

Essential Tremor: What the Experts Say

Collected Articles and Guidance

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International Essential
Tremor Foundation

Your Voice for Essential Tremor

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Preface

The International Essential Tremor Foundation (IETF) is pleased to publish this collection of informative articles about essential tremor (ET) written by some of the world's finest movement disorders specialists and experts in related health care fields. Now in its third edition, this comprehensive book is an invaluable resource to assist with understanding essential tremor. We are confident that this collection will help to enrich your life by offering you educational material that can be referenced again and again.

The IETF thanks the many physicians, health care providers, and other individuals that contributed time and effort to make these articles possible. These people have contributed greatly to the success of the IETF over the years.

About The International Essential Tremor Foundation (IETF)

Founded in 1988 as a 501(c) (3) non-profit organization, the IETF is guided by an executive board of directors, a medical advisory board and an executive director with a staff of three. The organization's membership consists of patients, physicians, educators, health care workers, parents, relatives, friends and volunteers.

Our Mission

The IETF funds research to find the cause of essential tremor (ET) that leads to treatments and a cure, increases awareness, and provides educational materials, tools, and support for healthcare providers, the public, and those affected by ET.

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Glossary of Common ET Terms

Glossary of ET Terms

Because knowledge is essential to making informed choices, we've developed this glossary to help ET patients, their loved ones, and caregivers better understand the terms healthcare providers may use in treating ET. You can find the complete glossary and many other helpful documents at www.essentialtremor.org/About-ET.

Action tremor

An action tremor is one that occurs during action as when performing a certain task such as reaching for an object, or bringing the object towards you. For example, a tremor that occurs when reaching for a cup or bringing a cup to the mouth.

Acupuncture

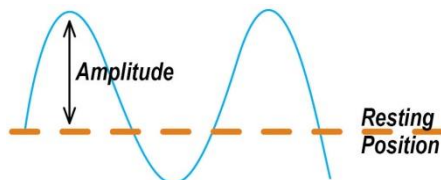
Acupuncture is an ancient Chinese therapy in which needles are inserted into certain points on the body in an attempt to control disease or pain. Acupuncture is considered an alternative (non-traditional) form of treatment for many disorders.

Alcohol

Consumption of alcohol may reduce tremor in many ET patients. Response to alcohol may be helpful to the physician in the diagnosis of ET vs. other causes of tremor. The effect of alcohol generally lasts only a few hours and excessive alcohol can actually worsen tremor. Alcohol should be used in moderation and responsibly to avoid injury or addiction.

Amplitude

Amplitude is the amount or range of movement caused by the tremor of a body part. Amplitude is most often measured in centimeters or inches.



Anxiety

Anxiety is a fundamental physiologic response to stress. Symptoms may include feeling of worry, rapid heart rate, shortness of breath, brief period of tingling sensations, as well as tremor. Virtually all types of tremor are aggravated by stress and anxiety.

Archimedes Spiral

This is a test commonly used to evaluate tremor. The patient is asked to draw a spiral without their hand touching a surface, through which the physician can determine the severity of the tremor. This simple, inexpensive test is also valuable in determining the effects of a drug on tremor.

The patient may be asked to draw the spiral before and after receiving a drug in order for the physician to evaluate the effect of the drug on the patient's tremor and at what dosages the drug is most effective.



Spiral drawn by a patient receiving no drug.



Spiral drawn by a patient receiving a trial dose of a drug.



Spiral drawn by a patient receiving a more effective dose of the same drug.

Ataxia

Ataxia is a neurological symptom that has many causes. It usually results in poor coordination, clumsiness, and abnormal walking in such a way that the feet are wide apart. Walking may resemble that of someone who is intoxicated. ET patients may experience mild ataxia.

Benzodiazepines

Benzodiazepines are a class of drug used to treat several neurological disorders. Their mechanism of action includes calming or depression of the central nervous system. They are used to treat certain types of seizures, muscle spasms, and anxiety. They can also be used as sedatives. They have been shown to help tremor to some degree. They can be habit-forming and tolerance to the medication can occur with chronic use. Clonazepam (Klonopin®), diazepam (Valium®), lorazepam (Ativan®), and alprazolam (Xanax®) are examples of benzodiazepines.

Beta-blockers

Also known as Beta-adrenergic blockers, beta-blockers are a class of drugs that block beta receptors in the brain. Beta-blockers decrease the effect of the sympathetic part of the autonomic nervous system and are commonly used to reduce high blood pressure and treat migraine. They can also suppress tremors in many patients with ET. There are several types of beta receptors. It is thought that the development of drugs that could act specifically on individual beta receptors would lead to more effective tremor control. Propranolol (Inderal®) is a beta-blocking drug.

Biofeedback

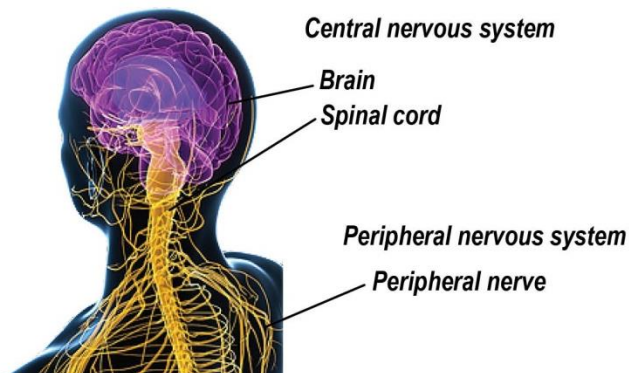
Biofeedback is a form of relaxation therapy in which body functions, such as heart beat or breathing rate, are consciously controlled through feedback from an outside device such as a heart monitor.

Botulinum toxin

Botulinum toxin is a toxin produced by bacteria often associated with food poisoning (botulism). It is injected in very small doses into the muscles to reduce their hyperactivity. It is used to treat some types of dystonia and tremor. It is marketed for this purpose as BOTOX® (botulinum toxin type A), Dysport® (botulinum toxin type A), Xeomin® (botulinum toxin type A) and MYOBLOC® (botulinum toxin type B). It can be used to reduce/treat head tremor that is not responsive to ET medications or DBS.

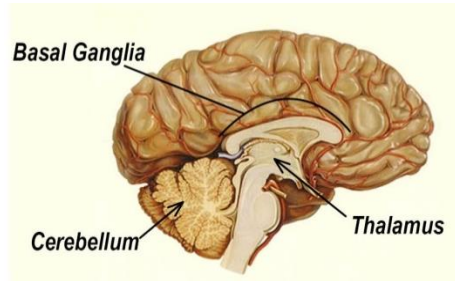
Central nervous system (CNS)

The CNS is made up of the brain and spinal cord. It controls voluntary acts, consciousness, and mental activities.



Cerebellum

The cerebellum is an area of the brain and is involved in many functions including coordination of movements, balance, and speech. Damage to the cerebellum can result in clumsiness and action tremor. Studies have shown that the cerebellum may be involved in ET.



Deep Brain Stimulation (DBS)

DBS is an adaptable therapy that uses mild electric pulses to stimulate the brain and block the signals that cause tremor. Therapy includes implanting an insulated wire lead in the thalamus (structure targeted for ET). The lead is connected to a pulse generator or battery (like a cardiac pacemaker) implanted beneath the skin in the chest. Physicians and trained health care providers activate and adjust the system. The patient can also turn the stimulation on and off with a patient programming unit or a hand held magnet. All parts of the system are under the skin.

Frequency

Because tremor is a rhythmical movement, it completes a cycle in the same way as does a clock pendulum. It swings in a direction and returns to its starting point, then swings out again and returns to its starting point, etc. The frequency of a tremor is the number of cycles it completes in a second. Frequency is measured in hertz. ET usually has a frequency of 4 to 10 hertz.

Gamma knife thalamotomy

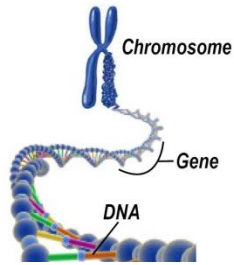
Gamma knife thalamotomy is a form of thalamotomy in which the area of the brain involved in tremor production is destroyed by creating a lesion by radiation beams. It can be very effective in reducing or diminishing tremor. The results may take several weeks to be seen and it is an irreversible procedure. It is used in patients who are very old or not good surgical candidates for DBS implantation.

Gabapentin (Neurontin®)

Gabapentin is an anti-seizure medication that can be used to treat ET.

Gene

Genes are the basis of heredity. Through a chemical called DNA, genes direct the information that determines all our characteristics such as how we look, color of our eyes, our height, formation of inner organs, etc., as well as the development of genetic diseases.

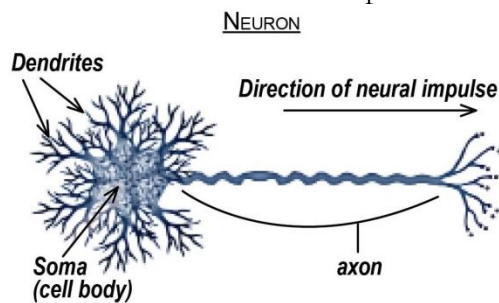


Head tremor

Most people who have head tremor actually have neck tremor that causes their heads to shake. With a yes-yes head tremor, the head nods up and down as if the person were saying yes. With a no-no head tremor, the head shakes from side to side as if the person were saying no. Some patients have head movement in all directions. Head tremor is seen in ET and dystonia.

Neuron

A neuron is a nerve cell made up of three parts. Dendrites with receptor sites receive information from other cells, a cell body integrates the information from all the receptor sites, and an axon releases neurotransmitters to pass on information.



Orthostatic tremor

The development of a tremor involving the upper legs, buttocks, and lower trunk after standing for a period of time is characteristic of orthostatic tremor. The tremor subsides when the individual sits, leans against something, or walks.

Positron emission tomography (PET) scan

This is an advanced scan that can detect and produce images of brain chemical activity.

Postural tremor

This is a tremor that occurs while the person voluntarily maintains a position against gravity. It is present when the individual holds his or her hands out-stretched in front. This tremor is often present in ET.

Sporadic occurrence

Sporadic occurrence refers to the development of a disease in an individual with no family history of the disease. In other words, the disease appears to occur by chance. ET can be either sporadic or inherited.

Thalamotomy

Thalamotomy is a type of brain surgery that involves making a lesion (controlled destruction of brain tissue) the size of a pea in the thalamus to alleviate tremor. Thalamotomy has been shown to effectively reduce tremor in ET.

Vocal tremor (voice tremor)

Vocal tremor causes a characteristic quivering of the voice, most evident when attempting to sustain a long note. Voice tremor is often seen in ET.

A special thank you goes to Dr. Kelly Lyons, Director of Research for the Parkinson's Disease and Movement Disorder Center at University of Kansas Medical Center in Kansas City, KS and IETF Board President, and Dr. Arshia Sadreddin, Director at Movement Disorders Center, California Pacific Medical Center in San Francisco, CA, and IETF Medical Advisory Board Member, for taking the time to write and review the information presented in this glossary.

What Is Essential Tremor?

The History of Tremor

By Jean P. Hubble, M.D.

"All the world is a stage and all the men and women merely players."

- Shakespeare (In As You Like It, 2.7.139)

Perhaps we are all players in life and as such, our words and actions are keenly regarded by others. In an effort to understand what it is to be human, we strive to capture our nature in our cave etchings, parchments, books, arts and videotapes. We take interest in what serves to distinguish one individual from another. This may include facial features, hair coloring, voice and body build. It also includes gestures and movements. Tremor can be a conspicuous body movement, so it is not surprising that it has been noted and interpreted throughout history.

The earliest depiction of tremor may come from Ayurveda, a sophisticated philosophy of life and science entwined with the mysticism of Hindu. It was developed four to six thousand years ago by ancient peoples in the region now known as India and is arguably the earliest formalized system of medicine. The Ayurvedic approach to disease and healing could be likened to what we know as "holistic medicine". At least one medical historian has suggested that references to hand tremor are contained within these records.

Medicine was also highly developed in early Egyptian culture. Scholars note a possible reference to tremor contained in medical documents from about 1200 B.C. The validity of such references could be challenged since the translation and interpretation of such records may be obscure. Assuming that these are accounts of tremor, it remains uncertain if ET or other tremorous maladies are being described.

A more certain description of tremor is found in the Bible. In Ecclesiastes (12.3), wisdom and advice are offered to the young. This includes a depiction of the physical prowess of advancing age, and comment is made of the trembling movements sometimes seen in the aged. Today most doctors would assert that tremor can occur during any time of life, although it is more common in the elderly. In another book of the Bible (Psalms 99.1), it is observed that fear can cause trembling movements. Most modern-day tremor patients would agree that strong emotions or stress accentuate their symptoms.

One of the most notable medical papers on tremor and involuntary movements was written in the second century A.D. by the Greek physician Galen. In *On Tremor, Palpitation, Spasm and Rigor*, he depicts the rhythmic nature of tremor and details other involuntary body movements. Many of his observations would still be valid.

Obviously healers such as Galen were interested in tremor; however, interest was also shown by writers and artists through the ages. Shakespeare, in the play *Troilus and Cressida* (1.3.172-175), describes the inability of an aged character to properly fasten his clothing because of tremor. The "faint defects of age must be the scene of mirth ... with a palsy, fumbling on his gorget, shake in and out the rivet."

Visual artists also contributed to our understanding of the function and malfunction of the human body. The 16th century work of Leonardo da Vinci provided detailed anatomical illustrations of the

human body. Da Vinci may have had specific interest in movement and its control when he wrote, “you will see...those who...move their trembling parts, such as their heads or hands without permission of the soul; (the) soul with all its forces cannot prevent these parts from trembling.” Rembrandt must also have been aware of the infirmities that can accompany aging when he sketched the elderly shopkeeper in *The Good Samaritan*. It has been suggested that the shopkeeper’s stooped position is indicative of Parkinson’s disease and that the depiction of the hands is consistent with the tremor of that disorder.

During the 18th and 19th centuries, accounts of tremor were produced by several physicians. In these, mention was made that once tremor occurs, it persists through the remainder of life. Comment was also made about life events or stresses that might cause or worsen tremor. The famous French neurologist Charcot pointed out that tremor is not an invariable feature of aging; he also called attention to head nodding or shaking as a manifestation of tremor. In 1817, the British physician James Parkinson prepared his landmark paper, *An Essay on The Shaking Palsy*, detailing the condition now known as Parkinson’s disease. In the essay, Parkinson distinguished parkinsonian tremor from other tremor of other disorders, including ET.

In 1887, the American physician C. L. Dana reported on a large family in which tremor was a frequent occurrence. This is the first detailed account of ET. Dana noted that various body parts could be affected and that the age of onset of tremor varied among the family members. Dana postulated that the family members with tremor were in some way distinguishable from the other, unaffected relatives. He speculated that the nervous system differed in these individuals. He called this a “neuropathic taint.”

This concept was likely influenced by the work of Sigmund Freud in the field of psychoanalysis. Effort was made to explain physical phenomena on the basis of individuals’ psychological makeup. Reported associations characterized tremor as being a sign of nervousness, drunkenness, anxiety, stupidity, intelligence, brilliance or brain damage. These assertions have not been substantiated in modern medical investigations.

Today we may be more knowledgeable about tremor, but we have yet to find an effective treatment for tremor. It is probable that various herbs or remedies have been tried through the ages. The Irish scientist Robert Boyle suggested chocolate as a remedy for tremor in 1688. It is not known if this helped the tremor, but it is suspected that the medicine was enjoyed.

The ability of alcohol to suppress tremor has been recognized for many decades. This caused some concern that higher rates of alcoholism among tremor patients would be the result. There does not appear to be a higher rate of alcohol abuse associated with tremor when this issue has been examined in modern times. The study of tremor in the second half of this century has served to dispel many old notions about tremor and tremor patients. Also, more customary medicines have been developed and have had modest success in the treatment of tremor.

Reviewing the history of tremor provides a glimpse at the observational skills of humankind and the development of modern medicine. More importantly, it is hoped that a firm grounding in the history of tremor will help expedite our modern-day search for effective therapy.

Toward a Definition of ET

By Shamaila Waseem, M.D. and Katrina Gwinn-Hardy, M.D.

The diagnosis of ET is made every day, and the term “essential tremor” is widely used by physicians and patients alike. However, how is ET actually characterized? Usually, the definition of ET is “tremor which occurs in the upper extremities, which occurs with activity and which is not due to another cause, and which may be associated with tremor of the head and voice”. However, there is actually a great deal of interest among neurologists, physiologists (people who study the function of the body), and other medical researchers regarding what ET actually is (and is not). We still have a great deal to learn about the causes of ET, how it differs fundamentally from other causes of tremor, and if there are more sophisticated ways of defining and diagnosing it than we now have.

ET is a common neurologic disorder, as much as eight times more common than Parkinson’s disease (PD). Although it does not affect life expectancy, it may cause considerable functional disability and serious psychological effects. There are, as yet, no specific anatomical, physiological, biochemical or genetic markers for the condition.

Positron emission tomographic (PET) studies in patients with ET reveal increased cerebellar activity even at rest, a finding that is consistent with the cerebellum having an important role in the generation of tremor. The cerebellum is a region in the back of the brain which is important in the timing and coordination of movements.

There is evidence from several large families in which ET occurs that there is a genetic component. However, the regions on certain chromosomes which have been found are only the cause in a few identified families known to date. While about 50% of people with ET have one or more affected family members, a clear genetic cause is not likely in most people. Rather, there are probably multiple inherited influences which contribute to the development of tremor. Still, it is very important to study families in which a large number of people have ET, as that is one way to learn more about the biological characteristics of the disease, and what is learned from those genetic cases can be used to look for the causes in other people.

The Diagnosis of ET

Diagnosis is currently based on the clinical finding of postural tremor, that is, tremor which is seen with the arms outstretched and in the hands and fingers during an activity, such as handwriting or drinking from a cup. This tremor typically predominantly affects the arms, but can affect the head and the voice also. Unlike Parkinson’s disease, this tremor does not occur at rest, and is not associated with any rigidity, trouble walking, or difficulty with balance.

Several neurologist/researchers have worked to develop diagnostic criteria for ET. This is important for those who work in patient care, such as doctors and nurses, to help them make consistent diagnoses. Such criteria are also important to allowing uniformity in how tremor is quantified and discussed in a research setting, as people strive to learn more about this disorder.

To be considered as having ET there must be moderate tremor in one arm, and activation tremor in most activities including pouring water from a cup, eating, writing, and/or drawing a spiral. The tremor must interfere with at least one activity of living or be associated with head tremor. Of course, one must make sure that there are not medications that the person is taking which cause

tremor, and that they don't, in fact, have Parkinson's disease (PD) or other diseases in which activation tremor will occur. It is important to keep in mind that not every patient fits into these criteria, and each doctor and researcher still uses their best judgment to make the diagnosis.

Measurement of Tremor

The term "oscillation" is very frequently used in thinking about the measurement of tremor and its origin in the nervous system. This term, oscillation, means "to move back and forth, to fluctuate", and in the nervous system, this very often means a regular back and forth movement of charge or energy within or between groups of nerve cells. Tremor may derive from mechanical oscillations (such as a jump one might get in a muscle every time the heart beats) or reflex oscillations (which would include, for example, the changes that happen in the nervous system in response to stimulation). Also, when considering the causes of tremor, researchers discuss normal central oscillators (changes which normally occur in all brains of people, which are important for normal brain timing and regulation functions) and pathological central oscillators (those which cause the back and forth movements between cells in the setting of illness or abnormality in diseases such as ET and PD).

Methods of studying tremor include accelerometry and electromyography (EMG). Accelerometry is used to measure tremor, in terms of amplitude (size) and frequency (speed) of the movements. EMG measures the movements of the actual muscles, and when several muscles on opposite sides of a limb, such as an arm, are compared, the tremor can be accurately measured and used in comparison with the tremor in different people or tremor in that same person at an earlier time. Tremor occurrence, tremor intensity and tremor frequency are highly reproducible and these methods are specific, sensitive, reliable and valid for detection of tremor. This allows differentiation and quantification of tremor and therefore, helps in differentiating between PD and ET. Unfortunately, only a few medical centers use this clinically and at most centers, it is only a research tool.

Parkinsonism and ET

Differentiating between PD and ET is the most important diagnostic skill in evaluating tremor. However, confusing the issue are the results of recent studies showing that PD and ET are actually related, at least, sometimes. In one such study, a large family with apparent autosomal dominant levodopa-responsive Lewy-body Parkinsonism (that is, rather typical PD) was investigated using genetic methods. After performing genome screen, a region of chromosome 4p was found to be strongly associated with PD in that family. Unexpectedly, however, this particular region was also associated with those who had only ET! This data demonstrates that a new genetic location for PD is associated with, as an alternative manifestation, ET. Thus, in some circumstances, postural tremor, resembling ET can occur rather than PD, as the result of a particular gene. What makes the disease go "one way or the other" is not known and an important area of further research.

What Can We Hope For The Future?

Currently, we rely on clinical examination by a doctor to make the diagnosis of ET. Other information can be obtained with the more sophisticated tests commented on above. PET scanning remains a research tool at this time and is not yet used to make the diagnosis of ET. But as it is becoming increasingly available, it is becoming more important in learning "what is ET and what is not ET?" Likewise, neurophysiology testing, such as accelerometry and EMG remains largely a research tool at most institutions but someday may be key in delineating different features and causes of tremor.

Brain examination after someone has passed away, in which a person donates their brain to science, will give us insight into the changes in the cerebellum and other structures, which are important in generating ET in the nervous system. Genetic studies will give us further clues regarding why some people get ET in a family, some might get PD, and others might not get either. The future then looks bright for increasing our knowledge and understanding of what causes ET. And only once we better understand the cause, can we better search for treatments and a cure.

The Natural History of ET

By Rodger J. Elble, M.D., Ph.D.

The clinical expression of ET is quite variable among members of the same family and among all patients in general. One family member may develop symptoms late in life while another may experience tremor since childhood or even infancy. Tremor is variably distributed in the head and neck, voice and upper extremities, so that one family member may have bothersome head tremor while another has tremor only in the hand.

The development of ET is usually so insidious that patients cannot date the onset of symptoms to within less than five years. Other patients claim to know within a year, but this usually means they recall their first troublesome incidents with tremors, not when the tremors started. Like most gradually progressive diseases that begin insidiously, ET is often unrecognized until someone brings it to the patient's attention, or until the tremor reaches sufficient magnitude to interfere with daily living.

The rate of progression of ET is also variable. Many patients are aware of tremor for years before seeking medical attention. Others report that their symptomatic tremor developed over a few years. Such a rapid course should warn that other exacerbating medical or neurologic problems may coexist.

The course of ET is variable even after it becomes a problem to the patient. Some deny progression of tremors over years. Remissions, though reported, are rare. However, spontaneous fluctuations in the amplitude of tremor are common and occur throughout the day and from day to day. Patients report inexplicable times when tremor is sufficiently quiet to write letters or to perform fine motor activities. The basis for these dramatic fluctuations is not known, but their occurrence suggests that ET is not irreversible. A better understanding of why this is so might lead to more effective treatments. Prospective studies are needed to carefully define the natural history of ET. Too much of our present knowledge is derived from anecdotal information.

The Pathology of ET

By Ali H. Rajput, M.D.

Every one of us has tremor that is neither visible nor handicapping. This type of tremor is called physiologic (normal) tremor. However, there are many forms of abnormal tremor. The most common of these is ET. In the past, it was believed that ET did not produce disability. More recent studies indicate that ET produces both psychological and functional handicaps in a significant proportion of cases.

Understanding the underlying abnormality which leads to the production of ET is important. ET is a disorder restricted to human beings – there is no similar naturally occurring disease in other animals. Additionally, we are unable to produce a good experimental animal model. The scientific studies in ET are therefore entirely dependent on observations in human beings.

In contrast to Parkinson's disease (PD), the interest in scientific studies of ET has been negligible. For example, there have been thousands of pathologic and biochemical studies on PD brains whereas only eight ET autopsy cases have been reported in the literature, and we have studied the brains of nine additional ET cases. Considering that ET is a far more common disorder than is PD, the lack of interest in studies aimed at understanding ET is remarkable and frustrating for those of us who do wish to investigate this disorder.

Because of some overlap in clinical features between the late-occurring ET cases and PD, it was postulated that these ET patients have an increased risk of developing PD. The most useful information is the recognition that the brain is normal upon examination by the naked eye and on routine microscopic observation. Careful histologic (study of tissue structure) examination has failed to uncover any consistent abnormality in the brains of patients with this disorder.

The nervous system damage in PD patients, on the other hand, is well known. Based on pathologic studies, it is now evident that ET is not an unusual variant of PD. Our studies have also concluded that ET patients do not have an increased risk for developing PD in later age. Since ET and PD are both common disorders in the elderly population, they could co-exist in some individuals.

The tremor of ET patients clinically resembles tremor seen in cerebellar diseases. It has, therefore, been suggested that the cerebellum or its connections with other parts of the brain may be abnormal in ET patients. One preliminary Positron Emission Tomography (PET) study of four cases has reported an abnormal metabolic pattern in the cerebelli of ET patients. The cerebellum and its connections, on the other hand, are reported normal when observed by the naked eye and by routine microscopic examination. It is possible that more refined studies using special stains, electron microscopy or neurochemistry of those regions will identify abnormalities specific to ET.

The biochemical studies of the ET brain are, so far, limited to one (our) patient. Many more neurochemical studies are necessary to be able to identify the pattern that is specific in ET. It is evident that if we are to increase our understanding of ET, far more work is needed. Members of the Medical Advisory Board of the IETF are embarking on studies that will include clinical and pharmacologic studies in ET patients during life and pathologic and biochemical analyses of brains at autopsy. Unfortunately, there are no shortcuts to such studies since the disease is exclusively a human disorder, and there are no good animal models on which to conduct research.

ET Fact Sheet

IETF publication

- ET is a neurological condition that causes shaking of the hands, head, and voice.
- ET can also cause legs and trunk to shake, and some people have a feeling of internal tremor.
- An estimated 10 million Americans have ET.
- Although there are many possible causes of tremor, ET is the most common. Still, misdiagnosis of tremor is a problem. According to the *Canadian Journal of Neuroscience*, general neurologists treating tremor have a diagnostic error rate of 25% to 35%.
- ET is often misdiagnosed as Parkinson's disease, yet according to the National Institute of Neurological Disorders and Stroke, eight times as many people have ET as have Parkinson's.
- ET is not confined to the elderly. Children and middle-aged people can also develop ET. In fact, newborns have been diagnosed with the condition.
- There is evidence that ET is genetic. Each child of a parent who has ET has a 50% chance of inheriting a gene that causes the condition. However, sometimes people with no family history of tremor develop ET.
- Few effective prescription medications are available for treating ET. Unfortunately, it is estimated that less than 60% of people with ET are helped by these medications.
- Researchers estimate that 4% to 5% of people age 40 to 60 have ET. The incidence rate for people age 60 and older is estimated at 6.3% to 9%.
- People who have ET become disabled at worst and feel frustrated or embarrassed at best.
- Quality of life is a big issue for people with ET. Daily activities such as feeding, drinking, grooming and writing become difficult if not impossible.
- Many people with ET are too embarrassed to go out in public and remain isolated in their homes, which can lead to depression.

Types of Tremor

Tremor Talk

Tremor is an involuntary shaking of one or more body parts. There are many causes and types of tremor. One way to classify tremor is by looking at whether tremor occurs when a person is at rest, when active, or when engaging in a specific task such as handwriting. This article discusses these classifications of tremor along with their sub-types and how they relate, or do not relate, to ET.

Rest tremor occurs in a body that is supported or relaxed in such a way that there is no intended movement such as when lying in bed or sitting in a chair. Amplitude, or the size, of a rest tremor often increases during mental stress such as when counting backwards or during movement of other body parts as in walking. Rest tremor is typically suppressed by intentional movement.

Pill-rolling hand tremor is a type of rest tremor that is seen most commonly in patients with Parkinson's disease. The fingers and wrist move in a way similar to when a small round object such as a pill is rolled with the hand. Pill-rolling hand tremor is not characteristic of ET.

Postural tremor occurs when a person attempts to hold a body part motionless against the force of gravity such as when extending the upper arms horizontally, pointing at objects, sitting up straight in a chair without support for the upper body or sticking out the tongue.

Physiologic tremor is a postural tremor that everyone experiences. Often invisible, it can be seen by placing a piece of paper between the fingers of an outstretched hand.

Enhanced physiologic tremor is a visible postural tremor that occurs in the absence of neurologic disease and is caused by medical conditions such as anxiety, hypoglycemia, certain medications, hyperthyroidism and withdrawal from alcohol or benzodiazepines. It is usually reversible once the cause is corrected.

Orthostatic tremor is characterized by rhythmic muscle contractions of the legs and trunk immediately after standing. Tremor is felt in the thighs and legs and may not be visible. The patient is bothered most by a sensation of unsteadiness while standing but not while walking. The shakiness stops when sitting or reclining. Symptoms resembling orthostatic tremor occasionally occur in ET, but most clinicians believe that classic orthostatic tremor and ET are different disorders.

Kinetic tremor occurs during a voluntary movement such as reaching, writing, drawing, pouring water, drinking from a cup, eating with utensils, and speaking.

Intention tremor is a kinetic tremor that occurs during visually-guided movements toward a target destination such as when looking at and then reaching for a pen lying on a table. Little or no tremor is present at the beginning of the movement, but as the hand approaches the pen, there is a dramatic increase in tremor. During a neurological examination, intention tremor can be seen when first touching one's own nose and then reaching out to touch the physician's finger. Tremor is seen as one's finger approaches the nose and the physician's finger but there is much less tremor in between the two targets.

Task specific tremor is largely or solely limited to a specific task or movement such as writing, speaking, or smiling.

Isometric tremor results from muscle contraction against stationary objects such as when squeezing a doctor's fingers during an examination, pushing against a wall, flexing the wrist against a table, making a fist.

Action tremor occurs during any voluntary contraction of skeletal muscle — any intended use of a body part. An action tremor may be any combination of postural, kinetic, and isometric tremor.

ET is typically a mixture of postural and kinetic tremor with intention tremor in advanced cases. Rest tremor is uncommon and occurs only in patients with advanced ET. In most instances, this rest tremor is actually a postural tremor caused by incomplete muscle relaxation because a body part is active against gravity, or not really at rest. Rest tremor with little or no action tremor is usually due to Parkinson's disease and is never caused by ET.

Who Gets Essential Tremor?

Genetic Concepts and ET

By Mitchell F. Brin, M.D.

ET occurs in all parts of the world. The highest prevalence rates reported are in Sweden and Finland. In the United States (U.S.), the estimated prevalence is 300 to 415 per 100,000 population. There is considerable variation in age of onset, and incidence rates increase with age with onset most common after age 40.

A study in the U.S. by Dr. Haerer and colleagues found ET to be more prevalent in women than in men and more prevalent in whites than in blacks. Dr. Rajput, though, found incidence rates to be no different for men and women.

The hereditary nature of ET has been recognized for 100 years. Dr. Critchley noted in 1949 that sporadic cases occur, but there is a strong genetic determinant, with an autosomal dominant mode of transmission being most common. It is generally accepted that 35% to 50% of the cases of ET are familial. For the purposes of this article, ET refers to the category of tremor that has no known cause and can be inherited.

There are various ways that a disorder can be inherited or passed on within a family (pattern or mode of inheritance), including autosomal recessive, autosomal dominant, x-linked recessive or multifactorial inheritance.

Autosomal Recessive

In a recessive pattern of inheritance, an individual with the disorder has received two genes for the disorder, one from each parent. The parents are referred to as “gene carriers” and do not exhibit the disorder themselves. Recessive disorders tend to be more common among specific ethnic groups; classic examples are Tay-Sachs disease among Ashkenazi (eastern European) Jews and sickle-cell anemia among blacks.

Autosomal Dominant

In a dominant pattern of inheritance, an individual has inherited the gene for the disorder from one of the parents, and this is sufficient to cause symptoms. If a disorder is dominant, many or few family members can be affected since each has a 50/50 chance of inheriting the gene. The disorder may be passed on from parent to child to grandchild, and thus continue for many generations. It is also possible for a dominant gene to be expressed in different ways so that some family members can be more severely affected than others.

When a dominant gene “skips a generation” or is passed on by an individual who shows no symptoms of the disorder, this is referred to as non-penetrance. The unaffected individual can have affected children. Some well-known examples of dominant disorders are Huntington’s disease (Woody Guthrie died with this) and neurofibromatosis (the so-called Elephant Man’s disease).

X-linked Recessive

An unusual pattern, with x-linked or sex-linked inheritance, males are typically affected by the disorder and unaffected females are carriers of the gene that is on the X (sex-determination) chromosome (females carry two X chromosomes, males have one X and one Y chromosome). Examples of x-linked disorders are hemophilia and muscular dystrophy.

Multifactorial Inheritance

One other pattern of inheritance is multifactorial inheritance, which means that many factors, both genetic and environmental, can combine together to cause a disorder.

