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EDITORIAL

Reassessing received wisdom in ALS – pain is common when studied systematically

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disorder that presents many challenges for patients and their families; in some cases, these may be compounded by commonly held beliefs about symptoms of the disease. ALS was traditionally held to spare cognition, now refuted by the findings of frontotemporal dementia in a small subset of patients and milder, more subtle cognitive dysfunction (of a predominantly executive nature) in a larger proportion [1]. A related misconception is that patients and caregivers do not wish to be told about the psychological consequences of the disease; most do [2]. Likewise, eye movements, once held to be unaffected in ALS, have been found to be subtly affected on closer examination [3]. Although conceptualized as a purely motor disorder, nearly a quarter of patients experience generalized sensory symptom abnormalities [4], and an fMRI study found higher level dysfunction in visual and auditory processing [5]. The characterization of patients with ALS as being particularly resilient to depression also fails to hold up in that under closer examination, this group appears to be no more or less depressed than patients with other neuromuscular disorders [6]; though, they probably have a lower rate of depression than patients with Parkinson's disease or multiple sclerosis.

Misconceptions about cognition and the sensory system may be understandable given they cannot be observed directly, but even in the case of observable phenomena such as pressure ulcers, Charcot's observation that 'there is no tendency to the formation of bedsores' was only recently refuted by systematic assessment, and the rate of pressure ulcers in ALS is no lower than other neurological conditions [7]. Such myths in the culture of understanding and managing ALS may adversely affect the level of attention and care that patients receive with regard to their symptoms.

In this issue, Chio *et al*. [8] describe a systematic evaluation of pain, another potentially under-recognized symptom of ALS [9]. Although pain as a consequence of stiffness or immobility is understood and managed clinically, patient literature still makes statements to the effect that ALS does not cause pain, pain is not a primary feature, or pain is not a recognized symptom of ALS. Using a prospective epidemiological register, the authors found that most of the patients with ALS (57%) reported pain in the previous week, with 14% in pain rated as 'severe' according to the Brief Pain Inventory. Pain in ALS was most commonly found in the extremities, shoulders, and hips and interfered with patients' enjoyment of life and relationships with others.

This is important work in systematically assessing a previously under-recognized symptom in a well-characterized population with an age-matched healthy control group. Future studies could use the same model to examine other symptoms, introduce a longitudinal element to record changes over time, or (more ambitiously) study the options available for treating the different manifestations of pain in this population.

Despite what we as researchers now know about pain in ALS, reasons given for under-reporting pain clinically might include patients not being asked by the physician, assuming it is an untreatable aspect of the disease, or patients failing to appropriately raise the issue with the clinical team [9]. Since 2006, we have been attempting to address these issues amongst 'e-patients' by systematically asking the 4522 members of PatientsLikeMe with ALS to rate their pain (amongst 10 other symptoms) on a self-report scale that can be brought to the doctor as a printed sheet. Patients can submit reports as often as they like, which has lead to 11 087 total reports of severity; the most recent report from 2664 users of the system with ALS indicates that 6% report pain that is 'severe', 21% 'moderate', 34% 'mild', and 39% 'none'. Empowering patients to report their symptoms, encouraging systematic symptom assessments in clinic, and updating the patient literature around pain may all contribute to improving our management of this important symptom.

With regard to other 'myths' about ALS, we must regularly challenge our preconceived notions of ALS and consider their origins. Although the clinical descriptions of ALS by our scientific forbears were clearly revolutionary, through the progress of neuroimaging, electromyography, genetics, and lengthened survival, we now have the opportunity to see patients with less typical symptoms during their lifetime than were available in the past. Even if a given symptom is rare, such as cognitive dysfunction, sensory disturbance, or problems with eye movements, there are sufficient numbers of these under-recognized symptoms that the chances are fair that a patient will experience one or more of these during the course of their disease. The pioneers that first described ALS did not have the advantage of large samples, prospective registries, validated measures, or suitable comparison groups. Perhaps in this context, we should re-examine the evidence we rely upon to inform our opinions about ALS;

were older studies that supported traditional notions somehow tainted by confirmation bias? Pain is an important exemplar that suggests we can take this opportunity to go back to patients themselves and listen intently and without prejudice to their experience of illness to guide how we support them in maximizing their outcomes.

P. Wicks

PatientsLikeMe, Research & Development, 155 2nd Street, Cambridge, MA 02141, USA (e-mail: pwicks@patientslikeme.com)

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