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BOOK REVIEW

Medical Mysteries and Conundrums —When Doctors May Not Help

Understanding Hashimoto's Encephalopathy: A Guide for Patients, Families, and Caregivers by Nicola Nelson, Susan Foley, and Shari Lawler for the Hashimoto's Encephalopathy SREAT Alliance. CreateSpace, 2013. 458 pp. $21.95 (paperback). ISBN 978-1484883099.

Some number of people feel ill, yet mainstream medicine tells them via their doctors that there is no discernable reason for their symptoms and therefore no clear way to offer any real help. Sometimes the sufferers may be told that it's psychological stress, "all in the head," that there's nothing "really" wrong even though the physical pains are indubitably felt; or perhaps that what they think they suffer from doesn't even exist.

Consider Chronic Lyme Disease, for example:

> The term 'chronic Lyme disease' is not recognized in the medical literature, and most medical authorities advise against long-term antibiotic treatment for 'chronic Lyme disease'. Studies have shown that most patients diagnosed with 'chronic Lyme disease' either have no objective evidence of previous or current infection with *B. burgdorferi* or are patients that should be classified as having post-Lyme disease syndrome, which is defined as continuing or relapsing non-specific symptoms (such as fatigue, musculoskeletal pain, and cognitive complaints) in a patient previously treated for Lyme disease. (Wikipedia; Science-Based Medicine)

Or chronic fatigue syndrome (CFS):

> Chronic fatigue syndrome is a complicated disorder characterized by extreme fatigue that can't be explained by any underlying medical condition. The fatigue may worsen with physical or mental activity, but doesn't improve with rest. The cause of chronic fatigue syndrome is unknown, although there are many theories—ranging from viral infections to psychological stress. Some experts believe chronic fatigue syndrome might be triggered by a combination of factors. There's no single test to confirm a diagnosis of chronic fatigue syndrome. You may have to undergo a variety of medical tests to rule out other health problems that have similar symptoms. Treatment for chronic fatigue syndrome focuses on symptom relief. (Mayo Clinic)

> Researchers have not yet identified what causes CFS, and there are no tests to diagnose CFS. Moreover, because many illnesses have fatigue as a symptom, doctors need to take care to rule out other conditions, which may be treatable. (Centers for Disease Control and Prevention)

> CFS may also be referred to as myalgic encephalomyelitis (ME), post-viral fatigue syndrome (PVFS), chronic fatigue immune dysfunction syndrome (CFIDS), or by several other terms. Biological, genetic, infectious, and psychological mechanisms have been proposed, but the etiology of CFS is not understood and may have multiple causes. (Wikipedia)

> CFS may in some cases be associated with immune deficiencies similar to those in AIDS. (Blogspot)

Or irritable bowel syndrome:

> In many cases, you can control irritable bowel syndrome by managing your diet, lifestyle, and stress. (Mayo Clinic)

> There are treatments that attempt to relieve symptoms, including dietary adjustments, medication and psychological interventions. Patient education and a good doctor–patient relationship are also important. (Wikipedia)

Those suffering from such symptoms or syndromes are unlikely to find any of these official pronouncements particularly helpful. They would be well advised to do some research for themselves. Sometimes, it turns out, they can come to understand what's wrong and even fix it, as Mohammed Aziz did when a family member turned out to have a deficiency of vitamin D (Aziz 2012, Bauer 2012a). With the officially non-existent Chronic Lyme, some sufferers have benefited from long-term antibiotic treatment offered by a few intrepid, empirical, evidence-based physicians who risk being disciplined by their profession and by government authorities (Bauer 2010).