Many patients with ET have no family history of tremor. These are called sporadic occurrences, and may reflect either the first time in a family that the gene is being expressed or a mutation or change in the genetic material. Other possibilities are that relatives who may have carried the gene for tremor died too young to show signs, or perhaps tremor was very mild in ancestors, was never diagnosed, or simply never discussed.

Genetics of ET

By Joseph Jankovic, M.D.

ET is inherited in an autosomal dominant pattern, which means that the affected individual inherited the gene from only one of the parents. Since each offspring has a 50/50 chance of inheriting the gene, the disorder is passed from one generation to the next. ET is expressed with a very high penetrance, which means that individuals inheriting the gene will exhibit symptoms.

ET is often accompanied by other genetic diseases such as dystonia (involuntary, sustained muscle contractions producing abnormal movements such as writer's cramp). In our study of 252 members in four large kindreds (a group of persons related to another) with ET, three of the kindreds had a total of 41 members with the combination of ET and dystonia and two had associated Parkinsonism. Besides the one kindred with pure ET without any associated disorders, we subsequently studied 216 individuals of another large kindred with pure ET. The observation in these families of earlier age at onset in successive generations suggests the phenomenon of anticipation, although the relatively small number of subjects and the possibility of ascertainment bias precludes any definite conclusions.

Thus far, three gene loci for ET have been identified. A genome scan of 16 ET Icelandic families containing 75 affected relatives with definite ET identified a marker for the familial essential tremor gene, FET1, on chromosome 3q13. An analysis of a large Czech-American family established linkage to a locus ETM on chromosome 2p22-p25. Two of our families with pure ET and one with ET-parkinsonism-dystonia also mapped to the same locus. Another marker for ET was mapped to chromosome 4p14-16.3 in a family with autosomal dominant form of Parkinson's disease. The marker was present not only in individuals who had Parkinsonism, but also in family members without Parkinsonism who exhibited isolated postural tremor clinically identical to ET. It is likely that additional gene loci will be identified in the future.

While the discovery of the gene loci is extremely exciting, a DNA test for ET will not be possible until the actual gene causing ET is identified. More importantly, the identification of the ET gene and the responsible mutation is absolutely critical for understanding the pathogenesis of this condition. Nevertheless, the recent discoveries of the gene loci represent the first and crucial steps in finding the cause of and eventually, a cure for ET.

Epidemiology of ET: An Update

By Ali H. Rajput, M.D., and Michele L. Rajput

Estimates of prevalence rates are dependent upon the definition of the disease and the ascertainment of cases. This is especially true in screening for possible cases of ET. It is necessary to exclude other types of tremor. Therefore, we must first form a clear definition of ET including type of tremor, site(s) of tremor, and duration of symptoms.

Some who self-report “tremor” may have physiological tremor. This is a tremor present in all human beings. It is not clinically evident but may appear under stress. Alternatively, some who have ET may have mild symptoms or no symptoms and may consider the tremor to be part of “normal aging.” This population has not been seen by a physician and so is not diagnosed as having ET. Any study including only clinic-based cases miss these kinds of ET cases. This results in an underestimate of the prevalence of ET in the general population.

Prevalence Rate

The number of ET patients per 100,000 of the general population at any given time is known as the prevalence rate. Studies in Rochester, MN, looked at the prevalence of physician-diagnosed ET in the general population including all ages. The prevalence rate was found to be 306 patients/100,000 population (0.3%). This study included only patients whose symptoms were diagnosed by a physician. As noted earlier, many persons with ET do not see a physician with tremor complaint. This prevalence rate is therefore likely an underestimate. If a careful screening for ET of all Rochester residents were conducted, more ET cases would be ascertained.

A few studies have undertaken a door-to-door survey for ET. One study, in Copiah County, MS, which included only persons 40 years and older with tremor of the limbs, head, or voice, reported a prevalence of 410/100,000 population (0.4%). This too is likely an underestimate as the definition included the provision that tremor is present for at least 10 years duration or the patient has a family history of ET. A similar study in Finland was also restricted to persons 40 years and older but included tremor present for one year or longer in duration. The prevalence rate reported was 5552 cases/100,000 population (5.6%).

Two other studies included people of all ages, one in India and one in Italy. Both studies defined ET as an action tremor of the limbs, head or voice, present for more than one year or with family history of ET. The prevalence rate in India was 1,663/100,000 (1.7%) for all ages and 2,763/100,000 (2.8%) for the population over age 40 years. Prevalence in Italy was lower, 405/100,000 (0.4%) for all ages and 1,074/100,000 (1.1%) for the population over 40 years old.

From these studies we can conclude that the prevalence of ET in the U.S. population is in excess of 400/100,000 (0.4%) and that a large number who suffer from mild ET go unrecognized. As life expectancy in the general population is increasing, we can expect higher prevalence of ET in the future.

Age

With advancing age, the incidence and prevalence of ET increases dramatically. Incidence is defined as the number of new cases per 100,000 population. Annual incidence of new medically diagnosed patients in the Rochester study was 2.3/100,000 for the population under age 19 and 84.3/100,000

for the population over age 80. In the Copiah County study, prevalence was 119/100,000 (0.1%) for ages 40 – 69 years, 1,181/100,000 (1.2%) for ages 70 –79 years, and 1,621/100,000 (1.6%) for those 80 and older. Thus the ET problem is concentrated in the elderly. In a survey of institutionalized persons 65 years and older, we observed that 10% had moderate to severe ET. None of these patients however, were aware of the diagnosis or were receiving treatment.

Gender

Males and females are affected equally, but the location of the tremor does show some sex-related differences. ET of the head and ET of the vocal chords are more common in females than males.

Race

Few studies have reported race-specific prevalence rates. The Copiah County study reported a higher prevalence of ET in whites than in blacks, but the difference was not statistically significant. A study of Medicare recipients in New York estimated ET prevalence in blacks, Hispanics, and whites. Whites had the highest prevalence (3.1%) and blacks had the lowest (1.8%) with Hispanics in the intermediate range (2.2%).

Family History and Genetics

In approximately half of all ET cases, there is a history of tremor in other family members. We have observed that if one member of the family had ET and another member also had tremor, 90% of the secondary cases had ET rather than any other cause of tremor.

In most patients whose family history is available, the mode of inheritance is autosomal dominant. Researchers have begun to look for the genes involved in familial ET. One study from Iceland revealed a specific location of the gene on chromosome 3. Future research in this area should prove valuable since there are probably several different genetic locations not yet identified.

Some studies suggest that ET patients are at an increased risk of developing Parkinson's disease. However, several clinical studies and autopsy studies have found that not to be the case. Since both of these disorders are concentrated in the older population, some patients would have both these diseases. Nearly one-third of ET cases in later age develop resting tremor on top of the typical ET. The resting tremor is usually regarded as a sign of Parkinson's disease. Such evolution of resting tremor that we confirmed at autopsy proved to be a natural course of ET in some patients. The second diagnosis of PD should only be made by an expert in movement disorders.

Quality of Life

ET was once known as a "benign" condition but this is a false characterization. It is true that a diagnosis of ET does not shorten life expectancy, but the quality of life is certainly affected. For patients with ET, the psychological effects can be significant and can drastically hamper the quality of life. A severe tremor can interfere with eating, writing, and speaking resulting in a significant handicap. In one study of the impact of ET on daily life, all patients reported some degree of measurable and/or perceived handicap in comparison to control subjects. With advancing age, tremor becomes slower and larger in amplitude, and increasingly interferes with daily activities.

ET research, including studies of genetic linkage, physiological/biochemical basis of ET, and better treatment is being conducted at many movement disorders centers. We look forward to future developments that would enhance our understanding of ET and allow for better management of

these patients. Since ET is restricted to human beings, patient participation in these studies is absolutely essential, especially because it has genetic implications for children of the ET patient.

Prevalence of ET

By Joseph Jankovic, M.D.

ET is the most common movement disorder, but its prevalence is still unknown (Table 1). There are three reasons. First, there is no biologic marker or diagnostic test, so the diagnosis of ET may be difficult to confirm. Most neurologists, especially movement disorders experts, agree on clinical criteria, but there is disagreement about which oscillatory motor disturbances represent typical ET or ET variants, or other movement disorders. While ET is considered to be a tremor present in the hands during activity or when holding a certain posture, some head, voice or trunk tremors are also included in the category of ET. In some cases, the tremor is manifested only during certain activities such as handwriting or positions such as holding a drinking glass at a particular distance from the mouth. My view is these task specific tremors should be included in the category of ET, but others do not agree.

Second, the epidemiology (branch of medicine dealing with causes, distribution, and control of a disease in population) of ET is difficult to study because tremor severity varies. It may be barely perceptible by the patient and not recognized by the next of kin or the examining physician. Such a “benign” tremor may produce minor inconvenience and be wrongly attributed to nervousness, alcohol, or a natural consequence of aging. In contrast, ET can also be manifested by a coarse, high amplitude tremor causing disability that ranges from social embarrassment to a loss of gainful employment (“malignant” ET).

Although ET is the most prevalent movement disorder, it is often mild, so few ET patients actually seek medical attention. Among 5,000 patients referred to the Baylor College of Medicine Movement Disorders Clinic, ET ranked third (16%) of all movement disorders, after parkinsonism (45%) and dystonia (28%).

Thirdly, to study the epidemiology of ET, inclusion/exclusion criteria must be defined. ET is either idiopathic (no identifiable cause) or inherited in an autosomal dominant pattern (offspring has a 50/50 chance of inheriting the gene). There is controversy whether patients with inherited peripheral neuropathy and postural tremor (Roussy-Levy syndrome) or patients with ET and associated dystonia (sustained twisting movements such as torticollis or writer’s cramp) and myoclonus (jerk-like movements) should be included in the epidemiologic studies of ET.

What about patients in whom postural tremor, seemingly identical to ET, occurs after injury to their peripheral nerves or after head trauma? These post-traumatic tremors may not always be easy to separate from the idiopathic tremors. Other secondary forms of tremor due to certain drugs, metabolic disorders and other causes may be difficult to separate from ET. A challenging problem in diagnosing tremor is to distinguish physiologic tremor from ET. These are some examples that epidemiologists must consider when designing protocols to study the prevalence (number of cases in a given population) and incidence (number of new cases in a given time period) of ET.

Despite difficulties, epidemiology has provided information about ET. Studying a total population or a random sampling of population, rather than a review of hospital or clinic records, provides the least biased epidemiologic data. Studies using these methods show the prevalence of ET in parts of the world varies from as low as 0.4% in a population aged 40 or older in Copiah County, MS, to as high 0.6% in a population living in southwest Finland. Although another study conducted in

northern Sweden, showed relatively high prevalence (3.7%), suggesting more frequent occurrence of ET in Scandinavia than in the U.S., the ten-fold difference can also be explained by differences in the diagnostic criteria.

For example, investigators in the Copiah County study insisted on very strict criteria including the presence of a family history or symptoms for at least 10 years and serious disability in handwriting, daily activities or speech. With less stringent criteria, such as in the Scandinavian studies, the U.S. prevalence would probably be similar.

The population-based study of ethnically and linguistically isolated districts in New Guinea showed a prevalence of ET in individuals older than 40 varying from 0.14% to 2.24% even in a relatively small geographic area. Although only 21% of the population was older than 40, there was a five-fold difference in the prevalence of ET in the general population (0.35%) and in persons aged 40 or older (1.64%).

Other studies have confirmed that the prevalence of ET increases with age; only about 20% of ET patients have symptoms before age 30, and the vast majority is 50 or older.

Table One

Prevalence of Movement Disorders (Per 100,000)

- Essential tremor 415
- Parkinson's disease 347
- Dystonia 33
- Gilles de la Tourette syndrome 29
- Progressive supranuclear palsy 8
- Hereditary ataxia 8
- Olivopontocerebellar atrophy 6
- Huntington's disease 5
- Multiple system atrophies 2
- Wilson's disease 1

Gender Differences in Tremor

By Jean P. Hubble, M.D.

The diagnosis is usually quite obvious - rhythmic shaking of the hands. Despite the fact that essential tremor (ET) is common and easily recognized, there are many aspects about the clinical presentation which are poorly understood.

Virtually all individuals with ET have tremor of the hands. The next most frequently affected body area is the head and voice. Head and voice tremor occurs in about 30% of ET patients in most reports. It is possible that age, duration of the tremor disorder, or gender might be predictive of the clinical expression of ET, including affected body region. To test this notion, we conducted a study at the University of Kansas Medical Center Tremor Clinic in 1992-1995. The results of this work were reported in the journal *Movement Disorders*, 1996, Volume 12, pages 969-972.

Clinical information obtained from 450 ET patients was analyzed. The average age was 67 years while the average duration of tremor was 25 years. There were 232 men and 209 women included in this analysis. Nearly all of the study subjects including men and women had tremor in the hands. However, only 30% of the men had tremor affecting the head and voice while 60% of the women had tremor of the head and voice. In addition, hand tremor was more severe in men and head and voice tremor was more severe in women. Thus, women were more often affected by tremor of the head and voice and had more severe head and voice tremor compared to men. There is no ready explanation for this finding. It is possible that the sex chromosome (X<Y) influence the expression of tremor in men compared to women.

Alternatively, the sex hormones (estrogen, progesterone, and testosterone) may influence the location and severity of tremor in some way. Another explanation for these findings is that women with tremor may have another neurological symptom called "dystonia." Dystonia refers to involuntary, sustained muscle contractions which can cause unusual postures including head turning. It is possible that some of the head tremor in subjects participating in the study was due to dystonia of the neck (torticollis). In contrast to gender, age and duration of tremor did not distinguish those individuals who had tremor affecting the head and voice from those who had hand tremor only.

Unfortunately, there are few effective remedies for head and voice tremor. Sometimes, the conventional tremor medications will help suppress head tremor to some degree. These medicines include propranolol, primidone, and clonazepam. Botulinum toxin injections into the neck muscles may help suppress head tremor. However, botulinum toxin injections can result in transient muscle weakness so that the patient who undergoes injections may have difficulties with head droop or difficulties with swallowing for several days or a few weeks. Botulinum toxin injections may help voice tremor when it is associated with dystonia of the vocal cords (muscle contractions of the vocal cords). Deep brain stimulation of the VIM nucleus of the thalamus has been shown to improve head and voice tremor in some patients.

Part of the limitation in our ability to better treat tremor is due to the fact that the causative brain mechanisms for tremor are not precisely understood. It is possible that a better understanding of the brain chemistry in ET will lead to better treatments.

Can a Child Have ET?

By George W. Paulson, M.D.

Every neurologist who sees children occasionally sees a child with what seems to be essential tremor. Furthermore, there are adult patients with ET who say that when they were children their heads or hands shook, particularly when they were anxious or tense. There is no reason to believe that ET that begins in childhood is any different than the adult form.

Tremor may be more delicate in childhood, and there should be no associated neurologic handicaps. By the time of adolescence most people have learned that in times of stress, shaking may occur; for example, when standing in front of a classroom or when dealing with a figure of authority. At such times tremor may be present in the voice or hands. Such a tremor can often be considered an exaggeration of the normal physiologic tremor due to anxiety, extreme exertion or fatigue that every person experiences occasionally.

Children with ET have more than an exaggerated physiologic tremor, although one of the possible mechanisms suggested for ET has been a failure of normal dampening of physiologic tremor caused by a central deficit. Most people feel, however, that ET is not solely generated by receptors in the nerves or muscles in the limbs, but also by complex central mechanisms, not fully understood at this time. Central input into the spinal cord may matter, but if one actually cuts the nerve roots into the cord, by doing rhizotomy, the tremor may remain in the limb. It appears that tremor relates to a combination of some segmental aspects, some spinal cord aspects and central circuits that are not well defined at the present time.

ET can be inherited in an autosomal dominant manner, but it is also seen frequently without any family basis and clearly may present in the first decade of life. If there are associated clinical features, one has to consider cerebral palsy and other central nervous system degenerative disorders. As a rule in the child (and also in the adult with ET) there are no abnormalities in intellect, speech or walking. Some children who have small amplitude tremors, which could be exaggerated physiologic tremors, will see such tremors improve as they age. The larger amplitude tremors or the head shaking, which is difficult to distinguish from the head-shaking of cerebellar disease, may also decrease with age, but in most instances in the later years of life a true ET type of movement will return.

Conditions such as juvenile Parkinson's disease may involve tremor, and there are conditions that involve both the eyes and head presenting as a rolling vertical tremor of the head (spasmus nutans). Dystonia musculorum deformans, or twisting due to a genetic disorder, can be associated with a tremor of the head or tremor with exertion. A treatable disorder that can occur in childhood and can cause tremor is Wilson's disease; many of these patients also have rigidity and stiffness.

Patients with isolated ET in childhood tend to do well and many respond to propranolol. Some young patients may require career counseling if they have significant tremors. They should not consider microsurgery or fine engraving as a career if the tremors are severe. Equally significant is the child's awareness that ET is a treatable problem, not a psychological disturbance or "weakness."

Talking to your child about tremor will go a long way toward helping them accept the problem. There is, as we all know, more than drugs to consider in treating ET.

Getting the Diagnosis

Tremor Analysis

By Seth L. Pullman, M.D., F.R.C.P. (C)

Tremor is one of the most common movement disorders. There are many kinds of tremor with numerous underlying causes. Some tremor disorders are more treatable than others, and most tremor disorders respond best to a particular kind of therapy. Therefore, it is important to determine the specific type of tremor so that the most appropriate treatment is started. Tremor analysis is a method of testing using specialized electronic recording devices that delineate the characteristics of tremors accurately and reproducibly.

The precise and objective findings of a tremor analysis test can help in making a proper diagnosis when the clinical picture is subtle or unclear. ET and Parkinson's disease are often confused with each other. Both conditions occur with increasing age, and the shaking movements may appear similar. Nevertheless, these two disorders can usually be distinguished on the basis of their objective physiologic characteristics. The two most important characteristics of any tremor are the amount of shaking per second, known as the tremor frequency, measured in cycles per second or hertz (Hz), and the size of the tremor movements, the tremor amplitude, measured in millimeters (mm).

In addition to its diagnostic utility, tremor analysis is useful in documenting levels of disability. The clinical exam alone cannot discern critical features of tremor with the same precision as an electronic transducer (a device used to convert one form of energy to another). For instance, sound can be connected to electrical impulses and then converted back to sound; this is how a telephone works.

Even if the underlying cause of a tremor is known, it may be important to quantify its severity on an objective basis. This is useful when comparing the changes in disability from one examination to the next or in documenting the effects of medication, physical therapy or surgery. Thus, results from the tremor analysis test supplement the clinical examination by providing clinicians with an objective and detailed evaluation of their patient's tremor movements.

Although there are several different techniques, the more commonly performed tremor studies use electronic detectors of motion (accelerometers) attached to the most tremorous body parts, typically the fingers or arms, legs, head and occasionally the trunk. Accelerometers indirectly measure how fast the body or limb moves. This information can be further analyzed mathematically to determine the frequency and amplitude of the tremor movements.

With an electromyogram (EMG), muscle activity may be recorded simultaneously with motion, usually using surface electrodes. The muscle activity will travel through the wires to the machine to be recorded on a graph overlying the most active muscles. This determines the relationship of the involved muscles to the tremor movements and reveals whether muscles that cause opposing movements (such as flexion and extension of the wrist) are working at the same time or alternatively to produce the tremor. A computer collects and manipulates the data from the accelerometers and the EMG electrodes. In this manner, frequencies and amplitudes can be determined accurately.

A typical tremor analysis test lasts less than an hour. The patient is usually seated comfortably with a series of electrodes attached over the surfaces of different parts of the body and limbs. Because tremors are categorized according to whether they occur at rest, during action or while maintaining a posture, the tremor analysis test functionally evaluates the patient during these same conditions.

Other maneuvers known to exaggerate or diminish tremor such as holding a cup of water or writing can be used to further delineate and assess movement abnormalities caused by the tremor. After the test is completed, a neurologist with experience in physiology analyzes the results and prepares a report for the referring clinician.

The tremor analysis test can provide a great amount of information and insight into the problems of tremor, and with further research, may someday help determine the underlying causes of those tremors whose etiologies are unknown.

Examination of the Patient with Tremor

By Jean P. Hubble, M.D.

Tremor, a symptom common to many conditions, frequently confounds both the general practitioner and the medical specialist.

The patient with tremor usually sees a doctor in one of three settings. First, tremor may be the sole complaint that brings the patient to the doctor. The tremor may annoy, embarrass or interfere during the usual daily activities of this otherwise well individual. In the second situation, the patient seeks help for another medical condition and tremor is incidentally noted. In the third scenario, the tremor is accompanied by other neurological problems, and thus, may suggest a progressive, neurologic disease.

In all three settings, a careful physical examination is in order that will usually allow the physician to establish the type of tremor. By identifying the tremor type, the doctor is better able to explore its implications on the patient's overall health and offer appropriate remedies.

Examination of the patient begins with the doctor asking questions such as: When did it begin? Is it always present? What makes it better or worse? Does it run in the family? Are there any other medical problems? After interviewing the patient, the physician establishes that the movement being described is tremor.

By definition, tremor is a rhythmic "back and forth" or "to and fro" movement produced by involuntary contractions of muscle. Next, the examiner will determine which parts of the body are affected. Tremor of the hands is most common, but the upper arms, head, trunk, legs, and voice may be affected either in isolation or together with hand tremor.

Then, by having the patient assume postures or perform activities, the examiner can characterize the tremor as resting, postural, or kinetic. Resting tremor is typically seen in Parkinson's disease patients. This tremor is observed when the patient loosely holds their hands in their lap when seated. Postural tremor is most prominent when maintaining a fixed posture such as the arms outstretched with the palms up. ET is usually the postural type. Kinetic tremor is seen when performing goal-directed motor tasks such as touching the tip of the nose with the index finger, and is seen in many conditions including ET.

Having characterized the tremor, a general physical examination is performed. The doctor is searching for evidence of other medical problems, particularly for signs of other nervous system disorders. Blood work or other laboratory testing may be indicated depending on individual circumstances. It may also be appropriate for the doctor to interview similarly affected family members, or talk with descendants of affected members who are deceased.

With the examination complete, the physician is better able to accurately diagnose the disorder, discuss the diagnosis with the patient, and consider treatment options.

Methods of Assessing Tremor in Clinical Trials and in Everyday Clinical Practice

By Rodger J. Elble, M.D., Ph.D.

Methods of assessing tremor severity and response to therapy

We all desire more effective methods of treating tremor. Testing a new treatment depends critically on the accurate, unbiased measurement of tremor amplitude and its impact on patients' lives. This article is a brief overview of the available measurement techniques used in research and clinical practice.

Tremor is measured with precision motion transducers and with ordinal rating scales. Motion transducers measure tremor objectively and very precisely. Rating scales are far less precise and more subjective. With ordinal rating scales, a trained examiner assesses tremor and assigns a grade or rating to tremor in a particular movement or posture. Rating scales require no instrumentation and are therefore inexpensive. However, these methods lack the precision and objectivity of motion transducers.

Any useful method of tremor assessment must be reliable and valid. A method is reliable if there is little variability in the measurement from time to time by the same investigator and by different investigators. A method is valid if it accurately reflects the patient's condition.

The main problem with rating scales is that most are very subjective, which limits their reliability and validity. Mild tremor to one examiner may seem moderate to another. In other words, there can be poor inter-rater agreement due to differences in rater experience and judgment. Consequently, investigators must come to some arbitrary agreement as to the meaning of normal, mild, moderate and severe, and they must develop adequate skill to rate tremor consistently. Unfortunately, it is often difficult for experienced clinicians to reach agreement on such issues. Furthermore, situations frequently arise in which a patient may not clearly fit one rating or another, and clinicians commonly waver in their judgment. The Tremor Research Group recently developed the Essential Tremor Rating Assessment Scale (TETRAS), which uses ratings that are tied to specific amplitude ranges in centimeters (Table). Raters are required to first estimate the amplitude of tremor and then assign the corresponding rating. This approach reduces the subjectivity of the ratings "normal, mild, moderate and severe".

Table: TETRAS ratings for upper limb tremor

- 0 = no tremor
- 1 = tremor is barely visible
- 1.5 = tremor is visible, but less than 1 cm
- 2 = tremor is 1- < 3 cm amplitude
- 2.5 = tremor is 3- < 5 cm amplitude
- 3 = tremor is 5- < 10 cm amplitude
- 3.5 = tremor is 10- < 20 cm amplitude
- 4 = tremor is \geq 20 cm amplitude

Motion transducers provide a very objective measurement of tremor (no rater is involved), and their superb accuracy and resolution make them capable of measuring changes in tremor that are not perceptible to the unaided eye. However, the advantages of high accuracy and resolution are usually

mitigated by the natural moment-to-moment variability in a patient's tremor amplitude. ET and other forms of tremor vary greatly from moment-to-moment and from day to day. Transducers easily measure this variability, which may be so great that it obscures the effect of a treatment. In other words, this variability can be so great that the advantages of transducers (accuracy and sensitivity) over rating scales are diminished because rating scales are less sensitive to random fluctuations in tremor severity. Thus, the main advantage of transducers in clinical studies is that tremor can be measured objectively without a skilled rater, and the current trend is to use transducers with scales in the assessment of ET.

Many patients exhibit tremor that depends greatly on a particular posture or movement. For example, some patients have little postural tremor but shake violently during a particular movement (e.g., writing or drinking from a cup). Consequently, tremor is usually assessed in several postures and movements. Unfortunately, the physical and electronic limitations of motion transducers may restrict their use to only a few movements and postures, and transducers, compared to rating scales, generally provide a more accurate but less comprehensive (i.e., valid) assessment of ET.

Why not simply ask the patients if they are better? This seems like a logical approach, but self-assessments are very subjective and are influenced by occupation, personal expectations, mood and personality. Nevertheless, many studies ask patients and clinicians to give a global impression of response to therapy (e.g., much improved, improved, unchanged, worse, much worse).

Some patients have learned to cope with their tremor, even though it clearly interferes with activities of daily living. Others are distraught with tremor that is barely visible. A treatment can make a patient better by treating an unrelated comorbidity such as depression, while another treatment may make the patient worse due to side effects, even though the tremor is clearly reduced. Consequently, ratings of patient satisfaction and quality of life often correlate poorly with tremor severity but are necessary for the comprehensive assessment of a treatment.

Methods of diagnosing tremor

We currently rely on clinical examination by an experienced physician to make the diagnosis of ET. Motion transducers and electromyography (recording electrical activity from the muscles with electrodes taped on the skin) are helpful in distinguishing ET from normal tremor, but there are no specific neurophysiologic or radiologic tests for ET. In short, there is still no test for ET.

Is Everything that Trembles Parkinson's Disease?

By Abraham Lieberman, M.D.

Introduction

A frequently asked question is, “Is every tremor Parkinson's disease?” Essential tremor (ET), not Parkinson's disease, is the most common cause of tremor. ET affects approximately 10,000,000 Americans, eight times as many people as Parkinson's disease.

Description

ET is sometimes called familial tremor, or hereditary tremor, or heredo-familial tremor, because in approximately 50% of patients there is a family history of tremor. ET is characterized by involuntary rhythmical tremor of a body part—usually the hands. ET usually begins in a finger, where it is apt to go unnoticed. This is unlike the tremor of Parkinson's disease which usually begins in the thumb and is more apt to be noticed. The tremor in ET may, for years, remain unchanged in amplitude, frequency, and distribution. Or the tremor may change in amplitude and distribution.

In some patients, tremor may begin in the head, the voice, or the tongue. The tremor in ET rarely begins in the legs. Tremor that begins in an arm and a leg on the same side of the body, or in the jaw, is more likely to be Parkinson's disease.

In most people with ET, the tremor worsens (or first becomes visible) when the person voluntarily maintains his/her hands in a fixed position as in holding his/her hands stretched in front of him/her. This type of tremor is called a postural tremor. In some people with ET, the tremor worsens (or first becomes visible) when the person voluntarily moves his/her hands, as in performing an action such as bringing a glass of water to his/her lips, tying his/her shoes, or threading a needle. This type of tremor is called an action tremor. Most people with ET have both postural and action trembling.

Diagnosis

The following are useful in diagnosing ET and excluding other causes of tremor.

- *A postural or action tremor that starts in both hands simultaneously* - This is unlike Parkinson's disease which is characterized by tremor that begins in one hand and is present when the hand is at rest, and the muscles relaxed. This is called a rest tremor.
- *Postural or action tremor present in both hands for at least two years* - Several conditions—including caffeine or nicotine use or withdrawal, alcohol abuse or withdrawal, lithium toxicity, electrolyte imbalance, calcium deficiency or excess, and thyroid disease— can result in tremor. This type of trembling is usually temporary and disappears when the underlying condition is corrected.
- *Head tremor* - Head tremor (as distinct from jaw trembling) with postural and/or action trembling of the hands suggests ET.
- *A strong family history* - Defined as several members of the same generation who have similar tremor, such as brothers and sisters or first cousins; several members of one or more older generations who have similar tremor, such as mother or father, aunts or uncles, grandmothers and grandfathers, great-aunts and great-uncles; several members of the younger generation who have similar tremor, such as children, nephews, and nieces.

- *The tremor of ET may be temporarily relieved by alcohol* - Alcohol, however, is not a treatment for ET. Over time, increasingly higher doses of alcohol are required to relieve tremor, and eventually alcohol fails to relieve it.
- *The tremor of ET may be relieved by primidone (Mysoline)* - Primidone (Mysoline™) doesn't relieve the tremor of Parkinson's disease. The trembling of ET may be relieved by propranolol (Inderal™). Propranolol (Inderal™), however, may also relieve, temporarily, the tremor of PD.

Prevalence

The prevalence of ET increases with age. A study that surveys people in a retirement community will come to a different conclusion than one that surveys people in a college town. A study that surveys people attending a specialty clinic (one that attracts patients with tremor) will come to a different conclusion from one that surveys people in a general medical clinic.

Among people of all ages, the prevalence of ET varies from 4,000 to 40,000 people per 1 million of population. Among people of age 60+ years, the prevalence varies from 13,000 to 50,000 people per 1 million of population. ET affects at least 10 million Americans, 3.7% of the population, and is eight times more frequent than Parkinson's disease. Although almost all people with Parkinson's disease see a physician, less than 1% of all people with ET see a physician. ET affects men and women equally. Parkinson's disease affects men more than women: 55 men for every 45 women. ET, like Parkinson's disease, can begin in adolescence.

Progression

In most people with ET, the tremor increases as time passes. And a few people with ET become disabled. The disability results solely from the tremor. This is unlike Parkinson's disease where the disability results from the slowness of movement rather than the tremor. Some people with ET may have difficulty doing everyday tasks such as handling small objects, tools, or utensils. And some people with ET have difficulty writing, drinking water from a cup or glass, feeding themselves, applying makeup, shaving, or dressing.

Cause

In most people with ET, the tremor is inherited. But how an inherited defect results in tremor is unknown. It is proposed that abnormalities, not visible, in specific brain structures—the olives, the cerebellum, the thalamus—result in these structures acting as oscillators, setting up self-propagating currents, resulting in tremor.

Parkinson's disease

This progressive disease affects 1.2 million people in America: 3,500 people with Parkinson's disease for each 1 million Americans. Typical onset is 60 years. Parkinson's disease affects 1% of all people over age 60 years; 2% of all people over age 70 years. But, Parkinson's disease is not just a disease of seniors: 15% of Parkinson's patients are 50 years or less.

Parkinson's disease is characterized by four main symptoms:

- (1) Rigidity or stiffness of one or more limbs. A stiffness, like arthritis.
- (2) Tremor of one or more limbs. The tremor is more prominent in the hands, is present when the hands are relaxed, or at rest and is asymmetrical. In 30% of people with Parkinson's, there is no tremor.

- (3) Bradykinesia or slowness of movement of the limbs and body. Rigidity, tremor, and slowness result, primarily, from a loss of dopamine in a region of the brain called the striatum. Dopamine is a chemical which facilitates the flow of impulses from nerve cells.
- (4) Postural instability results from impairment of the reflexes which allow people to adjust to abrupt changes in position. If you trip and start falling, and if your postural reflexes are normal, you're able to rapidly and without thinking adjust your body and limbs to stop falling. In Parkinson's disease, your postural reflexes are lost and if you trip, you're unable to stop falling.

ET vs Parkinson's disease: How do they differ?

By Kelly Lyons, Ph.D., Joseph Jankovic M.D., and Arik Herekar, M.D.

The characteristics listed in the table below can help differentiate between parkinsonian and essential tremor, but a medical professional should be consulted for proper diagnosis.

