often involve an initial physical focussed rehabilitation and physiotherapy approach becoming more psychologically focussed. Multidisciplinary treatment is recommended in all cases, and treatment of comorbid psychiatric disorders such as major depression forms an integral part of the management. The nature of the setting is often important and placement within a physical rehab unit or neurological hospital appears to maximise engagement and
dealing with the complex dynamics are the skills psychiatrists have in patients and occasionally psychosis are depression, anxiety, personality disorders. Partly because untreated psychiatry presence is nevertheless to ‘nerves’, can be addressed. A strong increasing fatigue or causing damage while concerns and fears, around increasing movement and mobility is most salient to them and they can because physiotherapists are able to find that most admissions require at least 12 weeks of treatment.

Units such as this are rare in the UK but not unique. It is essential that experiences in such units can be compared and contrasted and that outcomes measured and presented objectively. Clinical trials are of course the only means to develop truly evidence based interventions and are being planned.

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REFERENCES


