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PERSONAL HEALTH

Shedding Light On a Tremor Disorder

By JANE E. BRODY

“Essential” usually means vital, necessary, indispensable. But in medicine, the word can assume a different cast, meaning inherent or intrinsic, not symptomatic of anything else, lacking a known cause.

Since the mid-19th century, “essential tremor” has been the diagnosis for a disorder of uncontrollable shaking — usually of the hands but sometimes of the head and other body parts, or the voice — that is not due to some other condition. And without knowing what causes it, doctors have been slow to come up with treatments to subdue it.

As a result, millions of individuals suffer to varying degrees with embarrassment and humiliation, social isolation and difficulties holding down a job or performing the tasks of daily life. When you cannot drink a glass of water or eat soup without spilling it because your hand shakes violently, you are unlikely to join others for a dinner out. When you have to depend on someone else to button your shirt or zip your jacket, you may not go out at all.

Wherever those with essential tremor go, people are likely to stare at them and assume they have a drug or alcohol problem, said Catherine Rice, executive director of the International Essential Tremor Foundation in Lenexa, Kan. (Call it at 888-387-3667 or visit its Web site: www.essentialtremor.org.)

Now, thanks to the devoted efforts of a few researchers here and abroad, all this may change. Recent studies have begun to unravel the mysteries of essential tremor, and “essential” may someday be dropped from its name.

“Until very recently,” Dr. Elan D. Louis, a pioneering neurologist and epidemiologist at the College of Physicians and Surgeons at Columbia University, told me, “essential tremor was thought to have no known pathology, no changes in the brain, which led to a medical dead



RYAN McVAY/GETTY IMAGES

end.” But in the last five years, Dr. Louis said, discoveries in three areas — the brain, clinical findings and genetics and environment — “have changed our understanding of this disease.”

And as our understanding evolves, he predicts that rational therapies will follow.

Common Over Age 65

Essential tremor is a neurological disorder that causes uncontrollable shaking of one or more body parts during voluntary movement. The symptoms disappear at rest. In that way it differs from Parkinson’s disease, in which shaking at rest is a common symptom that disappears during movement. But those with essential tremor are four to five times as likely to develop Parkinson’s as people without tremor, and both conditions involve related changes in the brain.

Though essential tremor most often affects older people — as many as 1 in 5 over 65 have it — it can occur at any age, even in young children. It is typically progressive, getting worse as people age.

Stephen Remillard of Steamboat Springs, Colo., said he learned he had essential tremor while in kindergarten, when it affected just his hands. But the condition worsened as he got older, and by high school, Mr. Remillard said, “all my extremities as well as my voice were affected.” When he had to speak in class, he said, “it came off as if I was nervous, though I’ve always been a very confident person.”

The academic challenges related to tremor prompted him to drop out of college. But the biggest blow to Mr. Remillard’s self-esteem came when he tried to join the military and was rejected by the Army, Marines, Air Force and Coast Guard. Rather than feel sorry for himself, he returned to college, graduating last May, and started playing sports. Now 25, he works for a ski corporation and runs marathons to raise money for causes like the Lance Armstrong Foundation.

For Richard Crandell, a 66-year-old guitarist from Eugene, Ore., the problem began around age 60, forcing him to abandon his instrument. But he, too, was not to be defeated: he took up the mbira, an African thumb piano that he plays with two thumbs and an index finger.

Still, Mr. Crandell said, he has problems shaving, brushing his teeth, using a computer and slicing and dicing in the kitchen. And at the bank, he has to ask the teller to fill in his forms “because my handwriting is all over the place.”

Ms. Rice said essential tremor ran in her family. “My great-aunts used to shake uncontrollably, starting in their early 40s and becoming quite severe by the time they were 60,” she said. “They found it very difficult to cook, though their job was to feed the farmhands. They couldn’t pick up a heavy pan without spilling the contents. They had to give up crocheting and other things they truly loved.”

New Findings

Dr. Louis and colleagues have established a centralized brain repository that has revealed underlying abnormalities in essential tremor patients. The scientists collect detailed clinical and physiological data on each person, and after death their brains are shipped to Columbia, where they are analyzed and compared with the brains of normal individuals.

Of the 50 brains studied so far, Dr. Louis said, "all are degenerative and have very clear pathological changes, although there are several types, suggesting this is probably a family of diseases." In one subtype, Lewy bodies, which also occur in Parkinson's disease, are found in the brain but in a different area from Parkinson's. (Mr. Crandell's father died of Parkinson's, and there have been suggestions that the disorders may be linked.)

In about 80 percent of the brains, there are degenerative changes in the cerebellum, including a loss of cells that produce a major inhibitory neurotransmitter called GABA. Other abnormal findings include a messy arrangement of neurofilaments, which may interfere with nerve cell transmission.

Clinically, essential tremor is now considered a neuropsychiatric disease that can include unsteadiness, abnormal eye movements, problems with coordination and cognitive changes that sometimes progress to dementia.

Even certain personality types tend to be overrepresented among patients with essential tremor, Dr. Louis said. Many "are very detail-oriented and tightly wound and have higher harm-avoidance scores," he said.

Two environmental toxins have been found to be elevated in tremor patients:

lead and a dietary chemical called harmaline that occurs naturally in plants and animals. When meat is cooked for long periods or at high temperatures, as in barbecuing, levels of harmaline rise sharply. Dr. Louis called these "tantalizing leads."

Despite the problems caused by their disorder, most patients with essential tremor never seek treatment. Two drugs, propranolol (Inderal) and primidone (Mysoline), developed to treat other conditions, have proved helpful for many but not all patients. A costly surgical treatment, deep brain stimulation, has helped to reduce tremors in about 80 percent of patients who have tried it.

Caffeine, certain prescription drugs and undue stress can make symptoms worse and are best avoided. Though alcohol can temporarily relieve tremors, regular heavy drinking is a recognized cause of the disorder.