Parkinsonian Tremor Signs & Symptoms	Essential Tremor Signs & Symptoms
High amplitude. Lower, slower frequency.	Low amplitude. Amplitude is more variable, ranging from barely perceptible tremor to a high amplitude tremor. Higher, faster frequency.
Mostly seen at rest.	Mostly seen during action.
Generally involves slow movements (bradykinesia), rigidity (stiffness), and problems with walking or balance.	Tremor is primary symptom – slowness, stiffness, walking and balance problems are not commonly seen.
Rarely a family history (<10%).	Family history of tremor reported in the majority of patients (>50%).
Resting and postural (re-emergent); postural tremor observable after mean latency of 5 sec; rarely kinetic.	Postural, kinetic; postural tremor immediately observable; resting tremor less common.
Onset generally at ages between 55-65.	Onset most common in middle age but can occur at any time in the lifespan.
Usually starts on one side of the body and progresses to the other side; usually remains asymmetrical.	Usually affects both sides of the body initially (bilateral; symmetrical).
No effect from consumption of alcohol.	Alcohol often improves tremor.
Usually improves with levodopa treatment.	Improves with primidone and propranolol, in some cases.
Hands affected more than legs, voice and head almost never affected.	Hands predominantly affected, but tremor also present in the head and voice; rarely in the legs.
Worsens with emotional stress.	Worsens with emotional stress.

Many differences exist between essential tremor (ET) and Parkinson's disease (PD), the former being at least 8 times more common than PD.

Up to 20% of patients with ET may develop PD, and 10% report a family history of PD. However, whether ET is a risk factor for PD remains an unresolved and somewhat controversial issue. (Fekete R, Jankovic J. *Revisiting the relationship between essential tremor and Parkinson's disease*. Mov Disord 2011; 26:391-8).

As ET progresses, tremor frequency (number of repetitions per second) may decrease; however, tremor amplitude (magnitude/strength) may increase. Increased amplitude is associated with a decreased ability to manage fine, discrete motor tasks.

In those with PD, the most disabling symptoms are generally slowness, stiffness and problems with walking and balance, although tremor can also cause some disability. In PD, writing often becomes very small (micrographia) and therefore difficult to read.

The severity of ET and level of disability can vary greatly among patients. Some patients have a very mild tremor and therefore minimal to no disability. Others who have a severe tremor often cannot perform the majority of their daily activities and require help. The writing of a person with ET tends to be large and tremulous, rather than slow and small as in PD.

Anxiety can increase both ET and PD.

In addition to medications and botulinum toxin (Botox ®), deep brain stimulation (DBS) is a therapeutic surgical option for those with severe, disabling ET or PD.

ET is often associated with a strong family history (known as familial tremor), but no specific ET-related gene has been identified.

A special picture or scan of the brain that measures the uptake of dopamine, such as DaTscan, can help to differentiate between ET and parkinsonian tremor. Dopamine is depleted in patients with parkinsonism but not in patients with ET; therefore, scans showing depletion in dopamine are most likely parkinsonism and normal scans would suggest a more likely diagnosis of ET. These scans are used as a tool to help the physician make an accurate diagnosis in difficult cases.

The information above is intended to describe the fundamental differences between ET and PD. To obtain a diagnosis, please consult a neurologist, preferably a movement disorders specialist.

Misdiagnosis and its Ramifications

By Oscar S. Gershanik, M.D.

Frequently, individuals with ET are incorrectly labeled as having Parkinson's disease (PD), adding the emotional, economical and medical burden that this mistake implies. The following is a true story and will serve to illustrate the consequences of misdiagnosis.

A worried woman brought her mother for an expert opinion. After several visits to their family doctor, her mother was getting worse despite medication. The patient was a 72-year-old woman who first sought medical advice because of mild hand tremor. The tremor was most evident when she played cards or when she was holding a spoon or a cup of coffee. The doctor, without performing a thorough physical examination, diagnosed early PD and started her on Artane®. Months later she complained that the tremor was no better and she was becoming confused and her memory was suffering. The doctor said she was becoming senile and needed medication. In addition to the Artane®, she was given Flunarizine® (a calcium-channel blocker).

Months later, the patient noted worsening tremor that was present at all times, even with hands at rest, and slowness and rigidity. A visit to the doctor resulted in the addition of a second anti-parkinson medication; Sinemet®. He felt that her PD had progressed, and she needed a stronger medication. Unfortunately, this not only failed to improve the tremor, slowness and rigidity, but she developed hallucinations and delusions as well.

This time a desperate daughter and her tremulous, rigid, slow, confused and almost psychotic mother went again to the doctor. He explained to the daughter that her mother's condition was due to progression of both PD and a dementing illness and the side effects of medications she dearly needed. On top of all the drugs she was taking (Artane®, Sinemet® and Flunarizine®), he then prescribed Haldol® (a potent anti-psychotic).

Haldol® somewhat improved the psychiatric symptoms, but worsened the parkinsonian symptoms. Deciding to seek a second opinion, the patient was brought to our clinic. We saw a severely deteriorated patient. According to the family, she appeared to have aged considerably in the last year. She could barely walk; her face lacked expression. She stood with a stooped posture and tremor was evident at rest in both her arms. Her speech was almost inaudible and incomprehensible. A few questions brought us back to the initial complaint of postural and action hand tremor. The rest of her problems started after taking the prescribed medications. A diagnosis of ET was made and all medications were slowly withdrawn.

After a few months, she was again a bright, intelligent woman with mild hand tremor! Doctors need to remember that ET presents with postural or action tremor alone. PD is more complex usually presenting with unilateral rest tremor in combination with rigidity, slowness of movement and/or postural changes. ET is 8 times more frequently observed in individuals over age 65 than is PD. The symptoms this patient developed in addition to the original tremor were secondary to medications that she did not need. Artane® may cause memory problems and confusion in older individuals. Flunarizine® may cause or worsen depression and parkinsonism. Sinemet® may cause psychiatric symptoms in predisposed individuals. The main side-effect of Haldol® is parkinsonism.

Three Steps to Choosing a Doctor

By Margaret M. Maier Hoehn, M.D.

Whether you are searching for a physician for the first time, considering changing physicians, or seeking a second opinion, do not give it less care and attention than you do the choice of a new television set.

Consider first, basic credentials. Do not be embarrassed to ask physicians about their training and qualifications. In all English-speaking countries, four-year medical schools are inspected at regular intervals. The quality of patient care and teaching must conform to certain rigid standards in order for the school to maintain its continuing accreditation.

Similarly, approval is required for the year of internship and a minimum of three years of residency training in neurology that follows internship. The members of the boards charged with these inspections usually are leading educators and specialists in their fields.

In addition to the minimum eight years of formal training, many neurologists take additional post-graduate training and fellowships in subspecialties of neurology. Check the reputations of the institutions where they trained. Poor doctors can emerge from excellent training programs, but you can get a feeling about a neurologist from basic credentials and from opinions of your family physician and other doctors whom you trust.

At the conclusion of the training period, neurologists are tested to determine their competencies in neurology. Ask whether your prospective neurologist is certified by the American Board of Neurology, is a Fellow of the Royal College of Physicians and Surgeons of Canada, or is a member of the Royal College of Physicians of Great Britain. All of these specialty boards are recognized in the United States, Canada and England.

Secondly, be concerned about your particular medical problem. Almost every neurologist who has fulfilled the basic training requirements and passed the board examinations will be competent to treat ET patients. However, in any profession, proficiency is improved by real interest and experience. It is unrealistic to expect all neurologists to be equally interested in your disorder.

In addition, some physicians are uncomfortable treating patients with conditions for which there is no known cure and which progresses despite the best possible medical care. Ask a physician directly if they are particularly interested in your condition.

It is especially important to make inquiries through organizations such as the International Essential Tremor Foundation and local support groups where possible. These organizations usually have lists of physicians who are experts in the symptomatic treatment of the particular disorder to which the organization is devoted. They have obtained their information both from patients and from their personal knowledge of many specialists and are continually updating the list.

Finally, remember that doctors are people too. Your respective personalities may be incompatible and may be of less importance if you have a transient and quickly curable condition. However, if you have a chronic disease, your association with your neurologist will be a long and intimate one.

Second Opinions: Knowing Your Rights and Options

By The Patient Advocate Foundation, reprinted with permission

Facts and circumstances relating to the individual

Making decisions about healthcare is one of the most important aspects in a person's life. Many people are told that they have cancer or another life threatening illness and feel that they must make a decision and begin treatment as soon as possible. While this may be true in some instances, taking the time to learn about your disease, getting a second opinion or perhaps even a third opinion and weighing your options is a very reasonable approach. Proactive decision making will give you a greater degree of control over your treatment. Decisions regarding your health should be made after you have been thoroughly informed about your diagnosis, prognosis and available treatment options.

Will my doctor be upset if I get a second opinion?

Most doctors will welcome another physician's input. Your doctor may be able to recommend a specialist. A good doctor understands your right to be well informed and should support a second opinion. You will want to obtain a copy of your medical records and test results, to share with the new doctor that you see for the second opinion.

Doctors' opinions may differ

A different doctor may come up with a different diagnosis, or at least offer a different opinion as to treatment choices. Not every doctor will have the same opinion with regard to diseases and possible treatments. Factors which may have an effect on a doctor's opinion are technology available to that doctor, school of thought, where they were trained, individual methods of treatment and experience in dealing with that particular diagnosis.

Treatment methods may differ

Some doctors prefer to monitor the situation and use less aggressive procedures before moving to surgical intervention. Some doctors like to use more aggressive treatment methods from the beginning. By getting a second opinion, you can expand your options about different treatment methods which may be most suitable for you. Being informed is always your best option.

Second opinions don't hurt, and in fact, may even help

It never hurts to get a second opinion. Keep in mind that doctors are human and they too can make mistakes or be faced with unusual or challenging cases. When the first doctor's opinion is the same or similar to the second doctor's, your confidence will be increased. There is nothing lost by visiting one more doctor just to make sure that the first doctor's opinion is correct. With serious illnesses that may require extended treatment, you should feel confident that you have chosen the most appropriate treatment for your particular situation. A valid opinion and appropriate course of treatment is your best option for return to good health or grasping control of the chronic disease.

While second opinions may be awkward for doctor and patient at times, studies have shown that 30 percent of patients who sought second opinions for elective surgery and 18 percent of those who were required to obtain a second opinion by their insurance company, found that the two opinions were not in agreement. These studies are one more reason why you need to make sure you are educated properly to make the best decision for your health.

Choosing a physician

There are many professional doctors in practice today. Family physicians provide comprehensive medical care with an emphasis on caring for all members of the family. A general surgeon is able to perform all types of surgical procedures aimed at treating a range of diseases and conditions, including cancer. Medical oncologists are specialists that use various medications to treat and manage patients with cancer. This may include the use of chemotherapy to kill cancer cells, painkillers to manage cancer pain, and drugs that will eliminate or reduce the side effects of cancer treatment. Radiation oncologists use therapeutic applications of radiation to manage cancer and other diseases. They determine the type of radiation that will be used, as well as the amount or dose, and the number and length of treatments.

You may have a need for all four of these types of physicians at one time. It is beneficial to you to keep files or a journal with names and dates and copies of your medical records and procedures as your journey to wellness can be confusing and the doctors will need to know exactly what care you have previously received.

Patient rights

Second opinions are a way to learn about your diagnosis and choices for treatment options. Some doctors are more conservative while others tend to be more aggressive. A patient has rights and one of your most important rights is the ability to get a second opinion about your diagnosis. Being informed is critical in deciding your choice of treatment.

Statistics show over one-third of adults in the U.S. will never seek a second opinion. And almost 1/10 of newly diagnosed patients rarely or never understand their diagnosis. A second opinion means you are consulting with another doctor to confirm a diagnosis and/or find possible different treatment choices available to you. It is recommended to get a second opinion immediately to avoid delays in your treatment. Seven states currently have health laws pertaining to second opinions.

What will it cost me?

Assuming it is medically necessary, most insurance plans will pay for at least part of the cost while Medicare will pay 80% of the cost. As a matter of fact, if the second opinion doesn't agree with the first, Medicare will pay 80% of the cost of a third opinion. Patients that belong to a Medicare Health Maintenance Organization (HMO) are entitled to a second opinion, but some plans require a referral from your primary care physician, and like most HMO treatments, you must see an in-network physician. To learn about second opinions call 1-800-MEDICARE (1-800-633-4227) or for specific rules by state for a second opinion you can visit <http://www.medicare.gov/coverage/home.asp>.

Call your insurance provider before going for any treatment or second opinion to prevent any confusion or denial of the bill. You need to know exactly what will be covered, such as an out of network provider, any lab work or testing that may be required and what your responsibilities are before seeking the second opinion. Diagnostic tests can be very costly and many insurance providers will not pay for them if they were completed for the initial diagnosis. You have the right to have copies of the tests you already had done. Be an informed consumer and arrive for the second opinion with all of your previous medical records, contact information about the first physician, insurance card, list of prescribed medications and allergies, and any diagnostic test results.

Life with Essential Tremor

Addressing the Psychological Aspects of Essential Tremor ET's Impact on Emotional and Mental Processes

By Scott A. Wylie, Ph.D.

Our understanding of the psychological effects of essential tremor (ET) is at an early stage even though research on this topic has increased dramatically over the last 10 years. Brain areas often linked with ET, including the basal ganglia, thalamus, and cerebellum, play important roles in cognitive and emotional functions. The discovery that some individuals with ET experience changes in these areas is not too surprising.

ET can affect cognitive and emotional functioning. These changes can significantly impact quality of life by leading to frustration, social withdrawal, caregiver stress, and difficulties performing and participating in usual activities. For these reasons, efforts to improve the education, detection, and treatment of cognitive and emotional changes that can accompany ET are essential.

Cognitive Changes in ET

Cognition describes the mental processes we use to perceive, recognize, understand, and navigate the world around us. These specialized processes include learning and memory, decision-making, attention, and language.

Many neurological conditions impact cognition and can result in mild to severe impairment that affects one or more of these processes. When several aspects of cognition are severely impaired and interfere with the ability to perform usual activities, a diagnosis of dementia may be made. Based on these definitions, two general questions about the effects of ET on cognition are often on the minds of patients, families, and health providers.

Does a diagnosis of ET increase risk of developing dementia? Two relatively large studies conducted in Spain and in New York reported higher frequencies of dementia in ET groups (Spain: 11.4%; NY: 25.0%) compared to control groups (Spain: 6.0%; NY: 9.2%). While these figures should heighten our concern, important questions need to be resolved.

For example, in a follow-up Spanish study, increased risk of dementia was found only in individuals who were older than 65 when they developed ET. This finding raised the question of whether the increased risk for dementia is related to ET or to some other health or neurological issue unique to older age. More studies that track cognition across the age spectrum are needed to better determine risk of dementia in ET.

Since most people with ET do not show dementia, a second question follows.

Are milder changes in cognition impacted by ET? Recent research suggests that some individuals with ET may develop difficulties in cognitive abilities typically referred to as executive functions. Executive functions refer to the most complex aspects of human thinking, such as our abilities to reason solutions to problems, plan and organize steps to complete tasks, monitor our errors or mistakes, perform mental calculations, multi-task, and prevent distractions from interrupting our train of thought. Changes in these thinking abilities can significantly disrupt how well a person performs complex tasks at work and at home.

ET may also affect the cognitive ability to retrieve memories and words quickly during conversations. Everyone has experienced a time when a word or thought won't come to mind. These difficulties occur more frequently in ET than normal. Interestingly, changes in executive functions and word retrieval abilities are common to other movement disorders, such as Parkinson's disease, suggesting that similar brain circuits may be involved in these changes.

Emotional Changes in ET

A few months ago, I interviewed a 97-year-old woman who was brought into clinic for concerns about memory and depression. When asked if she had been feeling depressed, she smiled and said, "No, I'm fine." Deciding to press a little further, I asked, "If you were feeling depressed, would you tell me?" She chuckled and without missing a beat, replied, "No, probably not!" We shared a smile, and I kindly thanked her for her honesty.

Like this woman, many people find it uncomfortable to discuss or acknowledge emotional difficulties. However, research shows that rates of depression, anxiety, and apathy are alarmingly higher in ET compared to the general population.

In a study of 55 male veterans diagnosed with ET, 74% admitted to feeling embarrassed by their tremor, and 65% acknowledged avoiding social situations because of it. In a study of 349 ET patients seen in an outpatient clinic, 58% answered yes to the question "Does your tremor often embarrass you?" It remains unclear if all of the emotional changes are solely a reaction to ET or also related to neurological changes. Either way, these reactions and emotional challenges are very real and, if ignored or untreated, potentially harmful to one's health.

Helpful Strategies

What is the best strategy for managing cognitive and emotional changes? Discussing cognitive and emotional changes with your neurologist is the best place to start. If you have concerns about cognitive or emotional functions, a neuropsychological evaluation can be useful for precise characterization of these changes and tracking them over time. If problem areas are identified, there may be medications or behavioral strategies to improve or compensate for these changes.

Many individuals and families find that a series of counseling sessions with a psychologist can be very helpful to address issues of frustration, embarrassment, and overall coping with cognitive and emotional changes.

Finally, as with everyone, it is important to maximize the benefits to cognitive and emotional health by consistently engaging in mental, physical, and social activities.

Psychosocial Aspects of Tremor

By Glenn T. Stebbins, Ph.D.

Tremor has many effects on physical and psychosocial functioning. Difficulties in conducting simple daily activities such as eating and drinking may cause one to change habits and routines. The effects of these changes may signify nothing more than small nuisances or major inconveniences. Indeed, the effects of tremor may be so great as to lead to major psychological difficulties.

Small nuisances caused by tremor can often be addressed by making adjustments to how one functions. For example, difficulties in maintaining fine motor control while attempting to button clothes or tie shoelaces can often be overcome by using other methods to accomplish the same goals. The use of fasteners such as Velcro™ reduces the need for exact motor control for those tasks, or using a travel mug with top and built-in straw may help control spillage when drinking.

However, these adjustments may not help, or indeed, may increase the sense of embarrassment one feels around others. This social embarrassment may be one of the major impairments in tremor disorders. What may start out as a small increase in self-consciousness, can develop into a phobia or unrealistic fear of social situations and can affect all social activities. These fears may become so great that they limit one's desire to go out of the house or visit with friends.

When social embarrassment becomes great, one may become virtually housebound, unable to leave the house without feelings of panic developing. This situation must be differentiated from simple social phobias in which only some social situations lead to this unrealistic fear. Agoraphobia is when leaving the house becomes intolerable or it is a fear of being in places where help is not available. Agoraphobia may or may not be associated with panic attacks that involve feelings of dread or fears of dying, and physical symptoms such as shortness of breath, dizziness and/or heart palpitations.

There are ways to deal with the simple social fears that may develop from tremor. Just as you use different techniques to help you with the physical difficulties, you can use different techniques to help with the psychosocial problems. First, learn to relax. This may sound too simple, but it is true; tremor increases when nervous or under stress. By learning to relax, you can control that aspect or worsening of the tremor. Whether you make use of a program such as biofeedback, or suggestions from other tremor patients, learning to relax emotionally will help. This does not mean that the tremor will be eliminated, but it does mean that it may not be as severe as when you are not relaxed.

While you are learning, plan your social interactions so as to limit the nervousness you may experience. Try to socialize with friends who know about your tremor and who do not make a big deal about the movements. When you are meeting new people, socially or in business, you may wish to explain quietly and simply that you have ET. Finally, if the social fears do interfere with your interactions, talk with your prescribing physician. There are specific behavioral treatments that can address these problems directly and that are very successful in decreasing them. Your physician can be helpful personally or can refer you to a neuropsychologist with whom he works.

Emotional Aspects of ET

By George W. Paulson, M.D.

Few people have written about whether or not ET is actually “benign.” As you know, ET has carried that adjective for years, but is a tremor truly benign when one ceases to go to restaurants because of embarrassment? Is it benign when you have to explain to colleagues at an exciting board meeting why suddenly your hands are shaking? Is it benign when a proud young man is considered “nervous” because his hand or voice quivers? For some it would not even be considered benign when you have to avoid long earrings, can’t wear feathers in your hat, and dare not write in public. It can be distressing to have to write checks before going to the bank or store, simply to avoid embarrassment as others watch.

Most movement disorders are readily apparent to even the most casual observer. In addition, many patients feel a sense of trembling inside, particularly when the tremor is due to parkinsonism rather than ET, even though the tremor is not apparent to others. ET can affect the head, hands or voice; and the effects can be beyond simple self-consciousness or social embarrassment.

I remember the captain of the football team at the high school, who has now become a successful physician. He was failing draftsmanship in his senior year. When his mother said he was too “nervous,” his eyes flashed with anger as he said, “I told you Mother, I’m not nervous; it’s just that my hands shake.” When her own fingers were extended she could see tremor, but she never considered that to be linked to his “handicap,” or his poor performance in class.

ET is an action tremor, and therefore the physical disability from ET can involve handwriting or drawing. What of the cartoonist or the surgeon who really depends on his hands for delicate work? Surely to fail in your vocation because of ET is more than just embarrassing. There are settings in which public eating is mandatory. Some must avoid soups or must hold a coffee cup with both hands, and food spills sloppily from a fork. Since our ears are much attuned to pick up anxiety, anger or dishonesty in a voice, a quivering voice can be interpreted as anxiety.

For all these reasons ET patients may tend to view themselves as more disabled than is true for those with other conditions, perhaps because the problem rarely disappears. It may be difficult for some to overcome this sense of disability. Most neurologists would prefer individuals to have ET than to have PD, simply because ET is not associated with slowing down and does not shorten life or affect cognition.

In our culture, appearance and conformity are considered necessary. Deviation from the “norm” can result in unwanted attention, and the attention can make individuals feel anxious. Since tension and stress exacerbate ET, there may actually be more tremors in the presence of others. Although patients with other movement disorders, even very severe ones such as Huntington’s disease, develop techniques to disguise the movements, it is not easy for ET patients to disguise their shaking. For some, the harder they try the worse it becomes.

Can physical therapy help with the physical limitations of ET? Most of us would say “probably not,” but the optimism of therapists and their universal commitment to maximize potentials can be helpful. The patient’s ability, by force of will, to transiently overcome shaking or to calm the anxiety that increases the shaking can lead to self-improvement.

Emotional therapy (from a neuropsychologist) may be valuable for some because they learn to adjust to the tremor. Support groups can also help. They can offer opportunities to discuss common experiences and to learn tricks one can use to avoid being conspicuous. One of their best features is a reconfirmation that ET is indeed a real disorder, a disorder that has treatment, even though there is no ideal treatment.

One thing that the support groups, physicians, friends and indeed the patients themselves can do is to absolutely continue to remain active in society, to educate and, through self-help, to help others.

ET, Depression, and Anxiety

Tremor Talk

ET can have a devastating impact on the quality of life for individuals with moderate to severe tremor. Depression and anxiety—in particular, social anxiety—are common companions for many of these people.

Depression can generally be well-controlled with medication, supplemented in some cases with counseling. It can affect the way an individual eats, sleeps, feels and thinks. Often under-recognized and under-treated in persons with ET, it is important to know the symptoms of depression:

- Persistent sadness or anxiety
- Excessive crying or tearfulness
- Feeling hopeless, pessimistic, worthless, helpless or irritable
- Loss of interest or pleasure in activities that were once enjoyed
- Fatigue
- Difficulty concentrating, remembering, or making decisions
- Difficulty falling asleep, early-morning awakening, or oversleeping
- Loss or gain of appetite and weight
- Thoughts of death or suicide
- Persistent physical symptoms that do not respond to treatment, such as headaches and digestive disorders

If you have any of these symptoms, and suspect you might have depression, contact your doctor.

Anxiety disorders cause people to feel frightened, distressed and uneasy. Left untreated, ongoing anxiety can reduce productivity and diminish quality of life. Each year, more than 40 million Americans are affected by anxiety disorders. Many persons with ET experience social anxiety, a form of anxiety experienced as shyness or a fear of public places, performance, public speaking, social situations, interactions with others, and being evaluated or scrutinized by other people. The term social anxiety also is used to describe anxiety brought by anticipating embarrassment or shame.

Persons with social anxiety experience the physical symptoms of anxiety, including confusion, pounding heart, sweating, shaking, blushing, muscle tension, upset stomach, and diarrhea. For persons with ET, tremor often worsens. Social anxiety, according to WebMD, may be caused by an imbalance of the neurotransmitter serotonin. Neurotransmitters are chemical messengers moving information from nerve cell to nerve cell in the brain. If they are out of balance, the way the brain reacts to stressful situations is altered, possibly leading to anxiety. It is believed that social anxiety runs in families.

The most effective treatment of social anxiety is cognitive-behavior therapy (CBT). Several medications may also help ease the symptoms of social anxiety, but persons with ET need to ensure that their physician knows that some of these, including Selective Serotonin Reuptake Inhibitors (SSRIs) antidepressants like Paxil, can make tremor worse. Counseling to improve self-esteem and social skills, as well as relaxation techniques may also help. For more information about depression and social anxiety, visit the Mental Health America website at www.mentalhealthamerica.net to find a healthcare provider near you.

Tremor and Stress

By Mark Hallett, M.D.

Stress is a given of modern life. People work hard, problems need to be solved, and the future is a concern. Anxiety is a common emotion with its unpleasant sense of impending misfortune. People differ in their behaviors and reactions to life's problems.

Stress taxes the body, and too much leads to heart attacks, ulcers, high blood pressure, and emotional problems. Symptoms include irritability, nervousness, worry, jumpiness, fatigue, difficulty relaxing or sleeping, headaches, difficulty concentrating, and tremor. Stress also weakens the body's defenses against infection.

Although the tremor of excessive stress is typically of low amplitude and rapid rate, it can be prominent. If a person has a tremor disorder, such as ET or Parkinson's disease, the tremor will worsen. Indeed, all neurological symptoms will worsen with stress, but tremor is worsened by excessive stress; stress will not cause a permanent tremor.

Stress is associated with a hormonal response in the body characterized chiefly by an excessive outpouring of epinephrine and norepinephrine. These chemicals are responsible for many of the symptoms, including tremor. Increased tremor occurs in part due to receptors of adrenergic substances in the muscles being activated and increasing tremor.

A number of persons turn to alcohol for relief. This can have brief benefit, and alcohol will also dampen tremor. It is not a good long-term solution because chronic, excessive alcohol ingestion leads to its own problems.

It is possible to treat some symptoms of stress including tremor. Tremor can be relieved with beta-blocking drugs such as propranolol (Inderal ®) because these agents counteract the adrenergic substances. These agents may not relieve the anxiety and so will not reduce the stress.

Too much stress is bad for us all! People must adjust their lifestyles to avoid excessive stress. It is important to find time to relax. Regular exercise is healthful for its own sake, but it can also play a role in reducing stress.

Stress and ET

By Arif Dahvi, M.D, M.B.A.

A stressful situation such as writing a check while in line at a grocery store or using a pointer during a presentation can worsen essential tremor (ET). Observers may believe the tremor is due to nervousness, but in reality the increased tremor is caused by the additional mental and physical stress in such situations. What many people, including people with ET, are not aware of is that good stress, such as watching favorite teams win at sports, can also increase tremor.

Hans Selye, a pioneer in studying the mechanisms of stress, understood stress to include a wide range of strong external factors, both physical and mental, that can cause a physical response called general adaptation syndrome (GAS). The three stages of GAS, as defined by Selye, are alarm reaction, adaption, and exhaustion.

During an alarm reaction, adrenaline is released to create the fight-or-flight response. Muscles tense, the heart beats faster, breathing becomes deeper, pupils dilate, and perspiration increases. If the stress is removed, the body returns to normal. If the stress continues, the body moves into adaption stage.

Adaption is the body's response to long-term stress. Remaining in the adaption stage too long can cause fatigue, concentration lapses, and irritability. If the stress is not relieved, it leads to exhaustion, the third stage. The body's defenses begin to deplete in the exhaustion stage. Depression can also occur in this final stage of GAS.

A part of the brain called the limbic system, which processes emotion and memory, is greatly involved in this stress reaction. With any situation (stressor), the limbic system searches its memory for a similar situation from the past. Based on that memory, the limbic system determines how much of which neurotransmitter to release. Far from being something in the mind, stress causes a real physical and chemical change in the body. This can help explain why someone with ET in the checkout line at a grocery store can experience severe tremor.

The limbic system processes the emotional memory of having had difficulty signing one's name in the past and causes the initial alarm reaction of GAS. The associated emotional response of embarrassment at being unable to smoothly perform a relatively simple task increases the tremor. The guilt of holding people up in line also feeds the stress reaction. This causes the limbic system to release adrenaline, which increases tremor.

Understanding how stress affects the body – and the severity of tremor – and finding ways to cope with stress can have a significant effect on the severity and management of ET.

Functional Disability with ET

By William C. Koller, M.D., Ph.D.

If one looks in textbooks of medicine or neurology, one sees ET described by the prefix “benign.” ET is not life threatening and does not shorten life expectancy. However, ET is certainly not “benign” for all individuals. It is true that some people have only a minor tremor that does not interfere with many daily living activities. However, many people have severe tremor that interferes with a number of daily activities. This tremor should certainly be designated “not so benign.”

Many disabling problems in medicine do not shorten life expectancy or cause death. Even a stroke which leaves one side of the body paralyzed does not necessarily result in death or shortened life expectancy. Similarly in many other conditions, life is not threatened, but the quality of life certainly is.

Medical specialists have been increasingly interested in quality of life and how disease processes interfere with this. In one study, the most common complaint of ET patients was difficulty with handwriting, followed by difficulty drinking liquids, and by difficulty with fine manipulations and difficulty eating. Embarrassment also occurred in the majority of patients, and some patients had difficulty with dressing and speaking.

Because of the inability to use their hands for fine manipulations, patients such as dentists, surgeons and draftsmen could no longer pursue their careers. The patients expressed much frustration at their inability to hold a newspaper for reading or to do simple chores with a screwdriver around the house. While it appears that head tremor does not cause much functional disability, it can be a major cause of embarrassment. Hand tremor, too, can cause much embarrassment, and some patients tend to become reclusive, avoiding public places such as restaurants.

A national study on the impact of ET on daily activities was conducted. The “Sickness Impact Profile” is a standardized questionnaire that is used to determine the effect of a given disease on patients’ abilities to carry out normal activities and to interact socially. In this study, the questionnaire was also distributed to a number of volunteers from the general population and to a group of Parkinson’s disease patients so that comparisons could be made among the patients with ET, those with PD, and those with no neurologic disorder.

When compared with the healthy control group, ET patients had significantly greater dysfunction in all categories except eating. Those categories in which patients considered themselves to be most impaired were communication, work, emotional behavior, home management and recreation.

Replies from parkinsonian patients indicated that they experience greater dysfunction than do ET patients. We concluded that ET could cause significant disability. When compared with PD, ET tends to be less severe, yet causes greater psychosocial dysfunction. Stress associated with social situations often exacerbates tremor severity. In one study, 15 percent of patients with ET were forced into early retirement. Therefore, it is clear that ET can in some patients cause serious physical, social and psychological disability. Fortunately, drug treatment often decreases the tremor and can markedly reduce functional disability.

ET and Aging

By Rodger J. Elble, M.D., Ph.D.

“We shake more as we grow older.” This widely held opinion reflects the tendency of most lay people and many physicians to dismiss mild tremor as “normal aging.” Tremor in the hands, stooped posture, slow walking, and failing memory constitute a common caricature of aging, but in most instances, these problems, alone or in combination, are due to a neurological disease. Each of these problems exists in at least 5% of people older than 65 years, the age group commonly regarded as the elderly.

Mild tremor in the hands is seen in as many as 20% of healthy people older than 65 years. In most cases, elderly people shake because normal physiologic tremor is exacerbated by medications, systemic illness, or psychosocial stress (e.g., anxiety, depression). Hormonal disturbances, heart failure, pulmonary disease, renal failure and liver disease also make people more tremulous. Many medications exacerbate physiologic tremor.

Generalized shakiness “from head to toe” of duration less than 6-12 months is usually caused by medications (e.g., antidepressants, narcotics, asthma medications) or by occult systemic or neurologic disease. Tremor of this type should never be dismissed as essential tremor (ET).

ET is longstanding (> 3-5 years) action tremor in the hands and commonly in the head and voice, for which there is no discernible etiology. ET is not a specific disease. Many elderly people with ET have had tremor for decades, and these people commonly have a family history of tremor. Some of these patients have dystonia or a family history of dystonia, and it is debatable whether such tremor should be classified as ET or as dystonic tremor. Patients with ET unfortunately are not immune to other neurological disorders. For example, a patient with longstanding ET may develop Parkinson disease. ET generally progresses slowly over decades. A rapid change is usually due to an undiagnosed comorbidity.

The development of tremor after age 65 should not be dismissed as ET unless the patient is followed for several years and develops no other neurological abnormalities. In other words, late onset tremor should be viewed as a sign of undiagnosed systemic or neurologic disease until proven otherwise. Many neurologic diseases can begin with nonspecific tremulousness in the hands, including Parkinson disease, dystonia and Alzheimer disease. Dystonia should be suspected when late-onset tremor begins in the head or voice.

As presently defined, ET is a clinical syndrome, not a specific disease. Tremor-producing conditions are increasingly common with advancing age, and ET is a common misdiagnosis. Correct diagnosis is often difficult and usually requires a careful examination and follow-up by an experienced physician.

ET and Pregnancy

By Kathleen M. Shannon, M.D.

Concerns about pregnancy and ET can be divided into four broad categories: 1) genetic issues; 2) the effect of pregnancy on ET; 3) the effect of ET on pregnancy; and 4) the effects of medications on pregnancy and fetal development.

