Patient Handouts

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Essential Tremor

Introduction

Tremulousness is a very common symptom. Indeed everyone has a degree of tremulousness. On holding the arms straight out in front of us, we can observe a fine and barely noticeable trembling of our fingers, known as “physiological tremor” (physiological meaning related to normal function). Many normal people have noted that when under stress they get “the shakes.” This is usually due to accentuation of their physiological tremor. In addition to stress or anxiety, certain medications, such as asthma inhalers, also may exacerbate physiological tremor.

However, when tremor becomes noticeable in daily life, people often assume they have Parkinson disease. Although such persistent tremor is no longer physiological, it is much more likely to be essential tremor, a tremor with a much better prognosis than that of Parkinson disease. If there is a family history of tremor, essential tremor becomes even more likely. Other names that are used for essential tremor are benign essential tremor, familial tremor, or senile tremor (if onset is in old age).

Symptoms of essential tremor

The tremor is barely noticeable for a long time and typically progresses very slowly, usually over years or decades, and the symptom is often blamed on “bad nerves.” It is accentuated when maintaining certain positions or postures of a limb, such as when using utensils or carrying a cup, and during movement, particularly when precise movements and fine motor control are required, such as when reaching for an object or picking up a full glass. A common complaint is "Doctor, I can't carry a cup of coffee from the kitchen to the sitting room without spilling it!" Other complaints are that writing, threading a needle, or using a screwdriver are difficult. Although the hands are typically affected in a symmetrical manner, the feet, head, jaw, voice, or mouth can be affected as well. Since the tremor is worse when carrying out routine activities, people become aware of it before others begin to notice. As with most tremors, intense concentration, stress, and anxiety exacerbate essential tremor. A small quantity of alcohol may help relieve the tremor for an hour or two.

Why isn't it Parkinson disease?

This is a burning question and one that is uppermost in the mind of many. Hence, it is important that we explain some differences. People with Parkinson disease notice a tremor at rest, in other words, when they are not using the arms or hands in contrast to the tremor with use described in the previous section. This is the most specific difference that a neurologist looks for. There are often additional symptoms in Parkinson disease, such as a general slowing down, often both physically and mentally, and a general sense of stiffness, whereas essential tremor has none of these symptoms.

The vast majority of people (95%) with essential tremor notice the shakes in their hands, usually in both hands, but it obviously causes more trouble in the dominant hand (the hand that one uses to write, to throw a ball, etc.). If the tremulousness is restricted to only one hand or arm, then it is less likely to be essential tremor. Other parts of the body may also shake including the head (33%) in a characteristic bobbing fashion either up and down or side to side. This is called “titubation” and it is almost never

found in Parkinson disease. Rarely the voice (12%) may become tremulous and lastly the legs may shake, especially when standing still; this has been called an “orthostatic tremor” even though it is considered a variant of essential tremor in the broad sense.

Another useful distinction between Parkinson disease and essential tremor is the family history. Essential tremor is much more likely to run in a family, and more than 50% of the time patients will recall that their mother or father used to shake. Parkinson disease is very rarely familial.

There is one other interesting characteristic of essential tremor that differentiates it from the rest tremor of Parkinson disease. An essential tremor may often get better with alcohol whereas the tremor of Parkinson disease will not; so this is a unique situation in which your doctor may recommend a small glass of alcohol before situations of stress or when eating out socially, to settle the tremor down.

Epidemiology- Who gets essential tremor?

Many scientific studies have been done in many countries involving people of all ages from different racial and ethnic backgrounds. We know that the older you are the more likely you are to develop an essential tremor. Indeed, so common is this in people over about 70 years of age that it used to be called "senile tremor," but at present scientists believe that there is no point in differentiating essential tremor based on age. We know it is more common than idiopathic Parkinson disease. The prevalence in the community has been found to be between 2% to 20% depending upon which scientific study you read and on the country in which it was undertaken. We do know that although it is more prevalent in those older than 40 years, there are occasional adolescents who develop what appears to be isolated tremulousness. Often they are more aware of it when writing, something that they are perhaps doing frequently at college or school. Often the tremor with writing is associated with a rather painful, spasm-like feeling in the hand and fingers and the writer will consequently adopt unusual ways of holding a pen. This rather painful abnormal posturing of the hands, or fingers, or both, is called dystonia, and often tremor and dystonia go together. Fortunately the essential tremor in young adult life usually dissipates after a few years.

Many patients are concerned that essential tremor may progress and “turn into Parkinson disease”—an understandable concern. Epidemiological studies have given conflicting evidence—some suggest that the two conditions are related, whilst others have shown no relationship. There is, however, as mentioned before, a strong hereditary influence in essential tremor; between 20% to 75% of people with essential tremor will admit to a family history of tremulousness. The hereditary influence is so strong that it is passed onto the next generation with a risk of about 50%, consistent with a pattern of inheritance that is known as autosomal dominant. Such a pattern is rarely seen in idiopathic Parkinson disease. It is hardly more likely that someone with essential tremor will develop Parkinson disease than will someone without essential tremor.

Cause

The cause of essential tremor is not known. Your doctor will ask you about your family history, and importantly, about the medications you may be taking for other conditions. There are specific examples of very powerful drugs, especially those used to treat severe depression or other psychiatric illness, that over time may cause a person to look as though they have Parkinson disease. Stimulants, even caffeine in coffee, may make people shake, although this is usually due to an exaggerated physiological (normal) tremor. Once all the recognized drugs and toxins and other diseases that are associated with tremor are excluded, then we are left with this very common isolated tendency to shakiness of the hands, and because it is of unknown cause and the tremor occurs by itself it is described as "essential" tremor. If there is a strong family history then it is often called “essential familial tremor.” Considering how often

members of the same family are affected the abnormality is likely at the genetic level.