One of the children in our family became annoyingly unruly, strangely obsessive, a "problem child" in school. A succession of doctors advised counseling or medication with the ADHD favorite Ritalin or its ilk. The parents thought it might have had something to do with a recent infection, but the medicos pooh-poohed the idea. The persistent parents did their own research, and it turns out that their daughter suffered from PANDAS:

> PANDAS is an acronym for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections, a rare disease that usually appears in children. This term describes a hypothesis that there exists a subset of children with rapid onset of obsessive-compulsive disorder (OCD) and/or tic disorders and these symptoms are caused by group A beta-hemolytic

> streptococcal (GABHS) infections. The proposed link between infection and these disorders is that an initial autoimmune reaction to a GABHS infection produces antibodies that continues to interfere with basal ganglia function, causing symptom exacerbations.
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> The PANDAS hypothesis was based on observations in clinical case studies at the U.S. National Institutes of Health and in subsequent clinical trials where children appeared to have dramatic and sudden OCD exacerbations and tic disorders following infections. There is supportive evidence for the link between streptococcus infection and onset in some cases of OCD and tics, but proof of causality has remained elusive. The PANDAS hypothesis is controversial; whether it is a distinct entity differing from other cases of Tourette syndrome (TS)/OCD is debated.
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> PANDAS has not been validated as a disease entity; it is not listed as a diagnosis by the International Statistical Classification of Diseases and Related Health Problems (ICD) or the Diagnostic and Statistical Manual of Mental Disorders (DSM). Pediatric acute-onset neuropsychiatric syndrome (PANS) is a 2012 proposal describing another subset of acute-onset OCD cases including "not only disorders potentially associated with a preceding infection, but also acute-onset neuropsychiatric disorders without an apparent environmental precipitant or immune dysfunction."(Wikipedia)

Even if personal research turns up no real "scientific" certainty or actual cure, suffering individuals may get considerable help by learning of other people in similar situations and how they cope. And occasionally it may be that such contacts suggest the possibility of a known ailment so rare that it escaped the attention not only of local doctors but even that of such renowned institutions as the Mayo Clinic. That happened to someone who eventually discovered her problem to be Hashimoto's Encephalopathy:

> Hashimoto's Encephalopathy ('HE'), also commonly referred to as Steroid Responsive Encephalopathy Associated with Thyroiditis ('SREAT'), is a rare and devastating autoimmune disease in which a patient's antibodies mistakenly turn on and attack the patient's brain. The results of the attack may include severe cognitive impairment, speech disorders, seizures, memory loss, impaired balance, movement disorders, and sometimes psychosis. Patients sometimes fall into a coma and in rare cases die. Unfortunately, HE is not well understood and patients often face an enormous struggle trying to find the correct diagnosis for their confusing constellation of symptoms. Many neurologists have never even heard of the condition, although that has begun to change in the last few years. Misdiagnosis is the rule more often than the exception, at least in the early months of illness.

Those words introduce the book under review. Part I describes Hashimoto's Encephalopathy. Part II contains 35 personal testimonies from

individuals diagnosed with HE, at ages as young as 17 and as old as 66, in Australia, Denmark, England, Scotland, and the United States. Part III has 5 stories of children diagnosed as young as 10, as old as 17. In Part IV, friends and family of HE sufferers share their experiences. Then there are Resources: support groups (pp. 454–456), informational websites (pp. 457–458), and scientific articles (pp. 403–453).

Obviously this book is invaluable for anyone with HE, but most particularly for those who don't yet know that they have HE. It deserves to be common knowledge that a variety of debilitating symptoms might (but in rare cases only!) be signs of HE, symptoms easily mistaken for "mental" problems and difficult to diagnose:

> patients typically present with several of the following symptoms: cognitive impairment (including concentration and memory problems); emotional, behavioral, and personality changes; speech difficulties (including aphasia or dysphasia); tremor; muscle jerking (myoclonus); dizziness, vertigo, and impaired coordination and balance (ataxia); headaches; malaise, weakness, and fatigue; fluctuating consciousness; disorientation, confusion, or dementia; seizures or seizure-like events; stroke-like episodes; partial paralysis (often on the right side); sleep abnormalities such as insomnia or excessive sleepiness; sensory or motor difficulties, often on one side; status epilepticus; psychosis (including hallucinations, delusions or paranoia); coma.

Not necessarily helpful, since any of those can occur for a variety of reasons. Even worse: Some people experience episodes, in other words remissions and relapses, whereas others suffer progressive worsening. Consequently, HE is diagnosable only by excluding a whole host of other possible conditions, some of them also quite rare.