Genetic Issues

As many as two-thirds of ET patients have a positive family history of the disorder. We believe that ET is an inherited disorder which follows an autosomal dominant transmission with incomplete penetrance. Autosomal dominance means that it is only necessary to inherit one abnormal gene to manifest the disorder. This gene may be inherited from either parent. If one parent has the abnormal gene, each child has a 50 percent chance of inheriting an abnormal gene. Incomplete penetrance means that not all persons who inherit the abnormal gene will develop the disorder, although such a person is still able to pass the gene on to the next generation. This explains how a child with two healthy parents can develop ET: the phenomenon of “skipping” one or more generations.

In some hereditary disorders, testing can be done during pregnancy to determine whether the developing fetus has inherited the gene which produces the disorder. Since the gene(s) has not yet been identified, no such test exists yet for ET.

The Effect of Pregnancy on ET

The remarkable changes in hormone concentration, body weight and the body’s handling of drugs that occur during pregnancy could conceivably have potent effects on the severity of ET symptoms. Unfortunately, there has been no research in this area, so no information is available.

The Effect of ET on Pregnancy

There is no evidence that ET makes it more difficult to become pregnant, carry a pregnancy to term, or have normal labor and delivery. Although there is a lack of research in this area, we have no reason to believe that ET will interfere with the normal conduct of pregnancy.

Women who anticipate breastfeeding should be aware that certain medications, such as primidone and diazepam and related compounds, could enter the breast milk and cause sedation in the infant. Therefore, the breastfeeding mother should not use these medications.

Of course, it is important to discuss any concerns you might have about pregnancy with both your neurologist and obstetrician/gynecologist well in advance of becoming pregnant.

Table 1

DRUG	EFFECT
Ethanol, Mysoline ® (primidone)	Malformations, drug withdrawal in infant, bleeding disorder in infant
Inderal ® (propranolol), Blocadren® (timolol), etc.	Unknown
Catapres® (clonidine)	Unknown
Desyrel® (trazodone)	Unknown
Clozaril® (clozapine)	Unknown
Botox® (botulinum toxin injections)	Unknown
Valium ® (diazepam), Klonopin® (clonazepam), Xanax® (alprazolam), etc.	Malformations, drug withdrawal in infant, bleeding, disorder in infant

The Effect of Medications on Pregnancy and Fetal Development

Medications taken during pregnancy can adversely affect fetal growth and development. However, it is usually not possible to know the exact risk of a medication because they are not tested in pregnant women; it is also not possible to make conclusions from isolated cases in which normal or abnormal babies are born to women taking medications.

It is known that the risk of having a baby with a birth defect is approximately 2.5%. Taking medication during pregnancy increases this risk, but it is important to note that among women who take medications for chronic illnesses during pregnancy, the likelihood of having a normal infant is greater than 90 percent. The risk of malformation is highest when medications are taken during the first three months of pregnancy, the time of major organ development. Table 1 lists drugs reported to be beneficial in the symptomatic treatment of ET, and the risk of fetal malformation, if known.

The soundest recommendation to women with ET who are contemplating pregnancy is that they taper and discontinue medications before they become pregnant, and do not take any medications during the first three months of their pregnancies. If this is not possible, they should stay on the lowest possible dosages of the fewest medications possible.

Treatment Options

Medications Commonly Prescribed for ET

Reviewed by Rodger J. Elble M.D., Ph.D.

There is no cure for ET, and no medication is specifically designed to treat ET. However, there are several medications that may be of benefit - singularly or in combination - in the treatment of ET.

Considerations when taking medication:

- Always follow your physician's directions when taking any medication.
- Become aware of the side effects of the medications you take. Your physician and pharmacist can help you.
- Contact your physician if you experience harmful side effects.
- Inform your physician and pharmacist of all medications you are taking, including over-the-counter medications. Although not always possible, use one pharmacy for all of your prescriptions.
- Never take anyone else's medication, and do not share your medication.
- Although one medication may work well for one person, it may not work for others. Finding the correct medication, or combination of medications, is a trial-and-error process that will take time and patience.

Medications commonly prescribed for ET:

Propranolol (Inderal®) - Propranolol is the only medication approved by the Food and Drug Administration (FDA) for the treatment of limb and head ET and is available in immediate and long-acting formulations. Propranolol is a beta blocker that is used primarily for treating high blood pressure. Side effects of propranolol are usually mild and are more frequent at higher doses. The main side effects are decreased heart rate and blood pressure. Talk with your doctor if you have heart failure, diabetes, or asthma. Patients who respond usually require a total daily dosage of 240 mg or less. Other beta-blockers such as atenolol, metoprolol, and nadolol can also be beneficial for treating ET.

Primidone (Mysoline®) - Primidone is an anticonvulsant medicine that is effective in treating ET. Although it might have initial side effects such as nausea, poor balance, dizziness, fatigue, drowsiness, and flu-like symptoms, there are few long-term problems. To reduce the possibility of side effects, start with a small dose at bedtime (25 mg or one-half of a 50 mg tablet) and gradually increase the dosage according to your doctor's instructions. Patients who respond usually require a total daily dosage of 300 mg or less.

Clonazepam (Klonopin®), diazepam (Valium®), lorazepam (Ativan®), and alprazolam (Xanax®) - These anti-anxiety medications may be useful in patients who do not respond to other medications or who have associated anxiety. The main side effect is drowsiness, and these drugs can also affect mood and balance. These drugs are usually less effective than propranolol and primidone, and they can be addictive. There is also risk of withdrawal symptoms if they are stopped suddenly.

Other medications worth trying:

Topiramate (Topamax®) - Topiramate is an anticonvulsant that has been shown to be modestly effective in controlling tremor. Side effects include numbness or tingling, memory loss, and weight loss. Topiramate should be avoided by patients with a history of kidney stones and narrow angle glaucoma.

Gabapentin (Neurontin®) - Gabapentin is a generally well tolerated anticonvulsant, but controlled studies have produced conflicting results regarding its efficacy in ET. At best, this drug has a very modest benefit and seems to work best when it is taken alone. It is usually tried when there is no response to other medications. Side effects include fatigue, drowsiness, impaired balance, and nausea, especially when beginning therapy.

Miscellaneous agents:

Mirtazapine (Remeron®) - Mirtazapine is an antidepressant. Due to its lack of effectiveness for the majority of patients and its significant side effects, mirtazapine is not recommended for the routine treatment of ET. Adverse effects include confusion, dry mouth, weight gain, frequent urination, balance and gait difficulty, nausea, and blurred vision.

Botulinum Toxin Injections (BOTOX®, Myobloc®) - If other medications fail, you may consider injections of botulinum toxin into affected muscles. Botulinum toxin injections have been useful in the treatment of some patients with head and voice tremor and sometimes hand tremor. The toxin must be placed into target muscles by a trained specialist, and repeat injections are needed every 3-4 months. Transient weakness of the injected muscle is a potential side effect. This treatment can be expensive, so be sure to check with your insurance provider about coverage.

Alcohol - Adults with ET often notice that responsible drinking of alcohol - having one or two drinks before dinner or at social events - reduces tremor for one to two hours. However, a more severe rebound tremor can occur after the effects of alcohol have worn off. The addicting and intoxicating effects of alcohol limit its use.

Topiramate and ET

By Jean P. Hubble, M.D.

There is no one single medication uniformly safe and effective for the treatment of ET. The most frequently prescribed medications for this disorder are propranolol (Inderal®) and primidone (Mysoline®). Not all patients have adequate response to these medications, and some individuals can simply not tolerate these drugs because of side effects. There are numerous other medications that are less often used with success in essential tremor. The need for a safe and effective remedy for this condition is obvious.

Topiramate (Topamax®) is a prescription drug approved by the United States Food and Drug Administration (FDA) for the treatment of seizures. More specifically, topiramate is indicated as adjunctive (add-on) therapy for the treatment of partial-onset seizures and primary generalized tonic-clonic seizures. A limited study of the drug topiramate in ET patients has suggested that this drug may be useful in reducing tremor severity. It is important to note that seizures and tremor are not related conditions. Nevertheless, anti-seizure medications have often been tried as a treatment of tremor because of the effects of these drugs on the central nervous system.

For example, primidone (Mysoline®) has been used for many years in the treatment of ET even though the drug is actually FDA approved as an anti-seizure medication. This type of “off label” use of a medication is not unique to ET, but many medications can be safely and effectively used to treat disorders other than those for which they were initially intended. In the small tremor study cited above, Dr. Connor studied the effect of topiramate in 11 patients with ET. These individuals took topiramate at a dose of 75-400 mg per day. They were followed for 2-21 months. These patients reported improvement in hand, head and/or voice tremor ranging 25-80%. The only side effects noted were numbness, tingling and weight loss. It is important to note that this was an open label study. This means that patients and their doctors knew that they were on the active compound topiramate, and there was no comparison to placebo. Earlier studies of ET have demonstrated that many individuals will notice a mild benefit for a short time when given placebo.

While side effects were few in this limited study, relatively common side effects of topiramate in the study of seizure patients have revealed the following: sleepiness 29%, dizziness 25%, ataxia or decreased balance 16%, slurred speech or other difficulty speaking 13%, slowness in thinking or moving 13%, blurred vision or other visual problems 13%, difficulty with memory 12%, numbness or tingling 11% and double vision 10%. Weight loss occurred in 9% of adults with seizures treated with topiramate. Kidney stones occurred in 1.5% of adults receiving topiramate. Increase in fluid intake (improved hydration) may lessen the likelihood of the occurrence of kidney stones with a medication such as topiramate.

Adverse Drug Reactions

By William C. Koller, M.D., Ph.D.

The two classifications of drugs with proven efficacy in the treatment of ET include propranolol and primidone. These drugs, like all drugs, have the potential for adverse reactions. However, both propranolol and primidone are usually well tolerated.

Contraindications to propranolol use include: 1) heart failure, especially poorly controlled; 2) various forms of heart block; 3) asthma or other bronchospastic disease and; 4) insulin-dependent diabetes since the drug may block clinical manifestations of low blood sugar. The side effects of propranolol are usually mild and transient. The pulse rate and blood pressure can be lowered in some patients and certainly pulse rate has to be followed. Other less common side effects include fatigue, weight gain, nausea, diarrhea, rash, impotence and alterations in the mental state, particularly depression. It is important for patients to realize that certain side effects such as fatigue or impotence may be due to drug therapy. Patients in whom such side effects arise should discuss them with their physicians since dosages and drugs can often be changed. However, occasionally choices have to be made, and it is best to make such choices fully informed.

Most side effects of primidone occur when a patient first begins taking the drug, and as many as 30% to 40% of people will have short-term side effects following the first dose of the drug. These can include dizziness, headache, flu-like symptoms, nausea, vomiting and poor balance. Patients should be warned of these possible effects; the fact that they will go away with continued drug use should be underlined. The long-term side effects of primidone are really quite minimal. Some people get somewhat sedated; however, if the dosage is kept low this is not a major side effect. In general, most patients tolerate propranolol, a beta-adrenergic blocker, and primidone quite well. Any change in symptoms or the emergence of change in a patient's normal condition should prompt inquiries to the prescribing physician.

It sometimes happens that patients don't tell physicians about side effects, and simply stop taking the offending drug. This may be particularly true concerning sexual dysfunction. Patients may be embarrassed about sexual dysfunction or think it is just related to the aging process and not share this information with their physicians.

The main goal of treatment is to reverse functional disability. For tremor, this may include difficulties with handwriting, drinking liquids or with fine manipulation. If the drug treatment causes significant disability such as fatigue or impotence, these may be worse than the disease itself and cause the patient to decide the treatment isn't worth taking. However, the patient has to share this information with his doctor and ask: "Is it related to the disease; is it related to the treatment of the disease; is there anything we can do about this?" For instance, if impotence is not related to tremor or to the drugs being used to treat tremor, it may be due to depression or a urologic dysfunction that could be treated. As primidone may cause impotence, many physicians will slowly decrease dosage and see if there is a change. If so, another drug can be tried to treat the patient's tremor.

It is important that patients become their own advocates, be aware of changes they are experiencing, and share this information with their physicians. They may have to challenge their physicians and ask for explanations of the possible causes of such dysfunctions. Only with such cooperation will the patients obtain the best care and achieve the best results from medications.

Effects of Other Illnesses and Drugs on ET

By Anthony E. Lang, M.D., F.R.C.P.(C)

A number of drugs and illnesses are capable of causing tremor in their own right or worsening tremor in predisposed individuals. All of us have fine tremor, known as physiologic tremor that can be documented with sensitive recording devices. There are many factors capable of accentuating physiologic tremor to the point that it is evident to the individual and sometimes even mildly disabling. These same factors are capable of accentuating pre-existing essential tremor (ET) or bringing it out in someone who is previously asymptomatic but predisposed to developing it eventually (for example, someone who has inherited the gene(s) causing tremor).

Physicians specializing in treating tremor patients are commonly faced with the question of whether a drug or illness is the cause of the tremor or has simply accentuated a pre-existing tremor or precipitated it prematurely in someone who was predisposed. Often the only way of knowing is to withdraw the drug (if possible) or treat the underlying condition, and then wait to see if the tremor resolves. In the comments below I will give a few examples. Because it is impossible to comment on all drug treatments, when patients believe that specific drugs may be worsening their symptoms of ET, they should discuss these concerns in detail with their physicians.

Possibly the most common “drug” that we are all exposed to is caffeine. Typically, excessive caffeine intake temporarily worsens the symptoms of ET (as it does with physiologic tremor in all of us). However, an individual’s sensitivity to caffeine seems to be rather idiosyncratic. Some patients experience a clear reduction in tremor if they eliminate caffeine from their diets, and others note no change whatsoever.

Alcohol commonly lessens the symptoms of ET; however, many patients note “rebound” increases tremor the next day even when the amounts of alcohol they drank were not excessive.

Many “over-the-counter” drugs (including herbal or “natural” medicines) can have variable effects on preexisting tremor. Cold medications that contain epinephrine-like agents may accentuate tremor, while those with antihistamines may reduce tremor in a nonspecific fashion due to their sedating effects.

This essay was encouraged by a member’s letter questioning the influence of thyroid replacement therapy on tremor. Excessive thyroid secretion (hyperthyroidism) characteristically is associated with an accentuated physiologic tremor. In the same fashion, hypothyroid patients taking thyroid replacement drugs in higher dosages than necessary to normalize their thyroid states can present in a fashion identical to hyperthyroidism.

If an individual has pre-existing ET, the tremor will clearly worsen in a hyperthyroid state (spontaneously or due to excessive intake of thyroid replacement drugs). It is also possible that an individual who has inherited a genetic predisposition to ET but who was not previously symptomatic would be more prone to developing a more severe tremor at these times than might be seen in another individual who lacks such a genetic predisposition.

Sometimes hypothyroidism is present for months or even years before a diagnosis. If this occurs in ET patients (or genetically-predisposed individuals), then it is possible that the use of normal

replacement dosages of thyroid hormone will increase their tremors quite noticeably over those experienced while they were hypothyroid. Unfortunately, in this case, it is necessary to maintain proper thyroid replacement levels (making sure that they are not excessive) and accept the increase in tremor (if necessary, treating it with anti-tremor drugs) rather than not treating the hypothyroid state appropriately.

Treatments used for many other medical illnesses are also capable of worsening pre-existing ET. Another important example of this is the potential for a number of anti-asthma drugs (both oral medications and inhalers) to worsen tremor.

Unlike the thyroid situation described above, there are numerous alternatives that can be tried in hopes of avoiding drugs that are particularly prone to increasing tremor. Prednisone and other corticosteroid drugs are used in treating numerous illnesses. There are many reasons to maintain prednisone in as low a dosage as possible or to use other “steroid-sparing” agents.

A number of drugs used for treating psychiatric illnesses can accentuate or alter preexisting ET. Many of the older anti-depressants can increase tremor, but here again, there are a large number of alternatives that could be considered if increased ET becomes a problem.

Lithium is a drug that is used in manic depressive illness. Lithium commonly causes a tremor in its own right and usually worsens preexisting tremor. Once again, alternatives could be considered, but I have seen several patients whose psychiatric well-being was very dependent on lithium, and we have had no alternative but to accept the increases in tremor and treat this as necessary.

Older and to a lesser extent, newer (“atypical”) anti-psychotic agents (as used in schizophrenia) may cause a parkinsonian tremor in anyone. Occasionally we see a striking and severe whole-body tremor when a patient with pre-existing ET is given these drugs. This is uncommon but should be considered when patients are taking anti-psychotic agents.

Of the neurologic drug treatments that accentuate ET, certain anti-convulsants are probably the most common. The best known example is valproic acid. Again, alternatives are available, but sometimes this remains the most effective drug for the patient. Occasionally, switching to a slow-release or coated form of the drug reduces the tremor at least temporarily.

Just as drug treatment for underlying illnesses can sometimes accentuate ET symptoms, occasionally a reduction in tremor is seen. This serendipitous observation has often led to the use of these drugs in the management of ET. The two best examples of this are propranolol (used for cardiac disease and hypertension) and primidone (used for seizures). Similar “accidental” responses will almost certainly continue to be the source of future drug trials in ET patients, at least until we have a better understanding of the underlying neurochemical and physiologic abnormalities in the condition.

In concluding, I would again emphasize the importance of discussing concerns about the effects of underlying illnesses and their drug treatments on ET tremor with your physician. Even if you have experienced a clear increase in your tremor symptoms, this may be temporary, and the need for the causative drug may outweigh any increased disability you are experiencing from the tremor. Patients are advised to always consult their physician and not unilaterally discontinue treatments that they feel may be aggravating their tremors.

Alcohol, Alcoholism and ET

By William C. Koller, M.D., Ph.D.

Patients with ET often notice that ingesting small amounts of alcohol will substantially reduce tremor for a short period of time - 30 to 35 minutes. MacDonald Critchley, an eminent British neurologist, wrote in 1949 “a heavy dose of spirits will temporarily check the tremor.” It is now known that very small amounts of alcohol, a can of beer or a glass of wine, can dramatically decrease tremor, but the tremor may worsen the next day, however, if one overindulges with alcohol. In fact, tremor is a major sign of the alcohol withdrawal syndrome (the delirium tremens, DTs).

In a controlled study, we investigated the intravenous (IV) administration of ethyl alcohol and measured tremor by means of an accelerometer (a small device put on the finger and connected to a computer to quantify tremor amplitude and frequency). Tremor was reduced in all 15 ET patients studied by an average of 70%. Blood levels of alcohol were minimal, and there was neither sedation nor other adverse effect from the alcohol. A similar infusion of alcohol did not decrease tremor in parkinsonian patients. However, there are patients with ET whose tremors do not respond to alcohol. How alcohol works to decrease ET is unknown. It appears that the action of alcohol is on the central nervous system (the brain) rather than on the muscles. A better understanding of alcohol's effect on tremor could lead to the development of new and more efficacious drugs for the treatment of ET.

Alcohol does appear to be a most effective drug for ET. Its use is recommended before meals (perhaps patients should enjoy a glass of wine before dinner) in order to reduce the tremor thus making mealtime more comfortable. Because alcohol does diminish ET, there has been concern that ET patients may become chronic alcoholics, “that alcohol use in ET appears only too often to lead to habits of intemperance.” However, chronic alcoholism and ET are both very common medical conditions, and it is not surprising that the two disorders would occur in the same individual by chance alone.

We found in a study of 100 patients that individuals with ET did not use alcohol any more than did parkinsonian patients or people without neurologic disease. Likewise, similar surveys in Finland and Sweden found that ET patients neither used more alcohol nor had a higher rate of alcoholism than did the general population. It is interesting to note that a postural tremor may occur in chronic alcoholics, a tremor that may persist even after a year of abstinence.

It can be concluded that the occasional use of alcohol in ET patients is desirable and that the risk of alcoholism is quite low. Thus, judicious use of alcohol appears to be reasonable.

Marijuana and Tremor

By Monique Giroux, M.D.

The acceptance of medical marijuana is growing and so is interest in marijuana for tremor and other brain conditions. To date, 20 states have legalized medical marijuana and 13 more have pending legislation. Marijuana is known for its beneficial effects on pain, chemotherapy-related anorexia and nausea, anxiety and muscle spasms. The role of marijuana in brain disease is just beginning to be explored.

The growing interest in marijuana for tremor is not surprising since traditional medical therapy often falls short of the desired effect. In addition, tremor can change from moment to moment in response to many personal factors such as the tasks or activities a person is performing, emotional unrest, anxiety or stress. In other words, there is a complex interaction between symptom severity and psychological or physical wellbeing and a person's activity. Marijuana can potentially impact one or more of these factors.

As with any treatment, the following questions must be examined when exploring the role of marijuana as a treatment for tremor:

- Are there direct and favorable biochemical or physiological effects of marijuana on brain function that can improve tremor?
- Are observed effects related to other factors such as change in emotional wellbeing or stress?
- Are there side effects both immediate and long-term that must be taken in consideration?
- What is the role of other associated factors such as cost, change in activity or habits, and stigma that may result from treatment?

Scientific Background

Marijuana refers to the dried leaves, flowers, stems, and seeds from the hemp plant *Cannabis sativa*. This contains the chemical delta-9-tetrahydrocannabinol (THC). THC is only one of over 70 cannabinoid compounds found in the plant, each with different biological effects. Collectively these compounds are similar to natural occurring chemicals, such as anandamide, that exist in our bodies and make up the brain's endocannabinoid system, which is important for brain development and function.

These cannabinoid receptors are located throughout the brain especially in regions that influence cognition, pleasure, motor coordination, time and sensory perceptions. High concentrations of cannabinoid receptors exist in the hippocampus (memory), cerebellum (learning and motor coordination) and basal ganglia (motor control.) Activation of these receptors results in the "high" users experience; as well as change in coordination, problem solving, mood and altered perceptions such as paranoia or hallucinations.

Potential Medical Use

Marijuana is self-reported to help people manage symptoms of nausea, loss of appetite, muscle spasm and spasticity, pain and anxiety. There are over 70 identified cannabinoid chemicals in marijuana. These chemical isolates of the marijuana plant have different chemical properties and hence their individual use can be of potential benefit for targeted symptoms. Marinol® is an example of a chemical isolate of marijuana used for a specific medicinal purpose. Marinol®

(dronabinol) is synthetic delta-9-tetrahydrocannabinol (delta-9-THC) and an FDA approved medicine for nausea. Sativex® is a mouth spray approved in Europe for multiple sclerosis related muscle spasticity and contains delta-9 tetrahydrocannabinol (THC) and cannabidiol (CBD).

Our understanding of the chemical properties of these compounds is in its infancy, but laboratory research suggests that some of these compounds have brain protective effects. Both THC and CBD are powerful antioxidants exerting neuroprotective effects in research models of head injury and neurodegenerative disease. Cannabidiol (CBD) holds promise for medical use since it does not have the psychoactive affects described with the marijuana plant or THC compound. Cannabidiol has anti-inflammatory, antioxidant and immunosuppressive properties measured in the laboratory; properties that may be especially important to brain disease such as MS, head injury, Alzheimer's disease and Parkinson's disease.

Tremor is a symptom of many conditions including essential tremor, Parkinson's disease, MS, head injury, metabolic abnormalities and can be caused by some medications. There is limited research to guide the use of marijuana for tremor and even less understanding of how different types of tremor may or may not respond. An early study of five patients with Parkinson's disease by Frankel and colleagues (1990) showed no tremor benefit after smoking marijuana despite personal accounts of prior beneficial effects. The drug did have an effect on brain chemistry given the drowsiness and euphoria experienced during the study. In 2013, Lotan and colleagues studied 20 people with Parkinson's disease from Israel that smoked marijuana. (Medical marijuana is legal in Israel). They reported an improvement in sleep, pain, tremor and bradykinesia (motor slowness) 30 minutes after smoking marijuana in clinic.

It is not clear whether the benefits measured in these studies are from a direct effect of marijuana on tremor specific brain chemistry or physiology. Marijuana has a complex neurochemistry with combined psychoactive, behavioral and motor effects which alone or combined can impact tremor. For example, tremor will increase with stress and improve with treatments known to enhance relaxation. Marijuana's behavioral effects may lead to enhanced relaxation, euphoric mood or mitigate the stress response, and this alone could reduce tremor.

Marijuana "Side Effects"

The use of marijuana will likely increase as more states legalize marijuana, and societal attitudes about this drug evolve. Like any drug, marijuana use is not without risk. There are potential negative effects of marijuana and its chemical isolates that are noted below:

Behavioral Effects

Psychoactive effects of marijuana can have negative short-term and long-term consequences. For example, sedation, apathy and depression can worsen with use and exacerbate any sedating properties of anti-tremor medication.

Marijuana can cause psychosis or altered mental states. The elderly, people with cognitive problems, dementia or mental illness may be more susceptible to experiencing these side effects. The combined risk of marijuana in these populations is unknown.

Apathy can lead to lifestyle changes or habits such as lower activity and exercise levels, poorer eating habits and social withdrawal, which can impact the brain and general health.

Stress certainly worsens tremor and marijuana may indeed have a positive impact on stress physiology. Since there are risks to using marijuana, other alternatives that combat stress such as meditation, guided imagery, exercise and yoga should be considered as treatments with lower risk and perhaps broader general health gains. These anti-stress techniques should not be overlooked as part of a holistic approach to managing tremor.

General Health Effects

- Heart rate is increased by 20-100% for up to three hours after smoking marijuana with an increased risk of heart attack one hour after use.
- Pregnancy. Marijuana can alter the developing endocannabinoid system in the brain of the fetus. Consequences for the child may include problems with attention, memory, and problem solving.
- Smoking. The long-term impact of smoke inhalation and chemical particulates on lung function is unknown.
- Stroke. THC causes brain artery constriction and atherosclerosis which may lead to stroke. Whether this is true for all users is unknown.

Other Effects

- Medical marijuana is legal in some states but is still an unregulated drug with associated risks including potency, contamination and impurities. Synthetic marijuana is often "laced" with other psychoactive compounds to increase the users "high."
- Addiction and long-term use. About 9% of users become addicted to marijuana. Younger individuals and daily users have an increased risk. Withdrawal symptoms include irritability, sleeplessness, decreased appetite, anxiety, and drug craving.
- Long-term marijuana use can impact memory, learning, judgment and increase the risk of mental illness such as psychosis or depression and addiction.
- Impaired judgment and motor coordination can double the risk of a motor vehicle accident and this increases substantially if combined with alcohol.

As with any drug, there are pros and cons to using marijuana, and it is important to review these with your healthcare provider. In particular, the potential addictive, psychoactive and behavioral consequences of marijuana use must be taken into consideration. Nevertheless, specific cannabinoids found in marijuana hold promise as therapeutic agents for neurologic conditions offering yet another strategy for treatment, especially when traditional medications have failed or cause intolerable side effects.

Dr. Monique Giroux is co-founder of the Movement and Neuroperformance Center of Colorado in Englewood, CO and medical director of movement disorders for Swedish Medical Center. She is board certified in Neurology with specialized training in Botulinum toxin (Botox®) for dystonia, pain and spasticity as well as deep brain stimulation for tremor, dystonia and Parkinson's disease. She is also medical director of the Northwest Parkinson's Foundation Wellness Center, and project leader for the National Parkinson's Foundation Care Center Consortium Project. Along with Sierra Farris, PAC, she co-authored Every Victory Counts: Essential Information and Inspiration for a lifetime of wellness with Parkinson's, produced in collaboration with the Davis Phinney Foundation.

Botulinum Toxin Treatment in Tremor

By Joseph Jankovic, M.D.

Since its introduction in the 1980s, botulinum toxin (BTX) has revolutionized the treatment of various disorders associated with muscular spasms and involuntary movements, including tremors [Jankovic, 2004a]. Long-term follow-up of patients treated with repeat BTX injections for more than a dozen years show this treatment is effective and safe [Mejia and Jankovic, 2005].

Almost all types of tremor have been reported to benefit from BTX injections. Initially used in the treatment of dystonia (involuntary muscle contractions), BTX was observed to improve not only the abnormal movement and posturing, but also the accompanying dystonic tremor. This observation stimulated interest in BTX as a treatment modality for different types of tremor.

BTX treatment of limb tremor

In their initial pilot study, Jankovic and Schwartz [1991] found 67% of patients with disabling head-neck (42 patients) and hand (10 patients) tremor had moderate to marked functional improvement and a reduction in the amplitude of their tremor. In a subsequent study, Trosch and Pullman [1994] treated 12 patients with Parkinson's disease (PD) and 14 with ET for their hand tremor. Six weeks after the injection, five patients each with PD and ET reported improvement of 3 or greater on a 4-point scale. The only adverse reaction was weakness of digit extension, noted on examination in all patients, but this did not result in functional impairment.

Jankovic et al [1996] conducted the first double-blinded, placebo-controlled study in patients with ET hand tremor. Twenty-five patients with a 2+ to 4+ tremor severity score were randomized to receive placebo (12 patients) or 50 U of BTX (13 patients) injections into the wrist flexors and extensors. In addition to a statistically significant improvement in a tremor score in the BTX-treated group as compared to the group treated with a placebo, 75% of BTX-treated patients vs. 27% of placebo-treated patients reported significant improvement. This was also confirmed by postural accelerometry, amplitude of tremor. Adverse reaction was mild finger weakness in about half of the BTX patients at week four and persisted in some to week 16, but no patient reported interference with daily functions or activities.

Brin et al [2001] conducted a similar placebo-controlled trial involving 133 patients with hand ET recruited from 10 centers in North America. The patients were randomized to one of three treatment groups: low dose BTX, high dose BTX, and placebo. They were then followed up for 16 weeks. The postural tremor was rated 2 on a 0–4 scale. For the low-dose group, 15 U were injected into flexor carpi radialis and ulnaris and 10 U into extensor carpi radialis and ulnaris. For the high-dose group, 30 U were injected into the flexors and 20 U into the extensors. Members of the control group were injected with the comparable volume containing albumin and sodium chloride solution.

Based on tremor rating, both low and high BTX dose groups improved significantly by physician and subjective ratings, with peak effect at six to 16 weeks. Hand weakness was the most common side effect; 30 percent of the low-dose group and 70 percent of the high-dose group complained of decreased grip strength. Other adverse reactions included rash, pain, stiffness, cramping, hematoma (blood clot), and paresthesia (burning or prickling sensations). Since these initial studies, we have significantly modified our technique to focus chiefly on the flexor rather than extensor forearm muscles. As a result, hand weakness is now a rare adverse effect, and even if it does occur, it usually

is not troublesome and resolves spontaneously within days or weeks. Based on our long-term experience, for refractory tremor, we can conclude that BTX treatment is usually effective in reducing the tremor amplitude and improving function, and this treatment strategy should be considered before any surgical intervention.

BTX treatment in head tremor

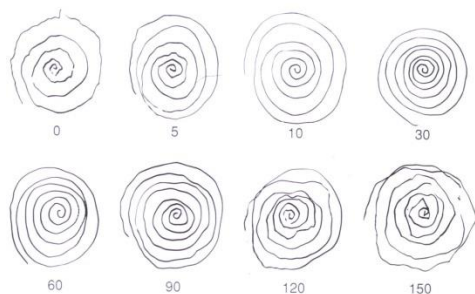
Head tremor, usually due to essential tremor, generally does not respond well to medications such as propranolol and primidone [Jankovic, 2002]. Intractable head tremor, therefore, is particularly suited for treatment with BTX injections. Besides ET, head-neck tremor also presents in up to 68% of patients with cervical dystonia (CD) [Pal et al, 2000]. Many studies have shown that BTX treatment not only improves CD, but also improves associated head tremor in the majority of CD patients [Jankovic, 2004b].

Pahwa et al [1995] conducted a double-blind, placebo-controlled study of BTX-A treatment in 10 ET patients with intractable head tremor. Each subject received normal saline as placebo or BTX injection three months apart. BTX was injected into each sternocleidomastoid muscle at 40 U and splenius capitis at 60 U under EMG guidance. Rating by “blinded” examiners showed that 50% had moderate to marked improvement with BTX as compared to only 10% improvement in patients who received placebo. Subjective moderate to marked improvement also occurred in 50% of patients treated with BTX as compared to 30% in those who received placebo. There was no statistically significant difference between the two groups, however, when the tremor was measured by accelerometry. Side effects were again transient and mild, mainly neck weakness, swallowing difficulty, and headache.

Wissel et al [1997] assessed 43 patients with head tremor: 29 suffered from tremulous CD and 14 had ET head tremor without dystonia. Average BTX-A (Dysport®) dosages were 500 U (range 320 to 720 U) in CD and 400 U (range 160 to 560 U) in ET. After two to three weeks, subjective improvements occurred in 100% of ET and 90% of CD patients. Side effects were mild and transient, including local pain, neck weakness, and dysphagia (difficulty swallowing) in 40% of ET and 39% of CD patients.

In summary, essentially all patients with head tremor of various etiologies (ET, CD, or cerebellar-rubral lesions) seem to benefit from BTX injections. According to different studies, at least half of the patients improve on objective examinations or measurements, while subjective improvement percentages can be even more robust. Response typically occurs a week after the injection and may last for eight to 12 weeks. Common side effects are mostly mild and transient, including neck weakness, swallowing problems, and local pain.