The current scientific belief is that the problem arises in those parts of the brain not so much concerned with the strength or power of muscles but rather in those which control the subtle and delicate way in which we move the muscles in a smooth and coordinated way and in a certain sequence. These particular parts of the brain are called the cerebellum and the basal ganglia, along with the nerves, which connect these areas with each other.

Quality-of-life issues

Even though we used to call this essential tremor "benign," it may be quite disabling to a small but significant number of patients. It may be socially embarrassing, often preventing people from going out to restaurants or even visiting their families. It may even ultimately make handwriting illegible. Reading the newspaper or holding the telephone up to the ear may be impossible. This, however, is a worst case scenario, and for most it is simply an inconvenience and primarily a social issue.

Treatment

It is important to admit at the outset, that because we don't know the cause, there is as yet no cure. The first step in management is probably the simplest and the best—to reassure the patient that he or she doesn't have idiopathic Parkinson disease. Education and explanation, such as the reading of this hand out, go a long way in reassuring patients that the problem is not sinister and indeed often influence patients to choose not to have any medication. Most patients do not need any treatment or can get by with a modest amount of alcohol before social events.

However, if disabling, either psychologically or physically, then treatment options at present rely on reducing the tremor, which is referred to as "symptomatic treatment." Almost all of the medications used for essential tremor were not specifically developed to treat tremor, but rather other medical conditions and it was then noted by some patients who also had an unrelated tremor that the drug seemed to reduce the shaking. The most common drugs are beta-blockers, which seem to block the autonomic nervous system and calm down the tremor. How this class of drugs work is not well understood, but beta-blockers provide symptomatic relief in about half of essential tremor patients, and because they are relatively innocuous drugs they are frequently prescribed.

Primidone is another widely used drug. It can cause sedation, but it is of proven benefit.

More recently, some patients were tried on anticonvulsants (the class of drugs used primarily for epilepsy), and there was a tendency to reduce tremor. Such drugs have now been subjected to controlled trials, which proved some benefit; these drugs include topiramate, gabapentin, and lamotrigine. In severe cases of tremor, if surgery is not an option, then combining two medications such as a beta-blocker and an anticonvulsant may be tried, but as always, the side effects are often magnified and without much dramatic benefit.

Alcohol may even be "prescribed" for essential tremor, and usually the advice is to use this judiciously, such as half an hour before attending an important function.

Even with optimum use of drugs, about 50% of patients continue to be disabled by their tremor, hence, other forms of treatment have been developed, including the use of botulinum toxin and surgery.

Botulinum toxin

There has been much research recently in the use of botulinum toxin, which quite simply partially

paralyzes the muscles for weeks to months. Whereas the drugs mentioned above are presumed to work in the brain, botulinum toxin weakens the muscles that shake so there is less tremor. Use of botulinum toxin is not without side effects, especially if not injected into the right muscles and in the correct amounts, although thankfully, any unexpected or excessive weakness eventually wears off. The injections must therefore be administered by a specialist who has an intricate knowledge of anatomy and the actions of the various muscles in the arm, hand, or neck.

Surgery
The very first operation for tremor in a human being took place in 1947. Surgeons found that if they created a small scar in the basal ganglia (that part of the brain that is responsible for the smooth and orderly control of motion that is abnormal in idiopathic Parkinson disease and essential tremor) the tremor would often almost miraculously disappear. However, the technique fell into disrepute because of unacceptable side effects. The scar had to be made so carefully and with such precision that any error could cause a stroke or impair language. In the past 10 years or so, using CT and MRI scans with three-dimensional images of the brain, neurosurgeons are able to much more accurately visualize the brain and to place the very thin wire into the exact part of the basal ganglia, producing a scar only a few millimeters in size.

In the 1980s neurosurgeons discovered that rather than making a scar in the basal ganglia (which is irreversible--an obvious disadvantage if the scar is not quite in the correct place), stimulating the thalamus of the basal ganglia using a high-frequency pulse wave could stop the tremor. This technique is called deep brain stimulation. It may be somewhat inconvenient for the patient, as they have to carry around a small electrical device (exactly the same principle as a heart pacemaker) and there is a wire left in place through a minute hole in the skull. The wire and pulse generator are inserted under the skin and the battery can be turned off and on at will by a small hand-held magnet. The advantages are obvious and the procedure is reversible. If the tremor reappears the stimulation may be increased in strength or duration. The most common side effect is slurred speech (dysarthria), which can be minimized by reducing the pulse stimulation. Because there is a piece of wire and a small metal box left in the body, there is an increased risk of local infection or inflammation. Another disadvantage is that the battery eventually needs to be replaced, which requires a small surgical excision.

Because the tremor is not present when asleep, patients must be awake during the operation. Despite the sophisticated technology involved in placing the wire, the best indicator that the wire is in the right place is the disappearance of the tremor when the wire electrode is stimulated. Once this happens, the surgeon is more confident that the operation will be successful. Naturally, the surgeon uses a powerful local anesthetic on the scalp so that there is no pain. Interestingly, the brain tissue itself does not feel pain.

Not every patient necessarily requires surgery and it is a decision made after much consultation, usually with an expert panel of neurologists, neurosurgeons, and psychologists. If the panel does agree that stereotactic surgery is the best option, then the next step is to decide whether to have a thalamotomy (scar) or thalamic stimulation. Again, there are many factors to consider when weighing up the advantages and risks of each procedure, and the patient and patient’s family would be involved in the ultimate decision.

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