One of the first symptoms seems often to be like a stroke or a seizure. Brain MRIs sometimes appear like those of much older individuals (e.g., p. 280). The book gives details about a number of tests and what they can and cannot prove. The report of a consulting neurologist, reproduced in full at pp. 269–775, offers insight into the difficulty of diagnosing and treating HE. It took 7 years after her first bad episode before Kelly got her diagnosis of HE (pp. 276–277).

Many will find it reassuring that "up to 90% of HE patients respond to steroid treatment" (p. 19). When seizures occur, anti-seizure drugs are appropriate. Thyroid hormone replacement may help when there is pronounced hypothyroidism.

The personal reports make emotionally difficult, heart-rending, shocking reading—extraordinarily debilitating experiences of mental confusion, physical pain, feeling completely ***lost***, described sometimes as

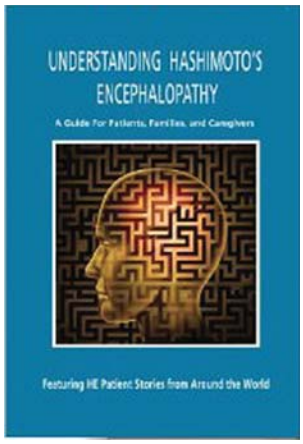
having an out-of-body experience, or feeling abducted by aliens, or suddenly experiencing full-blown dementia, or being on the point of dying—or even beyond it. Unable to talk, or unable to think, having no motivation to do anything at all, not knowing how to drive a car. Shari gives a very detailed description at pp. 82–97.

There is, however, a silver lining: These people somehow managed to carry on and eventually to understand what the problem is and often to go further with such ventures as this book and using social media to bring positive help to other people. There are tales here of astonishing courage and determination, for instance from Allison, who did much research, essentially developed her own treatment as a full partner with her doctors, survived a husband leaving her while she was ill, and not only held HE at bay but managed at the same time to have the baby she had always wished for (pp. 174–183).

Each tale is unique, but there are such common elements as long periods of tests, mis-diagnoses, innumerable doctors, and specialists. Often the doctors concentrate on what "objective" tests show while paying little attention to what the patients know about what they are feeling. One possible reason or excuse may be that patients often present with such a variety of complaints that hypochondria is a ready guess. In one case, Canadian doctors would not accept an HE diagnosis from the Mayo Clinic (pp. 363–364). Shari offers useful advice at pp. 99–116.

The adage that "hard cases make bad laws" cannot be applied to medicine: Systems should be designed with the needs of ***individuals*** in mind, just as democracies should care about minorities and avoid the tyranny of the majority. This book illustrates that the American "Health Care System" is worse than inadequate in several ways, routinely because there is little if any coordination among specialists and family doctors (see pp. 78–79, for instance), and disgracefully in individual cases when, for example, an insurance company refuses to pay for a diagnostic visit to the Mayo Clinic after local doctors have exhausted their expertise (p. 46); or when attending doctors could not get approval to do a PET scan (p. 60) when they were desperately seeking to diagnose a patient who had them puzzled; or having to wait more than a month to be seen by an appropriate specialist (p. 74), or being frustrated by bureaucratic paperwork (p. 77). In one instance,

> now that she is covered by Medicare it has opened doors to treatment options she didn't have with private insurance (that she paid for/worked for all of her life)! (p. 150)



—illustrating what's wrong with free-market, profit-driven healthcare and private insurance. There are the tens of millions like Tiffany (age 28; p. 256), who have no health insurance because they cannot afford it. For more such horror stories, read what Grace (p. 331 ff.) reports about insurance, bureaucracy, and not being allowed to defer payment on student loans. Or about a wait of 2 years for an appointment with a pediatric neurologist (p. 362).

That "up to 90% of HE patients respond to steroid treatment" is not unmitigated good news because the "side" effects can be dangerous in themselves, and some of the personal tales recount an astonishing number and variety of medications used to mitigate the consequences of the primary treatment, which is basically immune suppression. Allison, for example, was taking 21 drugs at one stage (p. 175).