Figure below: Improvement in drawing of a spiral after BTX injection into wrist flexors in a patient with ET during a 150-day follow up. Shows marked improvement during days 10-60.



BTX treatment in voice tremor

Spasmodic dysphonia (SD) had long been successfully treated with EMG-guided injections of BTX [Jankovic, 2004a]. The response of BTX injection on essential voice tremor (EVT), however, has not been thoroughly studied until recently. EVT is caused by oscillation of the vocalis muscle complex or posterior pharyngeal muscles with frequency about 5 to 7 Hz, most noticeable during sustained vowels. EMG recordings have shown that the intrinsic laryngeal muscles, specifically thyroarytenoid, are the most frequently involved. While the voice of SD is characterized by a strained, choked, strangled, and abrupt character (adductor type) or breathy, whispering voice (abductor type), EVT is characterized by pitch breaks and vocal arrests with excessive or interrupted glottal airflow. Approximately 25% of patients with ET have EVT.

The mechanism by which EVT is reduced with BTX injections is not completely clear. Hertegard et al [200] studied BTX injections in 15 patients with EVT. All patients were injected into thyroarytenoid muscles with additional cricothyroid or thyrohyoid muscles in some patients. Ten patients reported subjective improvement one month after the injection. There were also significant improvements when the voice was tested by perceptual evaluations of recordings of patients' connected speech and sustained vowels on the digital audiotapes rated by two phoniatricians.

This evaluation showed a significant decrease in voice tremor during connected speech. Twelve of 15 patients had a temporarily breathy and weak voice for one to two weeks. Three patients had hoarseness lasting up to four weeks. The authors concluded that the treatment was successful in 50% to 65% of patients depending on the method of evaluation. These results were confirmed by other investigators [Warrick et al, 2000].

In summary, about 30% to 50% of EVT patients improve based on objective acoustic analysis, and 65% to 80% of patients improved by subjective assessments. The objective response rates in patients with EVT are not as robust as those in SD. This is probably due to the fact that different factors in laryngeal, respiratory, and orofacial systems contribute to the generation of EVT. The beneficial effect of BTX injection is limited to the effect on the thyroarytenoid muscle, the most frequently injected muscle. The side effects are mostly mild and temporarily include hoarseness, breathiness, and weak voice. The various studies suggest that BTX injection is an ideal therapeutic option for patients with EVT who usually fail to obtain satisfactory response to oral medical treatment.

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Acupuncture in Neurological Conditions

Author Clare Donnellan combines Western and Traditional Chinese Medicine concepts of treatment, Tremor Talk.

Acupuncture expert Clare Donnellan, co-author of *Acupuncture in Neurological Conditions* (Churchill Livingstone) aims to improve patient care by combining Western and traditional Chinese medicine (TCM) concepts of treatment.

The language of TCM is uniquely combined with evidence-based clinical reasoning to provide an approach relevant to both acupuncture and physiotherapy clinical practice.

In the book, Chinese medical patterns relevant to the application of acupuncture are described, as well as key patterns of dysfunction based on a Western medical perspective. The place of acupuncture within the overall management of different neurological conditions is also discussed. The book offers clinical reasoning options from both TCM and Western medical perspectives and is illustrated by real cases from clinical practice.

IETF: What is the difference between the Western and TCM concepts of treatment?

Donnellan: Western Medical Acupuncture is based on insights gained from the scientific literature. For example, how information about pain is processed in the nervous system and how it may be modulated by acupuncture. Acupuncture based on TCM draws insights from the traditional Chinese literature on acupuncture as practiced over thousands of years.

IETF: What evidence is provided in the book to support these practices?

Donnellan: Relevant scientific papers have been examined and reported on. In addition, a wide range of actual patient case studies are reported, for example, the use of acupuncture for people who have Parkinson's disease, stroke, multiple sclerosis, Guillain-Barre syndrome, etc. Varying symptoms have been the targets of treatment, for example, pain, sleep dysfunction, mood dysfunction, spasticity, etc.

IETF: What are some key patterns of dysfunction based on a Western medical perspective?

Donnellan: A key pattern of dysfunction might relate to the motor system. Motor dysfunction in people with neurological conditions may include inability to move (paresis or paralysis) or "excessive motor activity," e.g. spasticity, rigidity or dyskinesia.

IETF: How can acupuncture work with, or apart from, Western medicine to treat neurological conditions?

Donnellan: People with neurological conditions report a wide range of problematic symptoms. Acupuncture can be an extremely valuable option alongside other treatments. For example, a person with an acute relapse of multiple sclerosis may also report difficulty sleeping. This in turn may contribute to daytime fatigue and poor concentration. This may impact directly on that individual's ability to participate in active physical therapy or occupational therapy. Acupuncture may therefore help the problematic symptom (e.g. sleep disturbance or pain) which in turn also allows an individual to gain the most benefit from any rehabilitation program that they may be receiving. Acupuncture may be used as a stand-alone therapy, but considering the complexity of presentation of many people with neurological conditions, its use within the context of a specialist multidisciplinary healthcare team can be invaluable.

IETF: The book covers many conditions ranging from Parkinson's disease to spinal cord injury. While there is nothing specific regarding ET, can you offer any comment on the practical application of acupuncture for this condition?

Donnellan: As with all neurological conditions, the use of acupuncture will be aiming to improve symptoms such as pain, anxiety, muscle dysfunction, sleep dysfunction, etc. While some symptoms may resolve entirely with acupuncture, often just an improvement in symptoms is useful to the individual. People with essential tremor may also report sleep disturbance, pain, etc., and therefore in this context, acupuncture may be very helpful.

IETF: What other key points might people want to consider when reading the book and consulting with a specialist about acupuncture?

Donnellan: I think it is important for people to have realistic expectations of treatment, i.e. not to expect a miracle cure, but to hope for some reduction of problematic symptoms. Regarding practitioners of acupuncture, you would want to know they were fully qualified and accredited by the relevant professional body.

Thinking About Deep Brain Stimulation Surgery? Factors to consider about surgical treatment of essential tremor

Tremor Talk

The idea of undergoing deep brain stimulation (DBS) surgery to treat essential tremor can be a daunting prospect. While fear of the unknown is natural, prospective patients can empower themselves with knowledge. Learning as much as possible about DBS and considering its implications can aid those with ET in making clear, informed decisions about whether or not to proceed with surgery.

About DBS Surgery

In the 1950s, a surgery known as thalamotomy was developed to treat tremor by creating a lesion or destroying a portion of the ventralis intermedius (VIM) nucleus of the thalamus. Since then, the less invasive deep brain stimulation (DBS) surgery of the thalamus was developed as an alternate treatment for ET and approved in 1997 by the Food and Drug Administration. DBS was shown to have beneficial effects with no destruction of the brain. In the procedure, a wire (electrode or lead) is placed in the thalamus portion of the brain. The wire is connected under the skin to a pacemaker-like device in the chest that provides mild electrical currents to control symptoms of ET.

The surgery can be performed unilaterally or bilaterally where a lead is implanted on one or both sides of the brain. Each half of the brain controls movement in the opposite side of the body. The aim of the surgery is to minimize the severity of the tremor. DBS surgery, or any surgery, cannot cure essential tremor. In fact, the effects of DBS surgery are reversible. If necessary, the implanted device can be removed without having damaged the brain. Also, the battery wears down and must be replaced every three to seven years depending on use. If the device is disconnected or removed, or if the battery is depleted, then the stimulation ceases and tremor may ensue.

Informed Decisions about DBS Surgery

Any surgical procedure carries risk that should be evaluated by both patient and the medical professional before the surgery. Because DBS surgery involves an operation on the brain, people should consider many factors as part of the decision-making process. Ultimately, it is the patient's responsibility to make this life-altering choice. Ask key questions, learn about the risks and benefits, and weigh them carefully.

"A patient should know that talking to a specialist about DBS isn't a commitment," says Dr. Jerrold Vitek, Chairman of Neurology at the University of Minnesota and a movement disorders specialist. Seek information from a variety of qualified sources. Begin with your primary physician and include a neurologist or movement disorders specialist with extensive knowledge about ET and DBS surgery. "Get information on what DBS is and how it works. Don't rely on the Internet," says Dr. Vitek. To gather facts and informed perspectives, "Find a DBS center and talk to the staff to learn about the surgery. Talk to people who have had the surgery done."

Several questions should be addressed in the early stages of evaluation.

- Have you been properly diagnosed with essential tremor by a physician or specialist experienced in diagnosing and treating ET?

- Have you tried medication as an initial treatment? A beta blocker such as propranolol (Inderal®), the anti-seizure medicine primidone (Mysoline®), or other drugs are typically prescribed first to counter tremor.
- How does the tremor impact your quality of life? Is the tremor severe enough to significantly affect essential daily tasks?
- Do you understand the steps involved with the initial DBS surgery, post-surgery, maintenance and subsequent procedures?
- Given your age and severity of tremor, is it worthwhile to consider DBS surgery a treatment option?
- Are you physically able to undergo surgery? Are there other health factors, i.e. diabetes or hypertension that would impact your overall health and ability to recover from surgery?
- How do you feel about undergoing brain surgery? How do your family members feel about the risks and possible benefits?
- Does ET affect one or both hands? Is the tremor stronger in the dominant hand or the alternate hand?

A patient may opt to have unilateral or bilateral DBS surgery, depending on the severity of the tremor, which hand it affects, and how much the patient depends on use of that hand. If a patient has a tremor but can function effectively to their satisfaction, then DBS will likely not interest them. “It’s their decision,” Dr. Vitek says. “Maybe their tremor is slight, but their job requires fine work with their hands, and therefore DBS may be a consideration.”

Since each side of the brain controls movement in the opposite side of the body, surgery can be performed on one side of the brain to effect change in a particular hand. A bilateral surgery would impact not only both sides of the brain, but also the tremor in both hands. Each type of surgery carries different implications and should be considered separately.

“With our approach, we do one side first and evaluate the results,” says Dr. Vitek. “Sometimes the patient’s quality of life improves with their ability to do tasks. Showing improvement in one arm may be enough. Some people would prefer to do the bilateral procedure since they are already in surgery.”

Does the DBS center have a qualified neurosurgeon and neurologist on staff with experience in treating ET? Dr. Vitek points out that additional staff such as a physical therapist or a neuropsychiatrist can be helpful in pre- and post-surgery treatment.

Does the facility have such professionals on staff?

Do you and your family members understand the risks of DBS surgery?

DBS surgery is not always 100% effective. This fact alone should be weighed carefully before undergoing surgery. As with any surgery, risk is involved. The surgical risk of DBS often depends upon proper patient selection and the experience and expertise of the neurosurgeon.

Surgical complications occur in fewer than 5% of patients. Risks and complications include bleeding in the brain, stroke, seizures, and infections. Most of these complications resolve within one month.

However, in approximately 1-2% of patients, these complications can result in permanent neurological deficit. Death results very rarely.

Other surgical complications can include confusion, headache and bleeding under the skin which are typically temporary and resolve without treatment. Dr. Vitek states, “There is approximately a 1% risk of significant bleeding per lead implanted in the brain.”

Regarding possible infection, stitches in the head and chest require attention to clean the wound after surgery. If an infection develops in the chest below the clavicle where the wire connects to the battery, then the extension wire is detached from the battery, and the infection is treated before reconnection.

Do you understand the programming procedure after the surgery and the need for subsequent follow-up visits? Given the all clear by specialists, the patient next undergoes programming usually one to six weeks after surgery to initiate deep brain stimulation. The programming of the battery takes place in the physician’s office. Several sessions may be needed to optimize the signal and maximize the benefit of DBS.

Has your specialist explained the possible side effects from the surgery?

In some cases, the patient may experience a transient tingling in the arm or face. This sensation is the result of the lead being placed close to an area of the brain next to the thalamus. Minor adjustments can be made to the signal if necessary to minimize the tingling. “Usually, the feeling goes away,” says Dr. Vitek. “It just depends on where the lead is placed.”

Other side effects include numbness, muscle spasms, speech difficulties, mood changes, balance difficulties, and pain. Adjusting the stimulation parameters can reduce most side effects. Speech and balance difficulties are generally more common with bilateral procedures.

The neurologist will examine the patient for any remaining signs of postural or action tremor and make adjustments to the device if necessary. With the latter type of tremor, the individual is attempting an action such as reaching to grab an object intentionally. “Action tremor is the hardest component to improve and resolve,” says Dr. Vitek.

Deep Brain Stimulation Outcomes

Several reports have demonstrated that DBS of the thalamus has a comparable improvement in tremor compared to thalamotomy, but with fewer complications. The majority of studies have reported improvements in tremor in 90% of patients on the side opposite of the side of surgery.

Long-term studies have shown that an improvement in tremor is maintained in the majority of patients up to seven years after the surgery. Multiple studies have demonstrated the immediate and long-term benefits of DBS in controlling tremor with improvements in hand tremor of approximately 90%, and improvements in functional ability and performance of activities of daily living of approximately 85%. Although all of the large studies have targeted patients with disabling hand tremor, in these studies head and voice tremor have had some improvement. The greatest improvements in head and voice tremor were seen with bilateral procedures.

The Role of Rehabilitation in the Treatment of Essential Tremor

By Sara S. Salles, D.O.

A physiatrist (fizz-ee-at'-trist) is a physician specializing in physical medicine and rehabilitation. As the population ages and individuals survive conditions and diseases that once would have been fatal, the resulting long-term disabilities become apparent. This development, coupled with concerns regarding quality of life, bring the field of physical medicine and rehabilitation to the forefront of medicine. The specialty—focused on maintaining and restoring function—serves all age groups and treats problems that touch on all major systems of the body.

The interdisciplinary rehabilitation team consists of various members that include physical, occupational and speech therapists. They work closely with the physiatrist to assess individual deficits and impairments to determine current functional status. The team can then make appropriate recommendations for medications, behavior modification techniques, therapies and the use of adaptive equipment to overcome impairments which can lead to disabilities.

Essential tremor (ET) is a condition that universally affects function. The goals of the rehabilitation team in these individuals include, but are not limited to, the individual's ability to effectively complete basic activities of daily living such as eating, bathing, dressing, communication and mobility as well as completing job-related tasks and hobbies. Furthermore, the objective is to minimize disability and reduce social handicap. The rehabilitation team works together with the individual to determine tremor severity, coexistent disease, current medication therapy, and response to previous therapies, if any. Appropriate therapy treatment plans are then established on a case-by-case basis to include the assessment of adaptive equipment to allow the individual to maintain and improve their functional status and quality of life.

Depending on the severity of the tremor, some individuals will need to re-learn activities using their non-dominant arm which can be taught and reinforced by occupational therapy. Occupational therapy focuses on strengthening the upper extremities and determines the benefits of adaptive equipment. This equipment may be purchased at a relatively low cost at a medical supply store or ordered by the individual therapist. Adaptations, such as wrist weights, can help stabilize the affected extremity and decrease tremor severity.

Other equipment may include the use of a sock aid to assist in donning and doffing one's socks or a button hook to assist with dressing. Long handle shoe horns and elastic shoe laces assist in the donning, doffing, and wearing of shoes. Weighted utensils may assist in eating and in meal preparation to avoid self-injury. For those individuals that use a computer for work or leisure, an adapted computer mouse to avoid multiple accidental clicks due to tremor may allow them better work efficiency. Examples of some of these devices may be found on IETF's website under the subheading of "Assistive Devices."

Physical therapy (PT) has been found to be helpful in individuals with ET. The role of PT is to help stretch and maintain general core and extremity strength. Additional features of PT may include biofeedback, which is the use of electronic monitoring of a normally automatic bodily function in order to train an individual to acquire voluntary control of their extremity and better tremor control. In conjunction with biofeedback, relaxation techniques such as deep breathing exercises, progressive

muscle relaxation techniques, massage therapy and meditation are helpful in lessening tremor as well as stress and anxiety.

Other therapies that help improve tremor intensity include exercises such as yoga, a therapeutic exercise technique that utilizes a number of stretches, poses and breathing for improved muscle tone, strength, balance and flexibility. Yoga exercises may help reduce essential tremor and its associated pain or discomfort. These exercises may also help reduce stress and anxiety, which are common triggers and symptoms associated with essential tremor. Discuss all possible treatment options with your physiatrist prior to initiation of therapy to avoid self-injury.

Individuals with ET may also experience voice tremor and the following challenges:

- 1) Significant difficulty initiating and maintaining phonation during conversation
- 2) Speaking that requires effort
- 3) Fewer communication attempts
- 4) Avoidance of social events and
- 5) Trouble sustaining relationships with others as a result of their voice tremor

Options for managing vocal tremor include medications, alcohol, botulinum toxin injections, and deep brain stimulation (DBS). Adjunctively, speech and language therapy (SLP) is important in helping to control voice tremor. Therapies, such as Lee Silverman Voice Treatment (LSVT), have been shown to improve voice loudness and speech intelligibility. Other compensatory strategies involve laryngeal maneuvers such as a yawn-sigh maneuver, relaxation, and breathing exercises.

Recently, a small study has demonstrated the use of incentive spirometry, a process that improves lung function by increasing lung volumes resulting in better throat muscle forces/vocal fold vibration and decrease in vocal tremor. A larger study is needed to further validate this technique.

With the progression of ET, some individuals may develop dysphagia or difficulty swallowing as a result of their worsening laryngeal or throat tremors. These individuals are at greater risk of aspiration and subsequent pneumonia. It is recommended that these individuals participate in a formal SLP evaluation including a clinical swallowing assessment in the office setting as well as a modified barium swallow, an x-ray study to objectively determine one's swallowing ability. SLP will also aid in teaching swallowing exercise to include throat muscle strengthening exercises as well as compensatory techniques to avoid pneumonia.

In addition to medical and surgical management of ET, researchers continue to seek adaptive devices to impact and improve the lives of those with ET. Current projects include adaptive utensils by Liftware intended to help control tremor and their ability to feed one's self by detecting and correcting for the tremor rather than the individual trying to control or stop their tremor.

At this time, there are numerous research studies across the country investigating various treatment options to help those individuals with ET. Currently, medical treatment, surgical options, and exercise remain the mainstay of therapy and treatment. People are encouraged to remain physically active and maintain a healthy lifestyle to avoid further medical problems. To learn more, please contact your local neurologist (www.aan.com) or physiatrist (www.aapmr.org).

The Role of Physiatry in Treating ET

By Sara S. Salles, D.O.

Physiatry is the practice of physical and rehabilitation medicine and is led by a physician called a physiatrist. The focus of the specialty is maintaining and restoring the ability to engage in everyday activities and tasks by treating problems that affect all body systems. Working with the physiatrist is a team of physical, occupational, and speech therapists that assesses individual problems and makes therapy recommendations.

In working with a person with ET, the rehabilitation team determines tremor severity, co-existing disease, current drug therapy, and response to previous therapy. Appropriate treatment plans are then recommended, including the use of adaptive devices that allow the individual to continue and increase daily activity, thus improving quality of life.

Depending upon tremor severity, some individuals with ET will need to re-learn activities using Constraint Induced Movement Therapy (CIMT). This involves constraining the tremor-affected arm to force use of the unaffected or less-affected arm. In general, CIMT is time-consuming and can be frustrating, but it has the potential to deliver great results as demonstrated by stroke survivors.

In addition to therapy sessions, all persons with ET are encouraged to remain physically active and to maintain healthy lifestyles to avoid further medical problems.

To learn more about rehabilitation in individuals with ET, please contact your local neurologist or physiatrist at www.aapmr.org.

Coping With Essential Tremor

Strategies for Coping with Chronic Disease

Tremor Talk

ET is a chronic progressive disease, and as with all chronic diseases, there are mild, moderate and severe cases. For many people with a moderate or severe case, living with any chronic disease can be a lonely, painful journey that often does not have a well-defined beginning, seems to have never-ending peaks and valleys, and certainly has no happy ending with a cure.

In the United States today, the healthcare system is geared towards treatment of acute illnesses — conditions with a beginning, middle and an end. Beyond providing services of a psychologist or a social worker, few organizations within a community are able to assist an individual and their loved ones with the life-altering challenges presented by a chronic illness.

One of the few organizations available to those affected by chronic disease is Turning Point: The Center for Hope and Healing, a Kansas City area non-profit that provides numerous programs and services to support individuals with the “psychological, social, emotional and physical needs that accompany chronic illness.”

Cathy Pendleton, the director of adult programming at Turning Point, recently shared valuable insights on the process of managing, accepting and living with a chronic disease.

The early symptoms of chronic disease often are ignored by many people or not recognized for what they truly represent. But, according to Cathy, with a diagnosis there is a sense of loss.

“Many of the feelings that go with loss and grief rise up including shock, denial, anger, sadness, depression and fear.”

At this time, a person’s personal coping style kicks in, which can range from researching a disease to learning all one can about it to choosing to remain in denial. Loss of control, both current and possible future losses, become huge issues at this time, and people often begin asking questions such as “What is going to happen now? Why me? Why now? What will my future hold?”

According to Cathy, learning strategies for self-care during this initial period can make dealing with future changes more manageable. But, “within our culture, self-care is often considered to be selfish. People who haven’t learned how to identify and express their needs and feelings often fear that showing their feelings will make them look weak.”

“While men are often taught not to express their feelings — especially sadness and fear— it would be a gross generalization to say that men do not express feelings and women do. Many women have also been taught not to talk about feelings,” says Cathy. “Research has shown, however, that people who are able to identify and express their feelings in ways that make them feel better are more able to manage the ups and downs that accompany illness.”

So, how does someone effectively deal with the realities of living with a chronic illness? Certain strategies can be of great help, including self-nurturing, exercising personal power, finding balance, soul-searching and self-talk.

Self-nurturing includes creating an effective support network. This is an important step not only for the person with the chronic disease, but for their primary caregivers as well.

As the first step in creating a support network, Cathy suggests making a list of priorities and needs, and then determining who can provide for a need based upon their individual abilities, and then keeping the list handy so that when someone calls and offers help, they can be given a specific and tangible thing to do.

“Sometimes we all need to ask for help and give others the opportunity to experience the joy of giving,” explains Cathy. “One friend might be the person that you call late at night if you need someone to talk to, while another friend might be the one that you call to go to a movie to enjoy yourself. Learning to ask for what you need is a wonderful way to exercise personal power.”

Personal power can also involve taking a socially awkward and potentially embarrassing situation and turning it into an opportunity to teach someone about a chronic disease. As an example, Cathy tells of a recent learning experience she had while attending an ET support group meeting at Turning Point.

“I went to hand a man a cup of coffee, and he looked up at me and laughed. He said that is was evident that I did not have ET, because the cup was filled to the top,” says Cathy. “That was a wonderful example of someone taking the opportunity to teach in a friendly, humorous way.” Even medical professionals can benefit from the education a person with a chronic condition can provide, as the staff of the IETF recently learned when Norma Doherty— support group leader from Centerville, OH — emailed her story of going to the doctor’s office and being asked to provide a urine sample using a small plastic cup. Norma had hand tremor.

“When you shake as much as I do, it is very difficult to hold and hit that little cup. After much frustration, I managed,” says Norma. “When I went to the doctor the next time, I mentioned it to the nurse. She told me that they have ‘a hat’ that fits in the toilet. I just hope that I can help someone else avoid the frustration that I had that day.”

Even though the nurse knew that Norma had ET—she had her medical chart—and possibly saw her tremor, she did not have the personal experience of living with ET to be able to put one and one together and understand that the “usual” way of doing things does not always work for someone with ET. Just because medical professionals have knowledge of a condition and can provide treatments, unless they have a condition they have no living knowledge of it.

Finding balance in our lives is important because we often forget to include ourselves in our “circle of care.” This can be especially true for the people supporting the person with the illness. They often put themselves at the bottom of their list of priorities until they feel like they’re running on empty.

It’s also important to realize that people find balance differently, explains Cathy. “The introverted person may need more solitude to replenish themselves while the more outgoing person may get recharged by being around others.”

Soul searching includes accepting that life will never be the way it was before and discovering one’s “new normal.” This involves looking for ways in which life still has meaning and being able to maintain a sense of hope, even in the midst of all of the changes that come with the illness.

As an exercise in soul searching and managing the feelings that come up in this process, Cathy suggests journaling. “Sometimes just writing about feelings gives people a release and can offer clarity about issues they may be struggling with.”

For people with ET, writing or typing can be problematic. For these people, journaling can be accomplished with the use of speech recognition software such as Nuance’s Dragon NaturallySpeaking voice recognition software, which is available at many computer and book stores for approximately \$100.

Other options for managing feelings,” suggests Cathy, “include talking with trusted friends, family, clergy, a support group or a counselor, and taking a walk or meditating.”

Self-talk is about the messages that people give themselves, having realistic expectations and remembering that they are more than the symptoms of their chronic disease.

An exercise she uses at Turning Point has two people sitting across from each other and talking for 10 minutes about themselves without mentioning anything related to their chronic illness. She says this is very difficult because they are so focused on illness that they to view themselves as being that illness.

These five strategies — self-nurturing, exercising personal power, finding balance, soul-searching and self-talking —can provide a more meaningful perspective on managing a chronic disease.

Coping with ET

Tremor Talk

Essential tremor (ET) is a life-altering condition that makes everyday living a test of ingenuity, perseverance and self-esteem. Daily activities such as writing a letter, dressing, and eating cause frustration that can lead to stress and a temporary worsening of tremor. In order to assist people who have ET in continuing to live full, meaningful lives, the IETF offers the following coping tips.

General suggestions

- Learn to use your tremor-free hand for as many activities as possible, including writing.
- Hold your chin toward your chest, or turn your head to the side to control head tremor.
- Use your tremor-free hand to steady your trembling hand, and whenever possible use two hands.
- Avoid caffeine, ma huang, ephedra and other over-the-counter medications and herbs containing ingredients that increase your heart rate and can increase tremor temporarily.
- Keep your elbows close to your body when performing tasks to help control hand tremor.
- Carry a small tape recorder with you to record notes.
- Carry and use larger, weighted pens and eating utensils.
- Use a signature stamp when possible for signing your name.
- Carry a strip of self-adhesive address labels to give to people who ask for your name and address.
- Fill out deposit and withdrawal slips at home before going to the bank.
- Consider using online banking to pay your monthly bills.
- If you write checks, do them all on a “good” tremor day.
- Consider using credit or debit cards instead of writing checks.

Eating, drinking, food preparation

- Use travel mugs with lids. When on the go, use lids for purchased beverages whenever possible.
- Carry straws with you. You can find sturdy, thick, plastic straws in many housewares sections of stores if thin, plastic straws are too flimsy.
- Use heavier glasses and mugs instead of light-weight cups. Soup mugs are also a good choice for drinking.
- When holding a mug or small glass, place your thumb along the rim and place your fingers across the bottom.
- Fill cups, mugs and glasses half-full.
- Request your meat be cut in the kitchen before being served.
- Consider ordering finger foods to eliminate the need for utensils.
- Ask that your soup be served in a mug.
- Consider using dishes that have vertical sides or buy rubber bumper guards from a medical supply store to place around the edges of your plates so you can more easily scoop your food.
- Try using covered ice-cube trays.
- Get a rubberized placemat that sticks to the table so plates do not slide.

- Put your microwave on a countertop or a low table so you can easily place food inside and remove it.

Dental visits

- Novocain increases tremor in some people. Ask your dentist if there is an alternative.
- Notify your dentist of all the medications you are taking.
- **Head tremor only:**
 - Request that your dentist stop periodically so you can massage and rest your jaw and your head.
 - Ask your dentist whether a bite block will help steady your jaw during dental procedures.
 - Talk with your dentist about having a person in addition to the dental assistant help with your procedure. The third person can gently hold your head to help control tremor.

Personal care

- Use an electric razor when shaving.
- Have a manicurist care for your nails.
- Have a cosmetologist wax or tweeze your eyebrows.
- Try using disposable floss holders when flossing your teeth.
- Hire a seamstress to do your mending, or find a volunteer to sew on buttons, thread needles and pin fabrics. Use Velcro® fasteners rather than buttons.
- Use an electric toothbrush or a child's toothbrush for better control when brushing your teeth.

Applying makeup and putting on jewelry

- Apply mascara by resting your elbows on the countertop. Put the wand in one hand and use the other hand to keep the wand steady.
- Apply eyebrow pencil, mascara, eye liner or lipstick by resting your finger or the palm of your hand on your face to steady your hand.
- Put on earrings by resting your elbows on a table. If you have head tremor, place your chin on an up-ended facial tissue box to steady your head.

Using technology

- Use a telephone with large buttons. Avoid phones with speed dial and redial buttons too close to the number buttons.
- Use a speakerphone, a headset or Bluetooth® device when using phones.
- Keep a small tape recorder next to the phone so you can record information when talking on the phone.
- Ask your security alarm representative to give you a remote to turn your system on and off.
- Use voice-activated dialing if available on your cell phone.
- Set your computer to omit double strikes on the keyboard and double clicks on the mouse. Go to your computer's "Control Panel" and click on "Accessibility Options" to set these options.

- Go to a store selling a number of different types of computer mice, try them out, and choose the one that works best for you.
- Check into speech-recognition software. Some computers are pre-loaded with this.
- When choosing a digital camera, pick one with image stabilization technology.

Airport security

- Getting through airport security can be especially difficult for people with ET. It can be a frustrating experience so be prepared.
- Give yourself plenty of time to arrive well before a flight so that you do not have to rush through security.
- When dressing for air travel, wear slip-on shoes that are easily removed to be put into checkpoint trays and easily put back on afterward.
- Place everything you usually carry in your pockets, such as coins, keys, and cell phone, into a plastic bag in advance so it is easier to place in the security tray.
- Have your ID and other travel documents together before entering security.

This information is not intended to replace your current medical therapy. Discuss your difficulties with your physician or other health care professional in order to help develop a well-rounded treatment plan that is right for you.

Patient Strategies for Good Outcomes

By Patricia Foote, Medical Journeys Network, reprinted with permission

Become the team leader of your medical team. Remember that you are paying, so hire professionals who are competent, compassionate and communicate well.

Understand your condition. With the use of trustworthy sites on the Internet such as the IETF, National Institutes of Health, the Office of Rare Disease, and the Genetic Alliance portal to more than 600 resource organizations, it is easier to find quality information. Lay advocacy groups are becoming experts in their specific condition. This enables consumers to prepare for doctor visits with a list of intelligent questions and to have information about different treatment options, including clinical trials.

Manage the routines of dealing with your condition. Remember to take medications and attend medical appointments. Keep a health history form up to date so that this can be easily sent to new doctors and also for your own ease in remembering what screenings, medications, etc., you have had. Monitor diet, rest and exercise.

Acknowledge emotions that come with diagnosis of, or life with, a genetic condition. Recognize fear, anger, depression and frustration. Seek professional help if things are bad or perhaps locate a support group for people with your condition. Find ways to reduce stress that are enjoyable such as walking, reading, or listening to music. Be sure to schedule a break for yourself if you are the primary caregiver.

Reach out to friends and family, even if activities have to be modified. Try to maintain relationships by clear communication: “I need”, “I feel”, “I’m afraid of.” Be specific when asking for help: “Could you please go grocery shopping for me on Tuesday?” “I’d love it if you could prepare dinner on Thursday.” “Will you please read the newspaper to me?” “Will you just sit next to me, hold my hand and say nothing?”

Stay apprised of health insurance issues. For example, what procedures require pre-approval; what is your annual deductible; what is your lifetime cap; what protections do HIPPA (Health Insurance Portability and Accountability Act of 1996) and the new Health and Human Services Privacy Regulations offer?

Stay sensitive to privacy issues. Who needs to know the information? If in doubt, don’t disclose. Never give blanket disclosure authorization.

Designate someone to coordinate your medical management for you if you are unable to take on a proactive role.

Learn to accept your diagnosis and live your new life. Integrate the business of living with a chronic condition into your life. Remember you are much more than your disorder. Accept and understand what is “normal” for you, which may include an increased amount of time spent on medical issues and some physical discomfort. It may not always be possible to “win” the fight with a genetic condition – some are terminal. The knowledge gained from the battle could be considered precious, and victory a deeper love of those who shared the journey.

Realize that sometimes blessings and insights come from living with a genetic condition. Perhaps you learn to set priorities, value the use of time, become more empathetic, and take care of unfinished business.

A recent PBS TV special, *Critical Condition*, stated the following: “The quality of your healthcare depends as much on you as on your doctor, on your being informed, on your asking the right questions, on your being your own best advocate. Your life may hang in the balance.”

Communicating with Your Doctor

Tremor Talk

Not long ago, patients deferred to physicians when making healthcare decisions. But research and experience show that patients who take responsibility for their own health care feel more confidence in their doctor and in the quality of their care.

An important aspect of the doctor-patient relationship is the ability to communicate honestly and comfortably. The following are guidelines to help you to prepare for a doctor's appointment, communicate with the doctor during the appointment, and follow-up afterward.

