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The National Institute of Neurological Disorders and Stroke has as its mission reducing the burden of illness due to stroke and hundreds of neurological disorders. Though stroke, epilepsy, headache are extremely common, we are the lead institute for 100's of rare neurological conditions. Many if not most of these conditions cause major disability in all or a segment of the affected population. Our efforts are focused on facilitating the discovery of new knowledge and new treatments for these extremely debilitating, and often fatal conditions. Some cause impairments that are obvious, such as paralysis, others are disabling in less obvious manners such as those that affect cognition, behaviour or cause pain. As compared to previous ages, in today's technologic age one's ability to function in the workplace seems dependent less and less on physical abilities, and more upon cognitive abilities. The IOM report "Improving the Social Security Disability Decision Process" reported that the percentage of SSI adults age 18-64 with mental disorders was 57% and mental disability has seen the largest growth in recent years. Because there are so many rare neurologic disorders we are acutely aware of the difficulty of being expert enough to be able to accurately make disability determinations in all. Because of the complexity of understanding all of these disorders clinical neuroscience itself has become more and more specialized.

As a neurologist with a busy practice and overseeing the practices of 30-40 residents prior to coming to NIH, I am cognizant of the troubles that certain patients have had in obtaining legitimate disability. Unfortunately, I can also testify that it was not infrequent to come across a case in which a person, perfectly able to make contributions in the workplace, would attempt to convert a "diagnosis" of neurological illness into a disability determination. Sometimes this seemed to be motivated by fear for their future health, sometimes simply a desire to establish a steady income. The SSA definition that the person be unable "to engage in any substantial gainful . . . work which exists in the national economy", as opposed to their previous job, is also very difficult for many patients to accept. Physicians therefore are often be pressured to "fill out disability forms" for patients who do not fit the definition in the Social Security Act. In some such cases physicians may object and indicate to the patient that remaining employed is in the best interest of their health. On the other hand the physician is also acutely sensitive to the importance of the patient-doctor relationship. They will often fill out the form truthfully in these cases, but hope that the SSA reviewer will see the facts clearly and make the correct decision. Worse, there are some patients with neurological symptoms who challenge the best diagnosticians to determine whether their symptoms are contrived or due to disease. SSA clearly has a tough job being sure that they don't deny benefits to those who are truly disabled, and don't grant benefits to those who are not really disabled. In many cases there is no gold standard.

Troubling as well is the patient unable to work due to damage to brain, spinal cord, nerve or muscle who struggles to obtain disability determination. In asking for input from patients the main issue seems to revolve around variability and consistency.

I received a number of replies about fronto-temporal dementia (FTD). It is a degenerative disorder that is not so well known. Its affects on behaviour and cognition can be insidious and there is considerable variability in its presentation. FTD therefore is a good example of a difficult disorder for SSA. However it is unfortunately progressive and leads to complete disability.

Let me share one very positive reply I received about a patient's interaction with the SSA system:

“My experience with helping my mother apply for Social Security Benefits was not as hard as I thought it would be. We went to the office together with a summary of the testing from a neuropsychologist. At the time the doctor had not used the exact phrase: frontal temporal dementia, but instead was describing brain atrophy of the frontal lobes, dementia and aphasia. The man at the social security office was very understanding, and it was obvious to him, or anyone, that my mother was impaired. There was also the fact that she had been fired from her job of 7 years, then got another job, but was fired two weeks later for not being able to perform the tasks. Our meeting was in December and her benefits started in March. I thought it would be harder.”

This may not be representative of all patients with FTD. Dr. Kent Jamison, vice chair of the Association for Frontotemporal dementia wrote me to say the “ambiguities” inherent to this disorder may make it difficult sometime. Patients can become cognitively impaired at a younger age. Another responder wrote that “when I called SSA I was asked if my husband was over 65, when I said no, the reply was “why not?”. FTD affects the patient’s frontal lobe executive function which may be necessary to persist in the pursuit of disability determination process. Dr. Jamison wrote that many do “not recognize their own deficiencies” and “it is little wonder then that final approval may get tripped up at the local or regional level causing delays and further hardship for caregivers”.

When I started caring for patients with Huntington’s disease in Boston in the mid 1980’s there was a constant battle to obtain disability for patients who were terribly disabled but walking, strong, not obviously aphasic. Their disability was due to cognitive and behavioural changes that sometimes even fooled their family. Something changed for the positive however by the mid 1990’s. I distinctly recall one day thinking how much smoother the process had become. The reason was never clear: Huntington’s disease was better known?; after filling out so many forms the State DDS staff came to know me?; HD was added to the list of degenerative disease?. I couldn’t tell but it did improve. The tough issue is how to systematize the knowledge base needed to make disability decisions in all the varied rare disorders. Clearly the rare conditions present the most problems because an individual staff person’s exposure to the wide array of presentations is surely to be limited.

This SSA administration has been extremely proactive in trying to problem solve. This summer representatives from multiple NIH institutes met to discuss how NIH research might help inform SSA with regard to how new scientific knowledge can impact on the disability determination process. A number of the NIH institute representatives were enthusiastic about participating in the process to uncover whether new technologies—neuroimaging, neurogenetics, or specialized testing might be best utilized to determine when a person is no longer able to work. Others were interested in the idea of incorporating commonly used functional or medical severity scales which have been well validated in clinical research arena, and could be correlated with inability to work. In so doing NIH would extract a greater knowledge of the burden of illness and functionally relevant endpoints for new treatment trials. All realized the difficulty inherent in making 100% accurate disability determinations on the first attempt in all 2.6 million applicants, but were attracted by the possibility of at least being able to make sure that the severely disabled patients could be identified out. This would directly help a significant number of affected individuals, improve processing time, but also indirectly help if the SSA resources could be better concentrated. NIH is very interested in working with SSA to develop the knowledge base for injecting the most current and valid mechanisms of determining the severity of illness into the disability determination process. This should be a work that is always in process as new techniques and information comes from research.

Thank you very much for your attention.

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