The thing to remember always about "side" effects is what Frank Ofner, MD, used to say: "'Side' effects are ***main*** effects that doctors don't want to talk about." Everything a drug does is an effect. That "side" effects are anything but negligible or ignorable is illustrated as drugs approved for one particular use are then ***often*** claimed to be effective in entirely different ailments. Drug companies like to "reposition" their drugs in this manner because it is cheaper and easier than having an entirely new drug approved: Once a drug has been approved earlier as "safe," fewer clinical trials are needed when approval is sought for the different use (Ashburn & Thor 2004, Healthtech 2012, Bauer 2012b). Of course, that a drug "works" does not necessarily mean that it is treating the actual cause of an ailment. As I read somewhere, the fact that aspirin "cures" pain doesn't mean that the pain was owing to lack of aspirin. Larry (p. 189) found that an anti-inflammatory and an anti-psychotic worked the same benefit for him in treating his HE, and asks the excellent question what that means.

Individual doctors are permitted to prescribe *any* approved drug for *any* condition, though drug manufacturers are not allowed to proselytize for such "off-label" uses. Well-informed patients may be able to persuade their doctor to try treatments for which only anecdotal evidence exists. A couple of the stories in this book mention naltrexone or low-dose naltrexone, officially approved to treat alcoholism but attested by a number of people as "helping those with HIV/AIDS, cancer, autoimmune diseases, and central nervous system disorders" (LDN).

The personal experiences published here are surely invaluable for medical professionals as well as for individuals suffering from mysteriously erratic, unpredictable, capricious collections of symptoms.

> [S]ince everyone who has HE has different symptoms, it's sometimes easy to dismiss some of my symptoms. I did that while I was in a period of denial. Now, I realize that the only way I can get better and other people can get help is if we all keep track of our symptoms and share them so that we can learn from each other and doctors can learn. (p. 163)

A priori assumptions don't work. For example, HE is an autoimmune disease; there is a genetic predisposition to some autoimmune ailments; autoimmune diseases run prominently in Lisa F.'s family; yet HE hit her only at age 40, while individuals with no such family history have been affected as early as age 10. Some early symptoms are reminiscent of paranoia or schizophrenia (see Larry at p. 183, for instance). The difficulty of ascribing characteristics to HE are redoubled because treatment always begins with, and has sometimes even preceded, diagnosis, so that some symptoms might be drug "side" effects rather than indicative of HE. Or, they may have to do with neither: Larry (pp. 196–197) twice had a toe turn blue and then black underneath the nail, then the nail dropped off and was replaced with a new one, and thought this might have been related to episodes of HE—but I had such an experience while perfectly healthy and attributed it to too tight a shoe.

HE seems more common among women than among men, but the reported ratio varies between 2-to-1 and 6-to-1 (pp. 9–10). Only 7 of the 35 personal stories in this book are from men, and only one of those was younger than 45 at diagnosis of HE. All 5 children's stories are about girls.

Before diagnosis of HE had been accomplished, some older HE patients were sometimes told that changes they had noticed were just age-related: not finding the right words, difficulty concentrating, decreased motivation, poor balance, general anxiety, loss of short-term memory while long-term memory remains. Those are indeed things that come with age, of course at quite different ages and with different intensities in individual cases; I (age 82) could empathize—fortunately to only minor degrees—with about half of the first 20 symptoms of HE listed by Larry at pp. 185–187. Essentially all the personal stories report that life is now quite different than before HE, which can also resonate with much older people who have seen their capacities decline and who spend almost all their time just coping with everyday matters. Anyone who has experienced a family member with Alzheimer's or other age-related dementia will appreciate how similar can be the symptoms of HE; those who have not had this experience can read

about the typical symptoms in the story of James (pp. 165–174).

This is a very important book. Mention it to everyone you know. The fact of very rare and difficult-to-diagnose diseases ought to be known to everyone, and that a large proportion of doctors and specialists might be ignorant about these conditions. Awareness of those facts might assist the few actual victims get help earlier than did so many of the individuals who tell all in this book.

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