Preparing for an appointment:

- Write down a list of your concerns and questions and prioritize them. Include symptoms, questions about medications, and other concerns.
- Make a list of all medications you take (including prescription drugs, over-the-counter medicines, vitamins, herbal remedies, teas or supplements.) Include the dosages and how often you take them OR put all of these items in a bag and take them with you to the appointment.
- Gather insurance cards, names and phone numbers of other doctors you have seen in the past. If this is your first visit, bring copies of your medical records.
- Remember to take your eyeglasses and hearing aids if you use them.
- Take a pen and paper to take notes or, if needed, bring someone with you.

During the appointment:

- Tell your doctor your most important concerns at the beginning of the appointment, and then discuss each in turn.
- When describing a symptom, include a brief description of when it started, how often it happens, and if it is getting worse or better.
- Be honest when answering questions and offering information. Don't make the doctor guess, and don't withhold important information. When discussing sensitive topics, bring copies of information you find on the Internet regarding the topic in order to help you ease into the subject.
- Tell the doctor if you have had any major life change such as the death of a loved one or the loss of a job.
- For any medications prescribed, ask the doctor what the medication will do, how long you will need to take it, and what side effect you can expect.
- Ask questions about anything you don't completely understand. Use "I" statements at the beginning of your question: "I don't know how often I need to take this medication. Can you tell me?"
- Tell the doctor if you feel confused, worried or rushed about anything that is said or that takes place during the appointment, or about how your treatment plan will affect your life. Again, use "I" statements. If there is not enough time for all of your concerns, ask for a follow-up visit.
- Take notes during the appointment. If need be use a tape recorder, but ask the doctor for permission.
- Ask the doctor for written literature about your health condition or treatments.

- Cultivate a good working relationship with the doctor's staff. They are part of your health care team too and can become important advocates for you.

After the appointment:

- If you forgot to ask a question during the appointment or have a new one, call the doctor's office.
- You might not be able to talk to anyone until the end of the day or until the next day, but make it clear that someone should return your call.
- If you have any unexpected reactions to medication or you experience any of the adverse side effects, call your doctor. If you believe it to be an emergency reaction, call 911.
- Make a follow-up appointment if needed.

Get the Most from Your Health Care Team

National Caregivers Library, reprinted with permission

Whether you are the primary caregiver for a family member or taking care of yourself, it pays to ask questions in a clear and assertive manner. Don't assume "the doctor will let us know if..." The best policy is to work actively with the health care team.

Use these suggestions to take the initiative.

Stay educated on each condition or treatment

- Research suggests that caregivers and patients who educate themselves get better results from doctors. Learn all you can to explore treatment options and alternatives knowledgeably.
- Ask the doctor for books, DVDs, or other materials that explain your loved one's condition and treatment.
- Get information from specific organizations such as the IETF, Alzheimer's Association and the American Heart Association.
- Speak up if you have questions or concerns. You have a right to question anyone involved with your loved one's care.

Discuss personal wishes

Before meeting with the doctor, get firm answers to the tough questions. Review these issues as early as possible before there is a crisis. And consult a lawyer about living wills, durable powers of attorney for health care, and other documents that can help insure your loved one's wishes are carried out.

- Who should make medical decisions if your loved one cannot?
- What kind of medical intervention does your loved one want? Under what circumstances heroic measures should not be taken?
- What medications or procedures should be avoided?
- What worries or fears does your loved one have?

Prepare for doctor's appointments

Before each meeting with the doctor, make a list of issues you want to discuss. Write down questions in advance, and make sure you have a pen and paper handy to take notes and record the doctor's answers. Consider asking the following types of questions:

- Can you explain the illness in non-medical terms? Where can I find more information?
- How has the situation changed since the last appointment?
- Are more tests required? A second opinion?
- What treatment options are available? Are there alternatives? What is likely to occur without any treatment?
- What are the side effects of these treatments? Of prescribed medications?
- How can you be reached? If you are unavailable, whom should we contact?
- What steps should we take in case of emergency? What is the likelihood of such an event?
- What are the next steps in the procedure or diagnosis?

Schedule regular discussions with all health care team members

A health care team may include a primary doctor, specialists, nurses, health aides, care professionals, family, and friends. In cases of complicated illness, you may want to draw these people together for a “health care conference” that will get everyone on the same page. Don’t assume all members of the health care team know the full picture; ask the primary care physician to take charge as “quarterback” to make sure everyone is clear about their roles.

Call in “the cavalry” when necessary

- If you are unable to get the results you want on your own, find professional assistance.
- If you are dealing with an elder care situation, consider hiring a geriatric care manager.
- Most health care facilities have resource persons such as social workers, patient advocates, chaplains, and nurses who will work for you and help clarify any concerns.
- If you are battling the “system,” enlist your state ombudsman’s help for managed or long-term care.

About this Article

This article is reprinted with permission of www.caregiverslibrary.org, a nationally recognized resource that provides families with interactive care planning tools, resource locators and helpful checklists to make caregiving easier. The company also provides corporations with a Work/Life program for employed caregivers. For more caregiving information, visit them on the web.

Learn New Ways of Doing Things Through Occupational Therapy

By Karen Mainzer, O.T.L., C.H.T.

“An occupational therapist who specializes in working with people who have neurological conditions can help a person with ET not only learn how to do tasks differently, but look at the world and the way it works differently,” says Karen Mainzer, an occupational therapist (OT) at St. Joseph Medical Center in Kansas City, MO.

Occupational therapy is the practice of assisting someone in returning independence to their occupation. Occupation in this case means an activity performed by anyone within any life role such as parent, homemaker, athlete, business person, volunteer, etc.

As Karen explains, OTs look at the “whole picture, including the person, the task and the environment in order to suggest necessary changes to maintain or achieve independence.”

“Society has a system of ways in which things are to be done, and we try to fit into that system. Problems arise when we cannot do a task in the way the system says it is to be done. As an occupational therapist, I help a person to see that the system can be changed,” says Karen.

As an OT, Karen helps people analyze a difficult task to determine what component, or small part of the task is prohibitive. Additionally, she teaches individuals to look at the component to see how it can be changed, or how the environment in which it operates can be changed. In this viewpoint, she explains, “The person is not inadequate for a task. The task is inadequate for the person.”

As an example of this process, Karen analyzes the task of clipping nails from the perspective of working with a person with ET.

“It’s not that the person with ET is not strong enough or smart enough to clip their nails. It’s that the clippers are not steady enough to do the task. A person with ET cannot change the fact that their hand is too unsteady to work the clippers, but they can change how they do the task, how they use the clippers,” explains Karen.

Five basic principles Karen suggests for people with ET to keep in mind when determining how a task can be completed differently than in the normal way include:

1. Stabilizing the object involved in the task through the use of non-slick surfaces, mounting tools, suction cups, clipboard and pumps;
2. Reducing fine motor demands through the use of weighted utensils, levers, hooks and pulls and Velcro.
3. Conserving energy by taking rest breaks, sitting instead of standing, and sliding objects instead of picking them up to move them.
4. Reducing the potential for not being successful by using covered mugs and glasses, eating finger foods, and using plate guards.
5. Using advances of technology such as electric mixers and can openers, electric tooth brushes, and adaptive keyboards for the computer.

According to Karen, the biggest obstacle some people face in learning new ways to do things is attitudes created by stereotypes. These include saying that: “I am too old to change the way I do things,” “I will look older than I am if I use certain devices”, or “I am not smart enough to change.”

If you believe occupational therapy could teach you new ways of doing tasks that are too difficult or impossible because of your tremor, ask your primary care physician or neurologist to write a prescription for an occupational therapy evaluation. Then discuss your service options with a representative of your insurance company. Be sure to ask for an OT who has experience working with people who have neurological conditions.

The Helping Hands of Occupational Therapy

Tremor Talk

People with essential tremor can relate to the frustration of hitting a wrong key repeatedly while typing, writing illegibly, or spilling rather than drinking a glass of water. Practice doesn't make perfect when hands shake from ET. Rather than struggle with or abandon daily activities, an occupational therapist may be able to help.

Occupational therapists (OTs) provide strategies and assistive devices to accommodate and improve limited motor skills. They use a client-centered rehabilitation approach to help people return to participation in the activities they need and want to do.

Physical exercise is one method to rehabilitate and "re-wire" motor skills. For instance, if your dominant hand becomes difficult to use because of tremor, an occupational therapist could teach you to write with your non-dominant hand.

Rehabilitation begins with gross motor movements, that is, big movements at major joints in the arm and hand, and then work to build accuracy and speed. OTs then move into increasing accuracy and speed in finer hand movements.

"For most people, adding speed is the most frustrating aspect to this therapy," says Kim Dorzweiler, a registered occupational therapist in Overland Park, KS. "People want to be better at something sooner rather than later. We realize the people we work with are already frustrated because they can no longer do what they had been used to doing so easily."

Performing exercises to increase hand dexterity also improves steadiness in people with hand tremor. Doing typing exercises, playing a musical instrument or completing simple arts and crafts projects are basic strategies to get hands working with more finesse. And don't worry if your project isn't quite worthy of entry into the Louvre! Refining the control and dexterity of your hand movements is more important than the artistic results of the activity.

Kim suggests using Chinese medicine balls to build hand strength and dexterity. Also known as Chinese meditation balls, they are made of various metals and have a hollow inner core. The balls are rotated repetitively in the palm of the hand.

Place mats, coasters and jar openers made from non-skid material, available at dycemshop.com, can prove useful. A specialized cutting board is another indispensable kitchen tool for people with hand tremor and can be found at elderstore.com or easierliving.com. Suction cups on the bottom of the board adhere to a counter, and strategically placed stainless steel pegs on the board hold the food to be cut.

Computers, smart phones and other technology play a crucial role in daily life. OTs have developed adaptive strategies to help people with tremor. For example, a large "target button"—bigger than the other buttons on the device—can ease operation, while some devices can even be programmed to work using voice activation.

For the smaller buttons of a mobile device, Kim suggests using a stylus (pointer stick) for a more precise reach. She says, “It’s slow, but effective.”

Some assistive devices are available at medical supply stores. Kim suggests calling the store in advance to make sure they have the item you are looking for in stock. Amazon often sells many assistive devices and is another option. Also, OTs have a variety of catalogs that list assistive devices for sale, if you cannot find what you need locally or online.

Having assistive devices available to ease completion of everyday tasks is the first step. Occupational therapists can offer guidance on helpful devices and exercises to improve hand dexterity, but you must be willing to use the devices and complete the exercises. Some people believe that seeking help is a sign of weakness, but that notion is a false hurdle to overcome so that real rehabilitation can begin.

“Sometimes the job of the occupational therapist is to help the person figure out why the device or strategy is difficult to accept,” Kim says. “That can sometimes open the door to acceptance.”

To learn more about occupational therapy visit The American Occupational Therapy Association website at www.aota.org.

Hypnotherapy and ET

By Judy Callihan Warfield, Certified Hypnotherapist

Let me begin by stating that I am a Certified Hypnotherapist and also suffer from essential tremor (ET). This gives me a unique viewpoint of solution and cause. I have found that ET is more difficult for me to manage when I am stressed, have not had enough sleep, or have not taken the time to eat properly. These are all areas that a qualified hypnotherapist can assist you in improving. The great thing about hypnotherapy is that it is a drug free method to learn techniques to lessen stress, improve sleep patterns, and help motivate you to lead a healthier lifestyle with exercise and diet.

What is Hypnotherapy?

Hypnosis is one of the most ancient and respected medical practices known to man. It's lasted through the centuries because it's a natural, safe, simple human process that all of us do every day, often without realizing it. If you meditate, if you zone out on the golf course, if you escape into music or a good book or movie and let the world drift away, that's more or less what a light hypnotic trance state feels like. The difference in a hypnotherapy session is that it is more focused and directed toward a specific goal.

A hypnotherapy session consists of talking about the issue at hand for approximately 30-40 minutes. The hypnotherapist then hypnotizes the client and suggestions are given directly to the subconscious. A session lasts typically 50 minutes. The effectiveness of this particular type of therapy is revealed in the following statistics by Alfred Barrios, Ph.D.:

- Psychoanalysis - 38% improvement after 600 sessions
- Behavior Therapy - 72% improvement after 22 sessions
- Hypnotherapy - 93% improvement after 22 sessions

This may be startling to many, but the rapid improvement and high success rate is due to the fact that we are dealing directly with the subconscious, where behavior, patterns, and habits reside. You are never out of control, but calm and relaxed and very aware of what the hypnotherapist is saying.

Advancement in Hypnotherapy

We're witnessing a resurgence of interest in hypnosis, within even the most conservative sectors of mainstream medicine because it is drug free, often successful, and rapid. Hypnosis has been approved as a method since 1958 by the American Medical Association, and it is now being actively investigated by the National Institutes of Health and practiced at the Mayo Clinic, medical schools from Stanford to Harvard, and endorsed by UCLA and Blue Cross as an effective adjunct to conventional medicine even for illnesses up to and including diseases such as cancer and HIV. The separation between mind and body has been closed, and the power of the subconscious is today being brought to bear as possibly the best ally you have for staying happy and healthy. Mind/body approaches work because they address the framework of attitudes and behaviors that surround health. It deals with the cycle of stress, anxiety, physical tension, symptoms, and disease. It teaches individuals to break old harmful patterns and develop positive attitudes and healthy behaviors.

Stress

The World Health Organization calls stress, "A global epidemic." Stress is felt to be a contributing factor in heart disease, high blood pressure, suppression of the immune system (which inhibits the

body's ability to fight cancer and HIV), deaths from smoking, deaths from obesity, arthritis, herpes, fibromyalgia, insomnia, migraines, chronic pain and infertility. Stress makes my ability to cope with ET more difficult.

Becoming more aware and improving your ability to reframe and change your perspective in stressful situations will give you a renewed sense of empowerment and a feeling of being in control of your life again. Many people have lost the ability to remember what relaxation feels like. Ask yourself when was the last time you were able to just “let go” and/or feel completely relaxed.

Our subconscious mind dictates our behavior, habits and our “knee jerk” response to stress. Your willpower, logic, and reasoning are located in your conscious mind and will continue to be overridden by the power of your subconscious mind unless you start making the changes where the behavior and habits originate.

The Process

A qualified hypnotherapist will touch on all areas that produce stress and address such issues as insomnia, lack of exercise, poor eating habits, and low self-esteem. Because ET affects people in every aspect of life, our relationships can suffer from the inability to see us as viable and attractive people. Motivation to exercise and sleep better will be instilled. Your ability to cope with others, job difficulties, loss of loved ones, relationship problems, and daily demands will be increased. Limiting destructive thoughts and beliefs will be amended to be more positive. A feeling of hope and optimism for the future will replace those negative thoughts that have now become a habit, and you will begin to see yourself in a more positive light, therefore increasing your self-image.

I cannot emphasize enough the power hypnotherapy has when combined with your doctor's protocol in battling disease and illness. The ability to reduce stress and feel like you are active in your own recovery greatly increases your body's ability to focus and respond to your physician's treatment.

Exercise: So Many Benefits—So Much Spice

Tremor Talk

All exercise—whether aerobics, weight training, strength building, mind-body, or movement—involves physical exertion. Physical exertion provides many health benefits including helping to prevent heart disease, type 2 diabetes, cancer, stroke, high blood pressure. It can also strengthen bones and muscles. The list of health benefits goes on and on.

Regrettably, no type of exercise will cure ET. What exercise will provide the person with ET, though, is still valuable. Other than the physical benefits listed above, most exercise, when practiced consistently as part of a lifestyle, can provide relief from the effects of stress, depression and anxiety—conditions many people who have ET deal with every day.

Although some of the exercises discussed in this article can be practiced in solitude, they can also be practiced within a group setting. For many people, talking and sharing experiences with others on a daily basis is what gives life its spice and is definitely an additional benefit. The lifestyle practice of exercise, according to guidelines from the United States Department of Agriculture and the Department of Health and Human Services, indicate that at least 30 minutes of daily physical activity is required. Before beginning any exercise program it is important that you check with your doctor, especially—according to MayoClinic.com—if you have or have had any of the following: heart attack, asthma or lung disease, diabetes or heart, liver or kidney disease, pain in the chest, joints or muscles during physical activity, arthritis or osteoporosis, joint replacement surgery, loss of balance, dizziness or loss of consciousness, a chronic health condition needing medication, untreated joint or muscle injury, or persistent symptoms after a joint or muscle injury, pregnancy, or an uncertain health status.

The American College of Sports Medicine also recommends you see your doctor if two or more of the following apply:

- Age 45 and older if a man, and 55 and older if a woman
- A family history of heart disease before age 55
- High blood pressure or high cholesterol
- Smoking or having quit smoking in the past six months
- Overweight or obese

Your doctor will be able to guide you to an exercise routine that is best suited for you. Of course, your preferences should also be considered when choosing an exercise program. If you dislike a particular type of exercise, you will not likely continue doing it day-in and day-out so choose several types of exercise in order to maintain interest and commitment. Incorporating exercise into your lifestyle may require time, patience and a lot of trial and error.

Once beginning a program, the Mayo Clinic suggests starting slowly and building up gradually. As stamina improves, gradually increase time and intensity. If you cannot exercise continuously for 30 minutes, break it up into two or three shorter sessions during the day. If you experience pain, shortness of breath, dizziness or nausea, you are pushing yourself too hard and you should take a break. If you are not feeling well, allow yourself to take a day or two or more, as needed, off from your routine. Being health-wise is also about knowing and honoring limitations.

On the following pages you will find short descriptions of popular types of exercise. When first beginning any new exercise program, it can be difficult and painful. It can even add stress to your life for a short time. However, eventually the positive transformations attained through regular exercise can provide opportunities for a great deal more participation and enjoyment in life.

Dancing

Many people enjoy watching other people dance. It appeals to something quite basic in human nature and can be invigorating and soothing and everything in-between. But participating in dance can also do these things while adding the benefits of a great low-impact, weight-bearing aerobic mind-body workout. And dancing also requires remembering dance steps and sequences and so it can improve memory skills.

In fact, according to AARP.org, researchers have found that people with Alzheimer's disease are able to recall forgotten memories when they dance to music they used to know. Some dance forms are more rigorous than others, but all styles of dance — including chair dancing for people with limitations — can provide physical, mental and social benefits including:

- Strengthens bones and muscles without harming joints
- Tones muscles
- Improves posture and balance
- Increases stamina and flexibility
- Reduces stress and tension
- Builds self confidence
- Provides social opportunities

In your community, you can find dance classes at dance schools, dance studios, health clubs, or community recreation centers.

Pilates

Pilates uses controlled movements in the form of mat exercises or equipment to tone and strengthen the body without adding muscle bulk. For decades, it's been the exercise of choice for dancers and gymnasts, but it was originally used to rehabilitate bedridden or immobile patients during WWI.

The Pilates mat program follows a set sequence, with exercises flowing from one another in a natural progression. Beginners start with basic exercises and build up to include additional exercises and more advanced positioning.

According to practitioners, the central aim of Pilates is to attempt to create a fusion of mind and body, so that the body will move with economy, grace, and balance.

As with any form of exercise, it is possible to injure yourself if you have a health condition or don't know exactly how to do the moves, so look for an instructor who is certified and knows the different ways to modify the exercises so new students don't get hurt.

For information, visit Pilatesmethodalliance.org or check with your community center for classes.

Yoga

Yoga is a unified mind-body alternative medical practice with ancient Indian origins. The various styles of yoga used for health purposes combine physical postures, breathing techniques, and meditation or relaxation to encourage flexibility, strength, stability and mobility.

According to the National Institute of Sciences' National Center for Complementary and Alternative Medicine's website (www.nccam.nih.gov), yoga is considered to be safe in healthy people when practiced appropriately. Studies have found it to be well tolerated with few side effects. People with certain medical conditions should not use some yoga practices, including people with spinal disc disease, extremely high or low blood pressure, glaucoma, retinal detachment, fragile or atherosclerotic arteries, a risk of blood clots, ear problems, severe osteoporosis, or cervical spondylitis.

Before practicing yoga, check with your physician. Be sure the instructor is certified and knows how to modify exercises for special needs and abilities. The 2002 National Health Interview Survey found that yoga is practiced for health purposes by nearly eight percent or 15.2 million American adults.

There are approximately 50 schools of yoga offering yoga therapy training in the U.S., and membership in the International Association of Yoga Therapists has more than tripled from 2003 to 2009. Yoga therapists include psychiatrists, psychologists and social workers.

For classes, check yoga studios, health clubs, and community centers. Yoga can look deceptively easy, but some classes that combine yoga postures with aerobics, Pilates or weights are quite vigorous.

Walking

The health benefits of walking are numerous, and it is the one physical activity that a great majority of people can safely enjoy. Besides reducing the risk of heart attack, stroke, hip fracture, glaucoma, type-2 diabetes, depression, relieving arthritis, strengthening bones, reducing blood pressure, and increasing good cholesterol, walking is wonderful for managing stress.

According to the AARP.org and WebMd.com, a 30-minute walk, five days a week is enough to reap benefits. You should be able to converse while walking. If you can't catch your breath slow down. When walking, start out slowly to warm-up, increase your pace in the middle, and slow your pace at the end to cool down. Stay motivated by walking with friends, family, coworkers, or pets. Set goals you can reach. For example, use a pedometer to count your steps. Wear it all day and try to take at least 2,000 more steps a day than you normally do and gradually increase your steps over time.

Water Aerobics

Think water aerobics is for sissies? Watch the WebMD video about the former marine instructor who teaches water aerobics (www.webmd.com/video/water-aerobics), and you will know that it can be for everyone—as long as the workout is tailored to the health needs and limitations of the participants.

Offering resistance training to build strength and endurance, water aerobic workouts usually combine a number of land-based activities such as walking or running backward and forward, jumping, dance movements, and exercises utilizing floatation devices. Classes are often conducted to

the tempo of music, and usually in shallow water, but some courses are in deep water and offer greater overall body resistance.

Water aerobics can be used to recuperate from injury and is great for anyone with arthritis because it puts little stress on joints. Exercise in water can also prevent overheating through continuous cooling of the body.

Water Aerobic classes usually last 45- 60 minutes. Contact your local community center or public swimming pool for classes in water aerobics.

Exercise for Relaxation: Improve Life's Quality

By Mona Reeva, Ph.D., M.P.H., L.C.S.W., Diplomate, and Doris Campbell, B.S., L.M.T.

Relaxing is important to good health and is essential to reduce tremor. There are many methods to calm our tremor through relaxation. The natural body cycle is a process of “tense and relax.” When we exert more tension than relaxation, our bodies are out of balance. This article is written to assist us in learning how to release muscle tension, relax the body and soothe the savage beast within.

Mona Reeva, co-author, is a psychotherapist in private practice in the San Francisco-East Bay Area of California. Professional experience coupled with her own journey with ET, Dr. Reeva first became aware of the way in which we attempt to cover or control our tremor. The most common method seems to be to tighten our muscles. It is as if by holding the areas the tremor will get better or at least not be visible to others. The opposite generally happens.

Tightening and holding the muscles in the area of tremor may be detrimental to reducing tremor and to general health. Feeling comfortable with the physical manifestation and learning how to relax these areas facilitates reduction in the tremor. This has been essential to Dr. Reeva's own ongoing process.

In our culture, the message of productivity is primary. In fact, it is so strong that we often lose track of our body sense and body messages. The body has signals that we have learned to ignore. Once we push against our own body's tolerance for extended and continuous activity, the tremor will worsen. Body-mind connection is not myth.

Dr. Reeva sometimes wonders if the purpose, not the cause, of her tremor is a reminder that she has gone beyond her body's tolerance for a particular activity. Dr. Reeva established relationships with practitioners who sooth the body. Since her head tremor manifested physically, she sought people who study, learn and practice in the realm of the body. She believes that tremor has physical, emotional and psychological components, and that it is difficult to differentiate between the tremor, its emotional aftermath and the emotions that influence its severity.

We understand that those who suffer with tremor do not cause it, and that if we work towards wholeness, than the notion of cause becomes less important. Clearly, if biochemical imbalance, genetics, environmental pollutants or other physical reasons cause tremor, the cure may be more readily treatable so we should research to uncover causation. Self-blame, though, is detrimental to wellbeing. As we begin to look beyond self-blame, taking responsibility for healing becomes the most significant part of feeling better. Healing is not the same as curing the tremor. Healing is about improving our quality of life and experiencing life as worthwhile.

Noting the physical manifestation, Dr. Reeva found Ms. Doris Campbell, who is a licensed massage therapist. Ms. Campbell, a former engineer, studies on a continuous basis, an attribute most notable. She is smart, funny and caring, a dynamite combination, and she is open to consulting with her teachers about symptoms she is puzzled about – another admirable attribute. We began to work together a few years ago, and while occasionally Ms. Campbell does full body massage, we have concentrated on cranial sacral work. Cranial sacral work assists in aligning and relaxing the tissue around the area of the head tremor. Because of Ms. Campbell's abilities, Dr. Reeva sought her as co-author.

Another person who works with orthopedic problems is Ms. Arlene Suda, a licensed physical therapist with many years of study and practice.

Ms. Suda is a master at understanding the mechanics of the body. She has an uncanny ability to shift and align the spine and other areas of the body. Her work reduces pressures within. Ms. Suda makes suggestions for appropriate exercises that promote healing aches and pains. She graciously consented to review our Exercise for Relaxation program.

There are many methods that will help us learn how to relax, soften the muscles and balance into our lives such as massage, physical therapy, Rosen Method, Feldenkrais, Hannah Somatic Stretching, Yoga, Tai Chi, Chi Gong, etc.

Choosing a Masseuse/Body Worker

Choosing a body worker is a challenging undertaking. The following is designed to help find words to describe what kind of experience you might like to have.

Type of Touch

- With lotion on the skin
- On the skin without lotion
- On the body while you are wearing clothing
- Above the skin in the space around the body

Intention of the Work

- Relaxation
- Movement of soft tissue (muscles, tendons, fascia, etc.) and bone for healing
- Stretching of the body
- Reduction of pain
- Movement of lymphatic system
- Work on the meridians of the body (Chinese medicine)

Depth of Touch

- Deep tissue
- Lighter tissue
- Subtle (extremely light touch)
- Energy (extremely light or just slightly above the body)

While these lists are not exclusive, they create a frame of reference from which you can begin. One approach might be to select a few of the above listed criteria that are important to you. Call a few practitioners and ask if their work is consistent with what you have chosen as vital.

Searching your neighborhood for practitioners will reap reward. Often classes are offered through adult education, senior centers and with private practitioners. The best way to find a body practitioner is through a referral. Ask friends and associates if they are receiving massages, and if so, get the name of their body worker. Most important, as with choosing any type of therapist, is that you feel there is a great deal of comfort and trust with that person.

One intention in writing this article is to acquaint readers with a number of exercises that can be undertaken in your home, in the garden, or elsewhere. They are designed to be simple, easy to follow and will add to your personal repertoire of relaxation methods. We recommend doing only one series at a time. Add to your routine on a weekly basis.

An important aspect of exercise is to develop a series that is doable. This means that they feel good, are not stressful and accomplish relaxation. We want tools by which we can relax. Part of this process of relaxing is to begin to relearn body signals that tell us when to slow down, to stop and take breathers with our goal oriented productivity. Sometimes, especially initially, we may find we like or gravitate to certain exercises, leaving out others. Frequently, the ones we leave out are addressing the area(s) of greatest tension! Here, we offer directions for “Exercise for Relaxation” that may change your life. Arlene Suda, physical therapist, has reviewed these.

Preliminary Guidelines

- Where there are pre-existing injuries, check with your health care provider
- Find a place that is peaceful
- If you enjoy relaxing and soothing music, play some while practicing
- Set aside a specific amount of time on a regular basis
- Ask others in your home to not disturb you for the length of time you choose to engage in your exercises and relaxation. If you feel better doing this work with someone, then ask another person to practice with you
- Consider turning off the ring of your telephone
- Wear loose clothing
- Work on the floor - a mat is useful, or in a straight backed chair or a stool. If you feel physically strong, some of these can be done while standing
- Perform each exercise slowly. Rushing to be done increases stress and tension.
- Repeat each exercise as desired
- Take note of where your body is tense when you begin
- Remember to breathe while doing each exercise
- Allow time before and after to breathe and rest
- If you feel yourself tensing, rest for a few minutes
- When you finish, note how your body feels
- In all exercise, move only as far as is comfortable without extreme or hurtful strain

About Breathing

While practicing these stretches, use your breath. Initially, this means just to pay attention to your breath, noticing when you are straining. If you notice that it is hard to breathe, allow yourself to slightly release from the tension of the exercise. If you find yourself straining by making faces or noises in order to hold the position, allow yourself to slightly back out of the position, release the tension and breathe clearly.

The breath can also be used to help you increase your range of motion in a particular position. Once you are in the stretch or position of tension think in your mind of sending your breath to the place in your body where there is tension or a slight pain from the position. Remember that pain is not the objective. Back out of the position until there is less pain or a general feeling of stretching and hold. Think of your breath as providing more space for release and motion to occur.

When trying to open and create space in the body we move back slightly out of the position on inhale and further into a position during exhale. This is to allow the space the body needs to hold the breath on inhale, and on exhale, taking advantage of the new space created by the exiting breath to increase the stretch.

In general, attention on the breath is a form of meditation. It is important to watch our breath. Often we may have shallow breathing, or we may hold our breath while we are concentrating and trying to do the exercises properly. When you notice that you are doing either, take one or two deep slow breaths and allow natural breathing to begin again. Feel your body relaxing.

The breath can be used to gauge the length of a movement or how long you stay in any one position. Start by holding a position for two to three breaths and see how it feels to hold for more. Note: At rest, most adults breathe 15 times per minute. Allow yourself to move on from there. You can also gauge some positions by your heartbeat. Notice if, and in which positions, you can sense your pulse or your heart.

About Repetitions – Getting In Touch With Your Body

The following are three series of exercises. Each one is a set and may be repeated to increase the state of strength and relaxation. Remember that it is important to start slowly, building strength as you do each exercise in each set. This series will allow repetition of each particular exercise and enable you to do more.

Reducing Negative Self Judgments

An important purpose of these exercises is to help each of us become familiar with our normally held tension. For example, while writing this article, Ms. Campbell noticed how high her shoulders had become. They almost touched her ears in her effort to concentrate. As she became aware of this tension, she began to repeatedly drop her shoulders to release the tension that had developed. Ms. Campbell indicated that she tries to not have a scolding air with herself such as “Oh no, these shoulders will never get it right.” What she does is to acknowledge whatever state she is in: “Good, I noticed that I had some tension, and it does feel better released.”

“Tense and release” is the normal cycle for working muscles. The muscles are bathed in a fluid, which contains chemicals that deliver the messages of when to tense. The contraction cycles help the body to move blood, lymph and the fluids between cells through the action of pumping. When we experience tension there is an opportunity to create release and complete the cycle that supports the movement of these fluids.

Series A

1. Shoulder Shrugs - Start: Either sitting, standing or lying on your back, arms at your sides, eyes forward. Begin by raising your shoulders upward towards the top of your head, moving them as far as they can go, tensing your muscles. When you feel your muscles beginning to tire, release your shoulders, letting them drop and relax as completely. Variation: Do one shoulder at a time.
2. Shoulder Rotation - Start: Sitting or standing, arms at your sides, eyes forward. Begin by rotating your shoulders up, back, down and forward. Repeat this rotation several times until you feel the release of tensions. Then reverse the direction, and as before, repeat.
3. Head Turning - Start: Either sitting, standing or lying on your back eyes forward. Begin with your shoulders relaxed. Keeping your chin level, lead with your eyes, moving them to the

right and turn your head to the right as far as it will go comfortably without strain. Now hold your head for a few seconds in that position. Return your head to center position and slowly relax, noting that your shoulders are in their dropped and relaxed position. Repeat the same rotation of your head and eyes to the left. Note that if you do this regularly, your head will begin to rotate more easily, and your range of motion will increase. Variation: Start the exercise by moving your eyes to the extreme left and then rotating your head to the right. When you switch directions, allow your eyes to move in the direction opposite from your head.

4. Arm Turning - Start: Either sitting, standing or lying on your back, arms straight and raised to shoulder level, palms facing down. Begin by rotating your arms in the shoulder socket first forward and then backward one direction then the other. Although your arms are straight, they should be soft and flexible at the elbow joints.
5. Hand Tightening and Relaxing - Start: In any position where you have enough room to move your hands. Begin by tightening your hands into a fist, holding them for a moment then releasing them. Allow a few moments to completely relax.
6. Waist Bend, Head Release - Start: In a standing position, feet shoulder width apart (6-8 inches), or sitting in a chair with your legs 6-8 inches apart at the knees with feet solidly on the floor. Begin by bending at the neck and folding forward, one vertebra at a time. Take your time. Bend forward as far as you can comfortably go, allowing your arms and head to hang loosely. Release everything completely so that you feel the loosening in your neck, shoulders, arms and hands. Rest for a moment or two. Slowly raise yourself upright, either in your standing or sitting position.

Series B

1. Wrist Circle Rotators - Start: Sit, stand or lie down with arms at sides, elbows bent. Begin by rotating your wrists slowly first in one direction then the other. You want to move slowly in areas that feel stiff or resistant. Remember to breathe.
2. Wrist Table Stretch - Start: Sit or stand no more than a foot in front of a stable object that you can push against. Begin by bending your elbows in front of you, with your upper arms held against the body and turning the hands so that your palms are facing upwards. Now, place your fingertips on the stationary object (table edge, wall) in front of you and gently bend your wrists leaning into the stationary object, so that the palms of your hands are moving towards the object. Eventually in this move towards the object, your wrists will be parallel with your body. Hold this position, gently breathing for a moment or two, and then release and repeat.
3. Ear to Shoulder - Start: Standing or sitting with head facing straight in front of you. Begin by allowing your head to drop towards your right shoulder until you feel a stretch in your neck on the left side. Keep both shoulders level. Do not lift your shoulder to meet your ear. Move slowly and deliberately. Hold for a few moments and slowly come back to the starting position (head forward). Then repeat the exercise on the left side.
4. Face Stretch – Scrunch and Release - Start: Standing, sitting or lying on your back. Begin by drawing the face muscles towards center, looking like you might if you had just bitten into a nice juicy lemon. Hold this position for a few moments and then release. Pay attention to whatever areas of your face that might still be hiding tension. Repeat: This is a great exercise to do while sitting in traffic.
5. Face Stretch – “O” - Start: Standing, Sitting or lying on your back. Begin by bringing your mouth and eyes into a big “O” position, as if you had just been surprised. Let the jaw drop

so that you get a nice big circle in the mouth. Hold then release, again, being aware of any areas where tension might be lingering.

6. Hip Tilt - Start: Lying on your back, knees gently bent, feet flat on the floor and roughly hip distance apart (4-6 inches). Begin by flexing the hips back and down bringing the lower back and spine into contact with the floor. After a moment, gently release the hips, pulling the lower back up off the floor and extending the spine. Breathing out assists this motion when flattening the spine against the floor tightening the stomach muscles. Then exhale and release the abdomen while extending the spine.

Series C

1. Wrist Flex and Extend - Start: Standing or sitting. Raise your arms so that they are parallel to the floor with palms facing the floor. Begin by bending your wrists downwards and draw your hands into fists feeling the stretch along the back of your hand. Be sure that your shoulders are not raised. Hold for a moment and then release, straightening the hand and bending the wrist in the opposite direction so the fingertips are headed for the ceiling. Repeat several times.
2. Chest/Shoulder/Arm/Wrist Stretch - Start: Standing near a wall or a doorway. Raise one arm so that it is at or nearly at shoulder level. Place palm of the hand on the flat surface of the doorway or wall with the wrist bent so that the fingertips are facing away from you, i.e. backwards. Feet should be shoulder width apart, one in front of the other. Begin by bending the forward knee so that you begin to feel a stretch across the shoulder and chest. You may also feel the stretch in any section of the arm. If you are using a wall instead of a doorway, you may need to twist in the waist in order to achieve a stretch. After a few moments, switch arms. Variation: Alter the height of the arm on the wall or doorway and note how that changes the location of the stretch.
3. Neck/Head Stretch - Start: Standing or sitting. It is important that the chest and abdomen are held in a firm and tightened posture. Begin by bringing your eyes downward and then allow your head to follow. Bend forward so that your chin is headed for or touching your chest. Do not allow your spine to bend below your neck and keep your shoulders soft and straight. Relax as much of your head and neck as you can. Then, slowly bring your head and eyes up so that you are again facing forward. Repeat as desired, feeling the stretch and holding your stretch softly.
4. Stretch of Life- Back and Leg Stretch - Start: In a standing position facing something that you can rest your leg upon, between knee and waist height (a little higher height for those of you who are more flexible). If the step stool or chair is a little higher than comfortable, bend your knee a bit more. Face forward with the heel of one of your feet resting upon your object. Begin by gently bending forward at the waist bringing your arms forward with the objective of touching your toes. Ideally, you want to keep your knee straight, but that is not necessary in order to feel a stretch. Hold this position for a good long time so don't go too far in the beginning. Really use your breath on this one to let yourself ease into the stretch. On inhaled, bring yourself slightly out of the stretch by straightening at the waist. As you exhale, work to straighten your knee while trying to get your hands close to your feet and head closer to the knee. Work on this gently for at least 10 breaths and then release, slowly coming back to standing one vertebra at a time. Repeat, using the other leg. This is a great stretch because you can easily measure your progress. You are addressing the important muscles in big muscles in the back of the leg. Caution: If you experience pain, tingling or numbness, slowly back off from this exercise as you have stretched too far.

5. Trunk Twist – Do The Twist - Start: In a standing position, near a wall. Face the wall and place both hands, palms forward, fingers facing upward, against the wall. This can also be done from a sitting position by placing a stool near the wall so that you can the back and the comfortably place your hands on the wall as above. Begin by slightly leaning against the wall. Begin to move both feet in the same direction so that your toes are not facing the wall. Taking small steps (carving an arc with the movement of the feet) continue to turn your feet until you feel a good stretch in the trunk. Allow your head to look back over your shoulder in the opposite direction than the feet. This is a good for a few moments as you are working so many muscles. Release the stretch a bit on inhale and really work to move into the stretch on exhale. Remember to slowly return to center. Repeat on other side.
6. Leg Bend and Stretch - Start: Lie on the floor on your back with arms at your sides. Place a pillow under your knees. Begin by raising one leg, bending it at the knee and tucking it to your chest by hugging it with stretch to hold your arms. Allow the other leg to rest gently on the floor holding the raised leg just tightly enough so that the straight leg can continue to rest on the floor. Stay in this position for several breaths. Feel the press of your leg against your abdomen. Allow the raised leg to return slowly to the floor and raise the other leg and repeat. Again, stay in the position for a good time, relaxing and noticing the press of the breathing against the pressure of the bent leg. Finally, bring both legs up and fold them against your chest, hugging your arms around your legs and holding for a nice long time. Use the opportunity your tremor has presented to improve the quality of your life.

You have now completed this entire series of stretch and relaxation exercises. Consider repeating two to four times per week. Keep track of your physical and emotional state of being before, during and after the exercises. Your tremor may or may not feel stressed by these movements, depending upon your individualized tremor and response to exercise. If they do, and the tremor has increased, give yourself sufficient time until your next activity to allow the tremor to return to its normal and/or improved state. If you find that any of these stress your tremor to the point that the tremor has increased for a long period of time, slow down the process or skip that particular movement. Perhaps you need to do less each time. For many, this series of stretches and relaxation will loosen your body, increase how you feel positively about yourself, and generally help you to feel fit.

Relaxation aids in coping with the stressors we find in daily living and in those that are part of the unusual events that occur in life. We all experience these events, as they are part of life. Trauma, losses, shocking experiences, loved one's pain, excitement, and tension all have a profound impact upon us whether we acknowledge them or not. Our bodies carry a great deal of stress that results from these kinds of situations. How we face our fears, our pain, how we find the inner strength to assist us in coping and working through these has influence on the quality of our lives. Inner strength and coping skills come not only from our own history and experience; they also emerge when we join with others in mutual support and caring. Learning comes within groups, formal or informal.

Learning coping skills also comes from good friends, a partner, and/or relative with whom you feel an inner resonance of communion. Spiritual relationships can be of great assistance as well. You have the power to choose the opportunity presented by your tremor to improve your quality of life. We trust that you will make this happen.

Meditation and Stress: Help for People with ET

Tremor Talk

Meditation has long been a part of Eastern religious practice. Now, according to the *2006 Annual Report of the Society of Neuroscience*, Western neuroscience is finding “measurable positive effects” of meditation that could be beneficial as a coping tool for some people with ET.

Recent research conducted by such prestigious research centers as Harvard Medical School, The University of Massachusetts Memorial Medical Center, and The University of Wisconsin has found that meditation, or what some neuroscientists call mindfulness, brings about feelings of well-being and emotional balance, increases the mind’s ability to focus and reduces stress, anxiety and pain.

Stress and anxiety present challenges for most people. They are particularly troublesome for people with ET because of the tremor-aggravating effects of adrenaline, the neurochemical released by the body during stress. Any method of reducing the effect of stress on the body and so limiting the effects of adrenaline is beneficial for people with ET.

According to the Society of Neuroscience, 10 million Americans currently practice some form of meditation. Historically associated with the attainment of higher spiritual goals, secular meditation practices use techniques that focus or cultivate one’s attention or awareness through the repetition of a word, sound or phrase or through breathing.

Specific research findings on the positive effects of meditation over a long period include:

- Higher level of gamma band rhythms, brain impulses associated with higher mental activity such as attention, learning and conscious perception
- Increased attention skills and the ability to stabilize the mind
- Thickening of the brain in regions involved with attention and sensory processing
- Thickening of the brain’s outer cortex, an area involved in the integration of emotional and cognitive processes
- Increase in antibodies, proteins used by the immune system to fight off bacteria and viruses.

Although no clinical research has been conducted on the effects of meditation on the tremor of ET, Elan D. Louis, MD, MS, professor of neurology and epidemiology, College of Physicians and Surgeons and Mailman School of Public Health, Columbia University, New York, says meditation may help. “Stress and anxiety can certainly temporarily exacerbate an underlying tremor disorder. To my knowledge, it has not been rigorously tested whether stress reduction through meditation temporarily lessens tremor. That is, I know of no clinical trials. However, meditation could, at least in theory, provide some mild temporary reduction in tremor.”

You can get more information about the effects of meditation as well as information about incorporating meditation or mindfulness into your daily life as a coping strategy against stress and anxiety through The Center for Mindfulness in Medicine, Health Care, and Society (CFM) Stress Reduction Program, University of Massachusetts Medical School. The program was developed by Jon Kabat-Zinn, Ph.D., and professor emeritus of the medical school in 1979.

Coping with ET: Tai Chi

Tremor Talk

Tai chi (pronounced “tie chee”) originated in China during the 12th century AD as a martial art, but today is a mind-body practice in the West. Also known as “moving meditation,” tai chi combines slow and gentle movements with deep breathing and meditation.

According to The National Center for Complementary and Alternative Medicine (NCCAM), part of the National Institutes of Health in Washington D.C., tai chi is gentle enough to be practiced by persons of all ages and varying disability.

Tai chi was developed as a set of exercises that combined imitation of the movements of animals for strength with flexibility and suppleness. While the entire body is relaxed and in an upright position, each exercise is performed at a slow uniform speed and easily flows into the next exercise. The entire body is in motion at the same time during exercise.

Besides movement, tai chi also incorporates deep, relaxed breathing and concentration, or meditation.

Although, according to NCCAM, there is no research to date that shows how tai chi affects the body, many practitioners believe that it promotes well-being, calmness and awareness; improves balance, muscle strength, coordination and flexibility; aids digestion; benefits bone health because it is a weight-bearing exercise; eases pain and stiffness; improves sleep; and modifies easily for older and disabled people.

Tai chi is practiced within a group of people or individually. It is recommended when learning tai chi that a teacher be consulted so that proper technique is instilled. Many community centers and schools offer courses in tai chi, and teachers can be found in your community by consulting a yellow pages or other directory.

As with any exercise, always consult your physician before beginning the practice of tai chi.

Biofeedback: Recognizing the Body's Need to Relax

Tremor Talk

For as long as Madonna Ozman has been teaching people to relax, as a certified biofeedback practitioner and registered nurse, she has noticed that many Americans tend to be bundles of tensed muscles and frayed nerves.

Tensed muscles and frayed nerves can have a negative impact on many health conditions. But Ozman believes that the ability to relax, learned through biofeedback, can overcome many of these negative influences. Research findings on biofeedback and ET tend to agree that for those people who see an increase in tremor as a result of stress, biofeedback may be of benefit.

A 1974 study, (Wake, et al) found progressive muscle relaxation was helpful in reducing ET. In 1995 and 1999 studies (Chung, Poppen & Lundervold and Lundervold, Belwood, Craney & Popper), ET and the disability it caused were reduced utilizing Behavioral Relaxation Training (BRT).

Tremor severity was reported reduced by 47 to 66 percent. In 2004, a study published in *Applied Psychophysiology and Biofeedback*, found that teaching BRT techniques along with EMG Biofeedback Training lead to decreased tremor during in-session assessments, self-reported assessments at home while engaged in activities of daily living and in follow-up in-clinic assessments. In addition, it was found that the more relaxation training during sessions, the greater overall tremor reduction. The researchers concluded that relaxation and EMG biofeedback training “hold promise for...adults coping with tremor and related disability.”

Biofeedback uses instruments to measure body temperature, muscle tension or brain waves, and requires a therapist to operate the instrument, explain what the readings mean, and to teach people relaxation techniques such as deep breathing, visualizations and meditation.

Biofeedback treatment sessions are usually held one-to-one with the patient and therapist. Therapy takes place in a comfortable chair in a distraction free room. Each aspect of treatment is thoroughly explained to the patient.

There are three common forms of biofeedback: EMG, EDR and EEG.

Electromyography (EMG), measures muscle tension and has been used in studies conducted with persons who have ET. EMG biofeedback training helps people learn to relax and reduce tension of overly contracted muscle groups.

Electodermal (EDR) measures the skin's ability to conduct electricity caused by emotions by measuring peripheral skin temperature. During stress, blood vessels constrict and blood cools. Overcoming the stress through relaxation techniques registers as an increase in skin temperature.

Neurofeedback or electroencephalography (EEG) measures brain wave activity. Sophisticated biofeedback instruments transfer information from the body, amplify it, and convert it into a readable signal. The instruments can be programmed to beep or light up a display as a person begins to relax and can confirm that a certain level of relaxation has been achieved. A training baseline, or

beginning level, is set and progressively more challenging levels occur during the course of treatment.

As new relaxation skills are learned, many people feel more confident that they can, to a great degree, control body responses that they once thought were completely uncontrollable. This ability requires regular practice between sessions in order to learn. In a typical session, instruments are used first to set a beginning point for the session. Next, therapist and client may review the stressors of the week, discuss symptoms, teach/learn new coping skills and discuss how to integrate these strategies in a lifestyle. Most of the session is then spent on practicing relaxation exercises, including progressive muscle relaxation, breathing exercises, visualization, imagery, autogenic training, or meditation. These can be narrated by the therapist or provided by CD that a client can take home. Many therapists provide clients with educational materials.

According to Ozman, biofeedback often gives mixed results. “There is a large psychological component to biofeedback,” says Ozman. “Success is dependent upon the client’s commitment to incorporate regular biofeedback practice into their everyday life.”

Reimbursement for biofeedback therapy varies by individual insurance policies, says Ozman, with many practitioners using a sliding fee scale to make biofeedback training affordable for everyone.

To find a certified biofeedback practitioner, ask your doctor for a referral. You can also go online to the Biofeedback Certification Institute of America’s website (bcia.org) to locate a certified practitioner in your area.

Mindfulness and Essential Tremor

By Dr. Monique Giroux

What is the next step when medical or surgical therapy does not control all of the symptoms of essential tremor? You may be surprised to hear that you have the answer to this common question within you. Understanding how your thoughts, ideas and experiences affect your tremor is the next step toward coping better with ET.

ET symptoms come and go, often without warning, and the mind is on constant alert to monitor for these changes. This internal alarm system brings us to heightened awareness with enhanced focus on tremor as our mind constantly monitors for a negative change. If not reset, this internal alarm system sets up an ever stronger daily reminder that you have a problem and things are just “not right.”

As a result, the experience of tremor becomes coupled to negative feelings and sets the stage for distress, worry, anxieties and fear - about how you feel now and how your tremor may change in the future. These feelings and your tremor symptom become a daily focus as the mind becomes preoccupied with negativity and fear. Left unchecked, negative thoughts can impact the rest of the day and/or interactions with other people.

Preoccupation with the negative impact of symptoms can disrupt the ability to live in the moment. The mind translates sensations from the body and can affect these very sensations. Similarly, the mind can translate the power of healing over the body.

What these observations tell us is that the power to heal can come from many places. Medication and surgery can treat symptoms and restore many of the body's functions that can be lost from disease. However, modern science and medicine may not be enough and does not treat the experience of the person in the moment of tremor. The power to heal, to be and feel whole, to know that you are OK, comes from you.

Mind Power

Our mind can be our strongest asset or biggest obstacle when it comes to feeling better. Taking steps to change our habits for healthier living toward a focus on healing requires a commitment from the mind. Your mind will influence how you feel with tremor.

Think about a time when stress was a major part of your life. Did your tremor get worse? Or perhaps tremor started during that time of stress? Stress can amplify tremor symptoms. Over time, repeated or runaway stress can change how we think and react and can negatively impact our health. This effect can also work in the opposite way. You might think of your favorite place, relaxing on vacation or other time when stress was at a minimum. How did your tremor respond? Was it less intense? Less bothersome?

Chronic stress negatively affects our body and mind. Conversely, low stress and enjoyable experiences positively affects our body and mind. Fortunately, we can modify how our mind and brain react to stress and reduce the impact on disease. A well-known and measurable example of this is the placebo effect. Innovative research shows that expectations we bring to a treatment will in part determine the strength of the placebo effect.

Hope, belief, and positive expectations that a treatment will work will increase the chances it will do so.

Mindfulness

Mindfulness therapy can reduce the negative impact of stress and sharpen the mind's potential for personal healing. It is a way to stay in the present moment, engaging in life and living life as fully as you can. Mindfulness is often defined as being present, in the moment, with intention in a non-judgmental way. Being mindful allows the mind to see things for what they are and then to let the thoughts go, instead of letting our thoughts control our body, mind and brain function. So how can this help you?

Being present in the moment helps one become aware of the many thoughts, ideas, distractions and assumptions that drive our behavior and impact our wellbeing on a daily basis. Our mind can become distracted by making assumptions, jumping to conclusions, and forming judgments that can have a negative impact on our thoughts. Becoming aware of these roaming or ruminating thoughts is the first step to living in the moment. Observing your thoughts without judgment helps you become more aware of your body, sensations, emotions and surroundings.

This technique is particularly powerful for people living with tremor. For example, when tremor increases you now have one of two choices—to react or respond. These choices at first glance may seem very similar but they are indeed very different in mindset and outcome.

React

By reacting to tremor you are giving control to the spontaneous thoughts and feelings that affect your behavior and how you feel. Your spontaneous thoughts are unique to you but often include judgments such as:

“Tremor is running my life.”

“My disease is worse.”

“I must fix this or stop this now.”

“If only I didn't have tremor everything would be fine.”

“What if someone sees my tremor?”

These reactions can lead to a “snowball effect” and further worsen stress leading to yet worsening tremor.

Respond

By responding to tremor you are no longer reacting but instead observing and choosing how to respond. How you respond is up to you.

“This problem will pass.”

“I will sit for a few minutes to let this pass.”

“I have experienced tremor before...”

“My tremor is in my hands but is not me.”

By choosing your response you gain a sense of control and change the relationship you have to your disease or symptom.

This does not mean that you have given into your symptoms or enjoy the fact that you have tremor (or any other medical condition or problem in your life), but it does reduce the downward spiral that can happen when life gives you problems or obstacles. Tremor is a good example of this downward spiral or “snowball effect.”

Practicing Mindfulness

Mindfulness can be an informal spontaneous practice or a formal and structured approach.

A spontaneous and informal practice is present many times, perhaps endless number of times throughout the day. These daily experiences and moments are used as an opportunity to take notice and be present. The next time you awaken to a sunrise, hear the laughter of a child or are stuck in traffic you can practice mindfulness. Simply bringing awareness to the moment, identifying any feelings the experience brings on and how this influences us.

Road rage is a good example of how our behavior can be driven by emotions and thoughts without our full awareness. This reaction will cause us stress, increase aggressive driving and affect our mood, behavior and perhaps tremor for the entire day.

Mindfulness is also practiced as a formal exercise with meditation. One way to practice mindfulness meditation is to sit quietly and calmly but with attention and awareness. Bringing your attention to your breathing is one way to focus your attention. During this time you will simply observe your thoughts, feelings sensations and perceptions, and let them pass without judging them, labeling them or controlling them. Meditation is a practice and as such, the benefits will improve with time and practice. Over time, you will learn to apply these skills to everyday events.

When something happens that requires your attention you now have the insight to respond and not simply react. Described in another way, tremor no longer drives your reactions as you can intentionally decide how to respond to it. This element of control and intention can liberate you from the hold of tremor on your emotions.

Mindfulness is not without risk that can come from paying attention to and increasing your awareness of thoughts and behavior. Individuals with psychotic disorders, significant depression or post- traumatic stress disorder should meditate under the care of a mental health professional. Mindfulness can be a helpful tool to enhance the effect of medicine and surgery on tremor control. The next time your tremor feels “out of control,” take a moment to reflect, and know that you have control in how you respond.

Dr. Monique Giroux is co-founder of the Movement and Neuroperformance Center of Colorado in Englewood, CO and medical director of movement disorders for Swedish Medical Center. She is board certified in neurology. She has specialized training in Botulinum toxin (Botox) for dystonia, pain and spasticity as well as deep brain stimulation for essential tremor, dystonia and Parkinson’s disease. Dr. Giroux is actively advancing quality care and wellness for the community in her positions as medical director of the Northwest Parkinson’s Foundation wellness center, medical faculty for the National Parkinson’s Foundation Allied Team Training Program and project leader for the National Parkinson’s Foundation Care Center Consortium project. Along with Sierra Farris, PAC she co-authored Every Victory Counts: Essential Information and Inspiration for a lifetime of wellness with Parkinson’s, produced in collaboration with the Davis Phinney Foundation. Please visit www.centerformovement.org for more information about the center and these programs.

Helpful Hints for a Good Night's Sleep

Tremor Talk

Fatigue can make tremor worse, so a good night's sleep is important in managing ET. But sleep does not always come easily. Short of taking sleeping pills, which potentially can create dependency, some people – the elderly, parents of young children, and those under a great deal of stress, among others – find it almost impossible to fall asleep or to stay asleep.

Following good sleep habits is the first step to better sleep, according to the University Of Maryland School Of Medicine. Here are suggestions for better sleep:

- Go to bed at the same time every night, and get up at the same time every morning, even on weekends and holidays.
- Don't nap, even during late afternoon when most people feel sleepy. If you must nap, sleep only 30-45 minutes.
- Cut off alcohol consumption four to six hours before bedtime. Just as there is a rebound effect when someone has more than one or two drinks to calm tremor, there is a sleep rebound effect from alcohol. When alcohol levels in the blood drop while sleeping, it creates a stimulating effect.
- Avoid caffeine from coffee, teas, sodas, and chocolate for four to six hours before bedtime.
- Sleep on comfortable bedding. What makes for comfortable bedding is greatly a matter of personal preference, but it is worth finding what makes you comfortable and then using it.
- Sleep in a cool room. If you are too hot or cold, it is hard to fall asleep. Studies have shown that 65 degrees is the average ideal sleeping temperature, but this is likely a matter of personal preference.
- Eliminate distracting noise and light. While some people may find the sounds of quiet music or waves relaxing, others will not. Some people can sleep in the middle of a sunny afternoon while others need blackout curtains on their windows at night. Whatever is distracting, eliminate it.
- Bed is only for sleeping. Don't work or watch television in bed.
- Relieve anxiety and reduce tension through relaxation before bed. Try a warm bath, deep breathing exercises, or reading.
- Don't allow yourself to worry while trying to sleep. This is easier said than done for many people. One technique to stop worrying at inconvenient times is to allow a short period of time during the day to worry and deal with issues. You might even try writing them down and setting them aside to banish them from your mind.
- Don't lie in bed awake. If you don't fall asleep within 15 to 30 minutes, get up and read a book or do something relaxing or boring. Do this also if you wake up in the middle of the night and can't go back to sleep. Do not watch television, work, or exercise.

If you continue to have problems falling asleep or staying asleep, talk to your doctor.

Music for Therapy and Relaxation

Tremor Talk

According to the 20th century Russian composer Igor Stravinsky, music provides order to chaos through structure, and it is this structure that “produces in us a unique emotion having nothing in common with our ordinary sensations and our responses to the impressions of daily life.”

Whether it’s music’s structure or the unique emotion it elicits—or both—researchers have found that music provides humans with numerous benefits, including decreasing depression and anxiety as well as boosting the immune system. In addition to providing a driving beat to energize someone out on the dance floor, it also gently rocks that person into a quiet state that helps them to relax.

Relaxation isn’t just a state of mind, but is a slackening of muscles and calming of the central nervous system. In theory, relaxation can help people manage their ET better. Research has found that music therapy can work well to help people relax by changing a person’s brainwaves through tempo, the beat of the music.

“Research has shown that music with a strong beat can stimulate brainwaves to resonate in sync with the beat, with faster beats bringing sharper concentration and more alert thinking, and a slower tempo promoting a calm, meditative state,” states Stress Management Guide Elizabeth Scott, MS, in her article, *Music and Your Body: How Music Affects Us and Why Music Therapy Promotes Health* on About.com.

Furthermore, in an article entitled *Music Therapy to Benefit Individuals with Parkinson’s Disease* by Dr. Concetta M. Tomaino, DA, MT-CS, LCAT, it is reported that many Parkinson patients who focus their attention on the beat of a specific music type display better movement and report an easier time performing successive tasks.

The Institute for Music and Neurologic Function, a nonprofit member of the Beth Abraham Family Hospital Family of Health Services, is located in The Bronx, New York City, NY. The Institute conducts clinical research studies on the effects of music on the human condition with the aim of restoring, maintaining and improving human physical, emotional and neurological functioning. Dr. Tomaino is the executive director and co-founder of the Institute.

The first step in music therapy treatment, according to Dr. Tomaino, “is to use music that a client identifies with to help them relax. That music may be different for each person, but—based on the music therapist’s evaluation—the therapist can create listening programs that will help reduce anxiety and enhance relaxation. The second technique is an interactive music therapy program.

“In an interactive music therapy situation, a therapist trained in music psychotherapy will engage the client actively in understanding the triggers that cause them to be anxious or nervous or exacerbate the tremors,” Tomaino explains.

The third technique is an intensive music therapy program, with which, says Dr. Tomaino, “The Music Therapy Institute at Beth Abraham Hospital has had significant success. However, this program is significantly reliant upon the determination of the patient to stay motivated.”

The actual type of music used in therapy is based upon an individual's preference. Some people are more sensitive to jazz while others are more sensitive to classical. However, a patient cannot limit therapy to just a single or few pieces of music or they lose their initial effectiveness.

“Our brains tend to get used to things,” explains Dr. Tomaino, “and because of the acclimation, the brain tends to just ignore it so the therapeutic effect isn’t as great as the first time they listen to it.” However, Dr. Tomaino also notes that if a person associates positive emotions with a particular piece of music, they will always have those positive feelings while listening to the piece, though it may lose its therapeutic influence.

Persons with ET, suggests Dr. Tomaino, can search for a music therapist who specializes in movement disorders by requesting a referral from the American Music Therapy Association (AMTA) at www.Musictherapy.org.

Finally—under no circumstances, stresses Dr. Tomaino—should a person in music therapy stop taking their medications without first talking to their physicians. Music therapists should also be made aware of medications being taken by patients. The more a music therapist knows about past and current treatments, the easier it is for them to create a program around the patient's individual needs.

Nutritional Issues in Individuals with Essential Tremor

By Sara Salles, D.O.

Essential tremor remains the most common movement disorder which affects men and women equally. The prevalence of ET continues to increase with age with mean age at presentation of 35-45 years of age. At the present time, the pathophysiology of ET is not well understood. Limited medication options are currently available in the treatment of ET. Additionally, there have been few clinical studies describing dietary and nutritional relationship in individuals with ET. Several studies have described decreased body mass index (BMI) compared to the general population, which may place individuals with ET at risk of nutritional deficiencies.¹

Due to tremor and decreased control of the upper extremities, some individuals may either abstain from certain foods or require a longer period of time to feed themselves, which may lead to decrease in overall intake from the increased effort of self-feeding. Additionally, it has been postulated that the decrease in BMI may also be associated with increased energy expenditure just from the presence of the tremor itself. From these studies, it has been recommended that BMI should be measured and followed in individuals with ET and other movement disorders. Furthermore, these individuals would benefit from consultation with a dietitian to ensure appropriate intake and to further avoid weight loss, which may lead to a decrease in bone density which can increase the risk of fractures, and to decrease long term mortality.¹

Other studies have also examined the relationship of dietary protein consumption in the form of meat and its increase in blood harmane levels. Harmane is an alkaloid that is thought to cause tremor. To date, there is still not enough evidence to prove that individuals with ET actually have a problem metabolizing of this chemical, thus making the levels higher in the blood when compared to normal controls.² Given the limited number of studies, there are no specific recommendations to avoid foods with large amounts of harmane.

Additionally, several studies have questioned the possibility of nutritional vitamin deficiencies such as vitamins E and C but again no specific link has been discovered.³

The adherence to a Mediterranean diet has been reviewed in individuals and there has been a decrease in Alzheimer's disease risk. The Mediterranean diet consists of fruits, vegetables, nuts, whole grains, fish, olive oil, red wine in moderation and limited use of red meat. One case control study has examined the relationship of the Mediterranean diet in ET and did suggest lower risk of ET in those with greatest adherence to the diet. The mechanism as to why this diet may decrease the risk of ET still requires further investigation.⁴

It is generally recommended that individuals with ET limit all caffeinated products since tremor does worsen with increased caffeine intake. Alcohol in moderation has been shown to decrease tremor in individuals with ET but intake needs to be monitored carefully to avoid dependence and further nutritional deficiencies due to high alcohol consumption.

At the present time, it is recommended that all individuals maintain a healthy diet to include proportions of meats, poultry, fish, vegetables, breads, cereals, grains, dairy products and limited fats and , as recommended by the Food Guide Pyramid.⁵

Unless there is an issue of decrease in BMI and poor dietary oral intake, additional vitamins or dietary supplements are not generally warranted.

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Helping Students with Essential Tremor

By Beth A. Prokesch

Donna tried to write words in straight lines and rounded curves. She knew that handwriting was important to her teacher and parents, but she couldn't help it. Her hand shook as she held a pencil and guided it across the page. Donna wished she could control herself.

Evan's voice quavered whenever he had to give an oral presentation in class. His teacher told him not to be nervous, but deep down he knew it wasn't nerves. Other kids in the room laughed which made him even more self-conscious and stressed. Lately, his voice has been shaking more often. He was less inclined to speak up in class, in the hallway with his friends, and even spoke less at the dinner table.

In the case of Donna and Evan, these children may be experiencing signs of essential tremor (ET). Teachers, parents, family members, caregivers, and medical professionals familiar with ET can help a child identify problems, get diagnosis, find solutions, provide support, and prepare a child with this neurological disorder to better face the challenges of growing up.

What is Essential Tremor?

ET is a neurological disorder with the primary symptom of shaking, or tremor, of the hands, head, voice, legs, and/or trunk, and can be so severe as to be disabling. Approximately 10 million people in the United States have ET, including children. Tremor occurs during action such as eating, drinking, speaking or writing. It also occurs when the body or body parts are held in positions against gravity such as when holding the arms out in front of the body.

Although often a genetic condition, ET also occurs with no known family history of tremor. It affects people of any age, gender, or race. Age of onset, severity of tremor, body parts affected, and response to treatment varies, even within the same family.

ET is frustrating and embarrassing for children and adolescents, and it can lead to anxiety, depression, and social isolation. As a teacher, the most important things you can do for a student with ET is to be patient and to give positive reinforcement. With your support, a student with ET can succeed academically and socially.

Signs of ET:

- A visible, mild to severe shaking of the hands, head, arms, legs or trunk; or a complaint of "trembling inside." Some types of tremor are not visible.
- A quavering or shaking of the voice.
- Poor, shaky handwriting that does not improve.
- Poor fine motor skills and/or poor manual dexterity.
- A worsening of tremor when tired, stressed or excited.
- A nervous, anxious or restless disposition.

Why it is necessary to accommodate the student with ET:

ET does not affect a student's ability to learn, however, the frustration and embarrassment associated with the symptoms can hinder a student's education if teachers are not aware of ET and accommodate for it.

Accommodation provides the following benefits for the student with ET:

- Reduces frustration and promotes task completion.
- Reinforces independence and accountability.
- Supports and strengthens self-image.

Basic classroom accommodations:

- Help students with ET fit in with other students. Provide them with support, but not to the point of it appearing that they are different.
- Avoid lengthy written assignments or extend due dates.
- Avoid grading handwriting for neatness.
- Provide extended time for assignments and testing.
- Provide short breaks.
- Encourage the use of thicker or weighted pens and pencils.
- Allow the wrist of the writing hand to be held by the other hand when writing on paper or when writing on the blackboard.
- Encourage oral responses, even in math.
- Teach and review speaking skills.
- Allow the use of a computer for written homework, and allow the use of speech recognition software.
- Provide student/teacher notes.
- Allow the use of heavy metal rulers and protractors and the use of pointed-end compasses.
- Allow the use of weighted eating utensils and cups, and the use of straws.
- Allow the use of technology such as overhead projectors, video or computer presentations, PC/PDA and/or audio recorders.
- Allow the use of a podium during presentations so that students can grasp it and steady their hands while still having materials readily available.
- When reading, allow the use of a bookstand to steady a book.

This information was developed by Beth A. Prokesch, M.Ed., teacher of middle school English. She prepared this information after her two-year-old son was diagnosed with essential tremor.

The Hardest Challenge: Getting a Family Member to Accept Help

By National Caregivers Library, reprinted with permission

Sometimes convincing a loved one to accept help is the toughest part of being a caregiver. Understanding the factors that contribute to resistance is the first step in meeting this challenge.

Factor: Accepting help is viewed as an admission that the individual is no longer self-sufficient or “in control.”

Possible Solution: If possible, it is best for the care receiver to be involved in the decision-making process. Have your loved one become as active as possible from the beginning such as making telephone calls or being present at interviews. This will allow the person to feel more in control and make the outcome more palatable. People feel in charge of their lives when they have all of the information. When “selling” a solution, be able to provide your loved one with the facts.

For example: The limits of service or the time frame involved. How much it will cost and who will pay. How many payments (including insurance co-payments) will be handled. When the service will be available and what is required to be eligible.

When the care receiver is advanced in age or confused, it is particularly important to make changes slowly. It may be necessary to discuss the need for a service many times before the individual understands and accepts the changes. Make changes in small steps, or introduce new activities for a very short period of time at first, so the care receiver is not confused or overwhelmed by the situation.

Factor: To some people, using community services feels like the equivalent of accepting charity or “going on welfare.”

Possible Solution: Community-based services (particularly those offered at reduced fees or free of charge) are frequently funded through taxes. If this is the case, you can help the family member understand that he or she has “pre-paid” for these services, and it is not the same as accepting charity.

Factor: An obstacle to using services like home health aides can be an unwillingness to trust outsiders. Many people do not want strangers in their homes. They feel vulnerable. While only a very few have been involved in incidents of abuse involving service people or repairmen, many have heard stories of such things.

Possible Solution: Everyone has a right to their feelings, and as a caregiver, it is important to acknowledge the fears your loved one may have. Instead of telling Mom that she doesn’t need to be afraid, recognize that a little fear is healthy. Explain what you have done to ensure her safety and protect her valuables, such as: performing criminal background checks on any in-home workers; only dealing with agencies that are reputable, licensed and bonded; and getting referrals from qualified professionals. Knowing that you take Mom’s concerns seriously will help her feel more comfortable.

Where Should Care Take Place?

By Michael Smith, Senior Writer, National Caregivers Library

One of the toughest problems family caregivers face is the housing issue. Almost everyone wants to remain in his or her own home, and while this can be the least expensive housing option, it also requires the most planning. On the other hand, when age, infirmity, or other factors make independent living difficult or unsafe, it may be time to look at alternatives. To determine the best housing situation for your loved one's needs, consider the following steps:

Step One: Do the assessment

- As you assess your loved one's situation, find the balance that protects this individual's well-being while preserving as much of his or her independence as possible.
- Think about the amount of assistance your loved one needs with basic activities such as dressing, bathing, preparing meals, eating, walking, etc. Consider every task a person living on his or her own should be able to accomplish. The accumulated activities of daily living (ADLs) can be the key factors for determining a person's level of required care and what kind of housing is best.
- It might help to draw up a quick checklist or chart. List all ADLs on one side of the page. Then divide the page into three columns, marked "Needs no help," "Needs some help," or "Needs much help." Complete this evaluation by marking appropriate areas with an "X" or checkmark.
- Have another family member do the same evaluation separately, and compare results to get an objective view.
- Consult your family doctor or health professional for help predicting future needs.
- A private care manager can assess your loved one's needs and provide appropriate recommendations.

Step Two: Have the conversation

- Getting a loved one to accept help may be harder than finding that help in the first place. Most people, however, are more willing to accept decisions that they're involved in making. So include your loved one in the process as much and as early as possible.
- Many people will not acknowledge even a small weakness because they dread loss of independence.
- As you explore alternatives, create opportunities for your loved one to express his or her concerns—and for you to provide additional information that may help overcome resistance to change.
- If you hold a family meeting to discuss this issue, respect your loved one's dignity and privacy.

Step Three: Match housing to needs

- Review the chart you completed earlier as you consider available choices, comparing the types of services offered by each. This can be a difficult and confusing time, so it's important to be organized and methodical in your approach.
- Don't wait for a crisis to gather information. The best decisions are made without time pressures.

- Consider your loved one's desire to continue a favorite activity or hobby and how a change in housing might affect that ability.
- Take stock of what is most important in the individual's life and make sure this can be maintained, if at all possible.
- Remember that no choice is going to be perfect or give everyone 100 percent satisfaction. Your loved one's safety and happiness come first.

Step Four: If the person can remain independent, look into support structures

- Repairs and modifications can make your loved one's home safer and easier to get around in. Consider how even these simple changes can make a large difference in at-home safety:
 - Set the thermostat of the water heater at 120 degrees F or lower to prevent accidental scalding.
 - As a reminder to keep the smoke alarm working, change the batteries on daylight saving time.
 - Locate appliances close to outlets, making sure cords run along walls and away from sinks or stoves.
 - Install handrails and non-slip mats in the bathroom and shower.
 - A ramp may allow easier access to the home.
- In addition, there may be services available from the government, non-profit, and for-profit organizations that can help your loved one maintain independence:
 - "Meals on Wheels" deliver hot meals to the homebound.
 - Local community or senior centers offer companionship, including classes, recreational opportunities, travel, volunteer opportunities, and meals,
 - Community groups or even the local police may sponsor friendly visitor or companion programs where volunteers make scheduled visits to isolated seniors.
 - Volunteers at telephone reassurance programs call people to chat or check on their wellbeing.
 - So-called "gatekeeper programs" may be offered through public utilities and the Postal Service. Workers who regularly visit the home are trained to look for changes in your loved one's condition.
 - Your Area Agency on Aging can provide the names of local organizations that offer legal, financial, healthcare and other services.

Step Five: If semi-independent living seems appropriate, examine choices

Sometimes it is best to relocate to a residence that is more manageable, or where the necessary care services and support are "part of the package." The distinctions between each kind of facility are not always clear, and different states may use the same term to describe very different types of facilities. Generally, however, the categories for semi-independent living and housing include:

Adult Residential Care Facilities. Licensed boarding homes provide room, board, and help with medications and personal care. Residents have limited supervision:

Most adult care homes are single-family residential units.

Typical residents include: frail elderly persons with chronic physical or mental disabilities, chronically mentally ill adults, mentally retarded adults, or other adults with diminished physical and mental conditions.

Adult Family Homes. These facilities accommodate couples and are licensed to care for up to six residents. They provide room, board, laundry, and necessary supervision, assistance with activities of daily living, personal care, and social services. In most cases, nursing services are not provided, and residents must manage their own care.

Assisted Living Facilities. This has become a catchall term for any boarding home that emphasizes privacy, independence, and personal choice. Services include meals, personal care, medication assistance, limited supervision, organized activities, and limited nursing services.

Assisted living is licensed by state governments and is known by as many as 26 different names including residential care, board and care, congregate care, and personal care. Services, style, and costs vary widely from facility to facility. Some may have a resort-style ambience while others are geared toward affordable living with few amenities.

This brand of residential care is not a substitute for a nursing facility.

Continuing Care Retirement Communities. CCRCs are designed to provide seamless transitions as care needs increase. A CCRC accepts seniors while they are still active and independent, and then provides an expanding range of caregiving services—including professional nursing care—as needed.

These facilities are privately owned. All levels of care take place on one “campus.”

These communities typically require a sizeable entry fee, plus monthly maintenance fees, in exchange for a living unit, meals, and eventual healthcare coverage, up to the nursing home level.

Many of these communities make their services available on a month-to-month rental basis. Remember that the way a facility describes itself is not necessarily a real indication of the level of care. FamilyCare America offers additional information, action plans, and checklists designed for caregivers exploring housing options.

Step Six: If the person is dependent, begin to evaluate facilities

Your loved one may be in a situation where he or she can no longer care for him or herself. Housing alternatives include:

- Intermediate care facilities for people who are physically and psychologically stable but need a nurse’s care for medications and other needs.
- Skilled nursing homes for people whose unstable or complex conditions require 24-hour supervision, help with medications, and frequent assistance with activities of daily living.
- Nursing homes provide rooms, meals, recreational activities, and protective supervision to residents. Services vary, yet all nursing homes are licensed by states and follow federal regulations. Keep in mind that some facilities specialize in Alzheimer’s disease or have sub-units for certain illnesses.

For more information:

National Institute on Aging, P.O. Box 8057, Gaithersburg, MD 20898, (800) 222-2225, www.nia.nih.gov.

Alzheimer's Association, 225 North Michigan Avenue, Floor 17, Chicago, IL 60601-7633, (800) 272-3900, www.alz.org.

National Caregivers Library, 901 Moorefield Park Drive, Suite 100, Richmond, VA 23236, (804) 327-1112, www.CareGiversLibrary.org.

Meals on Wheels Association of America, 203 South Union Street, Alexandria, VA 22314, (703) 548-5558, www.mowaa.org.

Other Types of Tremor & Similar Disorders

Tremor and Other Movement Disorders

By Peter A. LeWitt, M.D.

Readers of IETF materials no doubt may wonder if the definition of tremor has a medical designation that differs from its ordinary usage. There are several words that can convey a similar meaning, such as trembling, shaking, jittering, vibrating and so on. Despite the many varieties of subjective experiences that patients may report, this disorder is best described by the single term: tremor. Implicit in this designation is the concept of a rhythmic, to-and-fro movement with a high degree or regularity in its frequency and amplitude.

It is a characteristic sign (observed phenomenon) and symptom (subjective experience) of numerous medical conditions. For several of these, the patterns of tremor can be virtually identical despite different causative mechanisms. However, the presence of tremor is not always indicative of an illness or neurologic disorder, nor is what seems to be a tremor always deserving of this terminology. This article provides a short overview of several movement disorders in order to indicate how tremor is differentiated from each of them.

Not all involuntary movements are tremor. Other types of involuntary movements, rhythmic or otherwise, can also involve the hands, face and other regions of the body that are also commonly affected by tremor. Among these conditions is chorea (or dyskinesia), characterized by continuing “fidgety-like” movements which sometimes have a semi-purposeful appearance (e.g., incorporated into adjustments of the hair or clothes) or suggest a state of restlessness.

In contrast to tremor, these movements tend to have a “dancing” quality with a random fashion, although they cannot generally be defined as regular to-and-fro movements around any particular joint as can tremor. Furthermore, their tempo is generally slower. The causes of choreic movements include a number of neurologic disorders (e.g., Huntington’s disease, benign hereditary chorea and acanthocytosis-related disorders) as well as acquired or secondary conditions such as tardive dyskinesia, estrogen-induced dyskinesia, or damage to certain deep structures of the brain, among other causes. The distinction of these and other conditions responsible for involuntary movement disorders generally requires an evaluation by a specialist in neurologic movement disorders.

Other disorders with involuntary movements include tics, which are random, jerking movements often of a sudden, jolting nature. Tics can be characterized as facial grimaces, forceful blinks, head thrusts or sudden movements of the limbs; sometimes production of sounds or even words can occur involuntarily. Tic disorders can occur in any number of patterns and when multiple body parts are involved can signify a condition known as Gilles de la Tourette syndrome.

Sudden jerking movements occurring during sleep, termed nocturnal myoclonus, are part of another tic-like disorder that occurs in just about everyone, but which can be particularly troublesome when recurrent or severe. Other familiar conditions displaying regular or stereotyped involuntary movements which are not tremor include hiccups, shivering and behavioral mannerisms (e.g., so-called “nervous” habits).

Dystonia refers to sustained abnormal postures usually with a twisting component. Sometimes, as in torticollis, there can be a jerking quality to neck movements suggestive of tremors. Among the range of dystonic conditions are problems such as involuntary spasms of eye closure (blepharospasm),

exaggerated facial or mouth movements (orofacial dystonia or Meige syndrome) or strained or quavering qualities of the voice (spasmodic dysphonia). Sometimes, jerking or writhing movements take place only upon voluntary activity of a body part, a phenomenon termed action dystonia.

The examples described above with their medical terminology are indicative of the variety of movement disorders that could be confused with tremor. In fact, the same patient displaying one or more types of these conditions might also display typical features of tremor. In Parkinson's disease, choreic movements can be produced by overmedication in a patient who later, as the medication wears off, might display the typical tremor at rest. Many patients affected with torticollis are also affected by action tremor of the hands or voice.

It is also possible that tremor found in the context of another neurologic condition is not at all related; ET as a familial trait may coexist with other tremor disorders (such as PD) or any of the other movement disorders mentioned previously.

The differentiation and proper diagnosis of movement disorders call for careful observation and review, sometimes including the testing of the blood, urine or cerebrospinal fluid. CAT (computerized axial tomography), x-ray studies or MRI (magnetic resonance imaging) pictures of the head can be useful in this process. Medication approaches for controlling movement disorders other than tremor are different from the ways in which essential or parkinsonian tremors can be treated.

Essential Tremor of the Voice vs. Spasmodic Dysphonia

By Madeleine Petthan, M.S., C.C.C.-S.L.P., and Michael M. Johns, M.D.

Introduction

Certain neurologic conditions can cause people to have problems with their voice. These voice problems can often lead to more difficulty communicating throughout daily life. It is important that patients with neurological voice disorders are evaluated by an otolaryngologist, or ENT doctor, in addition to their neurologist to determine the diagnosis and discuss treatment options. Many patients with essential tremor also experience essential tremor of the voice. Essential tremor of the voice can often be confused with another neurologic voice disorder known as spasmodic dysphonia.

What is Essential Tremor?

Essential tremor is a disorder of the central nervous system that may result in tremulousness of the head, limbs, tongue, palate, and/or larynx. The tremor may occur while a structure is at rest or in action. For example, someone's hand may be tremulous while resting in their lap or it may not become tremulous until they reach for a cup of coffee on the table. Action tremor can occur while someone is purposefully moving the affected structure, or while postural muscles are contracting to hold the structure in the appropriate position. Essential tremor is characterized by rhythmic, involuntary movements of muscles during purposeful movements. Typically, essential tremor is absent at rest and maximal during the maintenance or termination of a movement. This is similar to essential tremor of the voice in that the voice box or larynx will only become tremulous when activated for voicing during speech.

What is Essential Tremor of the Voice?

Essential tremor may present as a generalized neurological disorder, affecting many structures of the body, or as an isolated symptom of the voice. Patients with essential tremor of the voice may not necessarily have generalized tremor in the limbs, trunk, or other major postural muscles. In other words, the voice tremor may be the only primary characteristic. The larynx, or voice box, is not the only structure which can cause essential tremor of the voice. Tremor of the voice can be caused when any of the structures in the speech system is affected. Essential tremor of the voice may be caused by tremor in the soft palate, tongue, pharynx, or even muscles of respiration. Extralaryngeal tremor (i.e., outside the voice box) has been reported in up to as many as 93% of patients with diagnosed essential tremor of the voice. Similarly, most patients with essential tremor of the voice also have tremor affecting their hands, leg, chin, or trunk.

Essential tremor seems to be associated with aging, although the reasons are still inconclusive. Most studies report average age of onset from the late 40s to early 50s. The highest prevalence of essential tremor of the voice is in the seventh decade of life. Heredity may affect the likelihood of developing tremor. The frequency (speed) of tremor is typically between three and seven times per second. Medications that often reduce generalized or extremity tremor, typically are not as effective for the voice symptoms.

The most prominent voice symptom and diagnostic indicator for essential tremor of the voice is a periodic or rhythmic modulation of either frequency (i.e., pitch) or intensity (i.e., loudness) in the voice. This modulation is most noticeable during prolongation of a vowel. For this reason, tremor will be most apparent when someone is speaking vowel laden sentences (i.e., you were away all year). Tremor may become so severe that a stoppage of voice occurs. This is one of the reasons that

essential tremor of the voice is often confused with spasmodic dysphonia. Both diseases, in certain speech contexts, will cause voice breaks, or stoppages during speech. Spasmodic dysphonia and essential tremor of the voice are also known to occur at the same time.

What is Spasmodic Dysphonia?

Spasmodic dysphonia (SD) is classified within a family of neurological conditions called focal dystonia. A focal dystonia is a condition in which movement is abnormal in an isolated part of the body. SD causes abnormal movement of the vocal folds similar to the way a condition called blepharospasm causes abnormal contraction of the eyelid. SD is different from blepharospasm, however, in that its effects are only noticed during meaningful tasks (i.e., speech). Interestingly, vegetative functions of the larynx, such as coughing, laughing, whispering, and even singing, may be normal.

Two types of SD are well described in the medical literature: abductor and adductor type. The two are clinically distinguishable by the way the voice sounds.

In the case of adductor type SD, adductor muscles—or “closing” muscles of the vocal folds—can spasm during connected speech. The voice sounds strained or strangled with intermittent stoppages of the voice. The severity can range from mild, or barely noticeable, to so severe that any attempt at producing a word brings great effort to the patient. Abductor type SD is caused by spasms of the muscles of abduction, or “opening,” which cause sudden and intermittent escapes of air during speech. These spasms occur during the transition from voiceless consonants (i.e., t, k, s) to vowels (i.e., i, u, a).

People with either type can often describe certain words which are harder for them to produce. For example, a patient with abductor type SD may report it is very hard for him or her to shout “nice shot!” at a sporting event due to the transition from the “sh” sound to the voiced vowel “o” in the word “shot.” The onset of spasmodic dysphonia is generally believed to be during middle age, somewhere between the late 40s and early 50s. Researchers have yet to establish an exact gender ratio; however, most clinicians agree that the majority of cases they see are women. The onset of spasmodic dysphonia is typically more gradual, but there are some reports of rapidly worsening symptoms.

What treatments are available?

Medical therapy is generally not effective for patients with SD, while medical therapy for ET of the voice is significantly less effective than for treatments for extremity tremor. Voice therapy is also not typically effective at improving voice problems caused by SD and has a limited role in essential tremor. Currently, the main treatment for both ET of the voice and SD involves the injection of very small amounts of botulinum toxin, otherwise known as Botox™ into the vocal folds or other muscles in the larynx. Botox injections to treat voice disorders are typically administered by otolaryngologists who specialize in voice disorders. Some neurologists also treat neurologic voice disorders with Botox. These injections can be done awake in the office directly through the skin of the neck, just under the thyroid notch or “Adam’s apple.” The physician will often use some form of numbing medicine to numb the skin around the injection site prior to administering the Botox.

With these injections, muscle contraction is temporarily blocked by the nerve endings, and the vocal folds are temporarily weakened. When the muscles are weakened from Botox, tremor and spasms are reduced. This leads to a more stable and effortless voice for many patients, allowing for more

effective communication. In the case of ET of the voice and SD, what we find is that the spasms and/or tremor do not return for a period of 3-5 months.

Summary

SD and essential tremor of the voice can significantly impair people's ability to speak and communicate effectively. Although the disorders are distinct, some overlap of symptoms can occur. Severity of voice changes can range from barely noticeable to patients being unable to effectively communicate. Treatment is available for both conditions. If you are experiencing voice changes relating to your tremor or suspect you may have SD in addition to your tremor, ask your neurologist about treatment and potentially a referral to an otolaryngologist.

Voice Tremor: Proper Diagnosis and Treatment

By Joseph Jankovic, M.D.

Traits

Awareness of voice tremor can cause social embarrassment and this itself can be a source of stress, thus exacerbating the condition. Some patients, in addition to shaking voice, may also experience some of the following physical traits:

- hoarseness
- low volume voice and difficulty projecting their voice
- increased effort during public speaking or when talking over the telephone
- decreased speech intelligibility
- throat and neck discomfort
- shortness of breath

While in most cases voice tremor may be hardly noticeable, except during periods of stress, in other cases it may be not only embarrassing, but quite troublesome and even disabling. Rarely, voice tremor can markedly interfere with or prematurely shorten a professional career of an announcer, auctioneer, singer, politician (e.g. senator Robert Byrd), or an actor (e.g. Katherine Hepburn).

Prevalence

While voice tremor may occur in isolation, it is more likely accompanied by or is a component of essential tremor (ET). The prevalence of voice tremor among patients with ET ranges from 25-62%, based on studies. This marked variability in reported prevalence of voice tremor is probably due to differences in populations studied, methodologies used to detect voice tremor, and the experience of the clinician.

In one study of 34 patients with voice tremor associated with ET, 93% were female, and the average age at onset was 63 years (Sulica and Louis, 2010). Although 68% also had tremor in their upper limbs, only 32% were aware of an arm tremor. A genetic cause for this tremor was suggested by the finding that up to one-half of the patients had at least one first-degree relative with tremor.

Misdiagnosis

Nearly one-third of patients with ET are misdiagnosed as having “spasmodic dysphonia.” This condition is produced by involuntary contractions (spasms) of the vocal cord muscles producing either a strained voice or a low-volume, whispering voice.

Spasmodic dysphonia is categorized as a form of focal, laryngeal dystonia. The word dystonia means abnormal involuntary contraction of muscles producing abnormal movement or posture such as torticollis (involuntary turning of the head and neck) or blepharospasm (involuntary contraction of the eyelids). These and other dystonia may be associated with an irregular tremor, the so-called dystonic tremor, which may be sometimes difficult to differentiate from ET, hence the frequent misdiagnosis.

Careful evaluation of patients with essential voice tremor using laryngoscopy usually shows rhythmic contractions of the vocal folds during sustained vowel phonation (e.g. aaaahhh, eeeehhh). These contractions frequently extend beyond the larynx into the pharynx and other areas adjacent to the voice box. Besides ET and spasmodic dysphonia, there are very few conditions that can cause voice

tremor. Parkinson's disease rarely ever causes voice and head tremor unless it is also accompanied by ET.

Treatment

Voice tremor is not easy to treat. It often does not respond as well as ET to the conventional anti-ET medications such as propranolol and primidone. If these two medications fail, topiramate, zonisamide, and benzodiazepines can be tried. While alcohol reduces tremor amplitude in vast majority of patients with ET, only about a quarter of patients with voice tremor report improvement with alcohol (Mostile and Jankovic, 2010).

Botulinum toxin injections into the vocal cords can be quite helpful, although the benefits usually last only three months. In contrast to focal dystonia, including laryngeal dystonia that produces spasmodic dysphonia, botulinum toxin injections for voice tremor are less predictable and may be associated with adverse effects such as low-volume voice, breathiness, and hoarseness.

In one study involving 27 patients with adductor spasmodic dysphonia and vocal tremor and in four patients with severe vocal tremor alone, a significant improvement in various acoustic measures was observed after botulinum toxin injections into laryngeal muscles (Kendall and Leonard, 2010).

Voice, tongue, and face tremors may also improve with deep brain stimulation, but this surgical procedure should be reserved only for those patients who are severely disabled by their tremor despite optimal medical and botulinum toxin therapy.

Voice Tremor

By Christy L. Ludlow, Ph.D.

Voice is the result of air from the lungs passing between the two vocal cords (or vocal folds) setting them into vibration. The vibration of the vocal folds is the sound source for speech when we talk. Voice tremor refers to a rhythmic shaking of the voice that can be heard during speech. These regular changes may be voice breaks, breathiness, regular pitch breaks or regular changes in voice loudness. These different rhythmic changes are due to certain muscles in the larynx (voice box) or the head and throat which affect the voice during speech. In some persons, these tremors affecting the larynx, throat or head are present all the time that the person is awake but are detected only when the person goes to speak.

Voice tremor is upsetting and interferes significantly with a person's ability to communicate because it makes them difficult to understand particularly over the telephone and also makes them sound like they are crying or are upset. Often persons with voice tremor find that others think they are elderly when they first hear them over the telephone.

Voice tremor can start at any time, but usually affects persons later in life, beginning around 60-70 years of age. It can begin slowly, progressing over a year and then remaining chronic, or it can first occur when a person is very upset by a life event and then remains a chronic disorder. Sometimes, it appears first in the larynx and then after a few years the person begins to see tremor emerge in other areas such as affecting head movement or the hands.

In our clinic, we see voice tremor more often in women than in men and sometimes see it in families, where the grandmother, mother and daughter all have voice tremor. Unfortunately, many of the persons we see have not sought treatment for their voice disorder because they are embarrassed by their shaky voice. They avoid using the telephone and meetings with strangers as much as possible. Thus, their voice disorder infringes upon their social and work lives, making them withdrawn from society.

Voice tremor can be a significant disability particularly when it causes regular voice breaks making the person difficult to understand and limiting their ability to communicate through speech with others. Voice tremor can be a significant occupational disability when it limits the types of work a person can perform. Teachers, lawyers, receptionists, salespersons, and businesspersons are just a few of the occupations that are unavailable when a person has been affected with voice tremor. In addition, to the communication problems, persons with voice tremor usually complain about the great amount of effort they have talking. Often, the more a person tries to control the shaking of their voice, the more difficult it becomes for them to speak.

Types of Voice Tremor

Voice tremor is a symptom. That is, it can be caused by many types of neurological disorders or can be a disorder occurring on its own for an unknown reason. In voice tremor, only a few or many of the laryngeal muscles can be affected. Depending upon which muscles are affected by regular spasms, the particular characteristics of a persons' voice tremor may differ. Voice tremor is most easily heard and detected when a person is asked to prolong a single vowel for up to 10 seconds. At this time, the regular shaking of the voice can be heard, usually at a rate of five times per second.

Adductor Voice Tremor

In some persons, the tremor causes the voice to break off completely. Muscles in the vocal folds spasm and they close the larynx. This makes it impossible for the person to push air through the larynx and make the vocal folds vibrate for sound. When these voice breaks are regular they are referred to as a voice tremor. Sometimes they are irregular and then are referred to as adductor spasmodic dysphonia. Adductor tremor and adductor spasmodic dysphonia often occur together in the same person. Usually when persons have adductor voice tremor, the adductor muscles of the larynx are affected by uncontrollable muscle spasms—the thyroarytenoid, lateral cricoarytenoid and/or interarytenoid muscles of the larynx. These same muscles may be affected in persons with adductor spasmodic dysphonia. When adductor muscle spasms are less severe, they produce regular alterations in the loudness, or wavering of the voice.

Abductor Voice Tremor

Regular breathy breaks occur in the voices of persons with abductor voice tremor. This is because the muscles that open the larynx, primarily the posterior cricoarytenoid muscle, have spasms and open the vocal folds, interfering with vocal fold vibration, and making the voice intermittently become a whisper. Some persons also have pitch breaks in their voices as well as breathiness. This is often due to spasms in the cricothyroid muscle that lengthens the vocal folds and produces rapid changes in voice pitch.

Treatment

The most effective treatment for voice tremor is an injection of a small amount of botulinum toxin into the muscles that are spasming. The result usually is a smoother voice and persons usually find it much easier to talk. This treatment is most helpful in persons with adductor voice tremor if only one or two muscles in the vocal folds are affected by spasms. If many muscles of the throat are affected besides the laryngeal muscles, botulinum toxin injections will be less effective in controlling the tremor. Side effects that occur in some persons after injection include some breathiness of the voice for about two weeks and some difficulty with swallowing liquids rapidly for about five days. Usually if the person sips liquids through a straw slowly they can avoid coughing because of swallowing problems.

Once the period of side effects is over, persons usually find that their voice remains clear and easy to produce for about two to three months, and then they need another injection. Injections of botulinum toxin can be given in one or both sides of the larynx. In general, the bilateral injections do not last as long as the one sided injections, although both techniques are effective.

When voice tremor is associated with neurological disorders such as Parkinson's disease, multiple sclerosis or cerebellar disease, usually voice tremors occur in association with head and/or throat tremor and cannot be managed well by botulinum toxin injections. Most medications are not helpful in voice tremor; although in some persons, medications such as propranolol may reduce the effort and degree of voice tremor to some degree.

Persons with abductor voice tremor usually find it more difficult to gain benefit with botulinum toxin injections. This is usually because the muscle with tremor in these persons is often the posterior cricoarytenoid, which is a more difficult muscle to inject with botulinum toxin. In these cases, the side effects can include stridor (an abnormal, high-pitched sound heard during breathing) if too much toxin is injected.

Future Research

Persons with voice tremor often report that other members of their family have similar or related disorders. Scientists are working with geneticists in hope of identifying the genes that may lead some people to be more likely to develop voice tremor. Scientists are also working to develop other treatments, which might be effective in persons with abductor voice tremor, such as muscle stimulators. It is too early at this time to know whether these new approaches have potential or not.

Straight Talk on Therapy for Vocal Tremor

By Rosemary A. Lester, M.A., CCC-SLP

How often do you think about how your voice sounds or how it feels when you speak? Normally, few of us think about our voices on a daily basis because speaking is usually effortless. In fact, many of us are unaware of our voices unless we have a cold and sound hoarse or we are in a noisy room and have to shout to be heard. But some people with essential tremor think about their voices all the time. They notice that their voices sound shaky when they speak. They feel that their voices are difficult to control and that speaking is tiring. These are the people who have vocal tremor, a voice disorder that affects approximately 18-30% of people with essential tremor. Fortunately, some people with vocal tremor can benefit from therapy with a speech-language pathologist (SLP) to learn how to make the voice sound less shaky and how to use less effort when speaking.

What Causes Vocal Tremor?

Many people assume that a shaky voice is caused by tremor within the larynx (the “voice box”). This is true for some people with vocal tremor. But vocal tremor can also be caused by tremor affecting the chest, abdomen, mouth, or throat. We use all of these parts of the body to speak. Before we speak, we take a breath and then we slowly release the breath to speak. We control the breath using our chest and abdominal muscles. As we release the breath, we also use muscles in the larynx to allow the vocal folds (often called the vocal cords) to vibrate and produce sound. In addition, we use muscles in the mouth and throat to shape the voice into sounds that can be understood as speech. Because muscles in all of these parts of the body are used to speak, tremor affecting any of these areas can make the voice sound shaky.

How is Vocal Tremor Treated?

The medications that are used to treat tremor affecting the arms and legs are not often effective in treating vocal tremor. But some people with vocal tremor benefit from injections of small amounts of botulinum toxin (Botox®) into the larynx. This can sometimes make the voice sound less shaky and can make it feel less effortful to speak. Unfortunately, this treatment does not help everyone with vocal tremor, and it may have some negative side effects including difficulty swallowing. For people who do benefit from Botox®, the effects are only temporary so the injections are typically repeated every few months. For these reasons, some people seek a different treatment approach for vocal tremor that involves therapy with an SLP.

What is the Research on Therapy for Vocal Tremor?

Recent research has demonstrated that therapy can help make the voice sound more stable and make it feel easier to speak for some people with vocal tremor. But more research is needed in this area to determine who the best candidates for therapy are and what the best treatment approaches are for these individuals. Because so many muscles are used to produce voice and because essential tremor can affect many different muscles, it can be challenging to study vocal tremor in people who have essential tremor. That is one of the reasons why researchers at the University of Arizona are using computer models to simulate vocal tremor involving the different parts of the body that are used to speak. Using these models, they can isolate tremor to one part of the speech mechanism or simulate combinations of tremor affecting multiple parts of the speech mechanism. They can then make adjustments to the voice and determine which ones reduce how shaky the voice sounds to listeners. The ultimate goal is to use these findings to help determine the most effective and efficient therapeutic approaches for treating vocal tremor.

How Can I Try Therapy for Vocal Tremor?

If you think that therapy might help you, ask your doctor about a referral to an SLP who specializes in voice. You may also visit www.asha.org/findpro or call (toll free) 800.638.8255 to find an SLP in your area. To determine if you would benefit from therapy, the SLP will ask you about the changes in your voice, listen to and record your voice, and watch the way you speak. The SLP will see if tremor affects your larynx, chest, abdomen, mouth, or throat while you speak. Depending on the results of the evaluation, the SLP might teach you ways to adjust the pitch or loudness of your voice, change your breathing patterns, or alter the way you produce speech sounds. The SLP will then help you learn to use these techniques in your everyday speaking activities to improve the way your voice sounds and the way it feels when you communicate with your family, friends, and colleagues.

Head Tremor Explained

By Joseph Jankovic, M.D.

Why do people get head tremor?

There are many reasons why some people shake their heads besides indicating their agreement or disagreement. Most patients with head tremor have either essential tremor (ET), neck (cervical) dystonia, or both. About one-third of all patients with ET in the hands also have tremor of their head due to the backward and forward movement of the cervical muscles. When the head tremor is regular in rhythm, either in a side-to-side (no-no, or negation) or anterior-posterior (yes-yes, or affirmation) direction, then the likely cause is ET head tremor, particularly if it occurs in women and is associated with hand and voice tremor.

Dystonia

Dystonia is a neurological disorder that involves involuntary, patterned muscle contractions. These cause twisting and other abnormal movements or postures that are often initiated or worsened by voluntary action, and that are relieved by sleep or various “alleviating maneuvers” (also referred to as sensory tricks or “geste antagoniste”).

When it affects the neck, it is referred to as cervical dystonia. Cervical dystonia causes an abnormal position and/or movement of the head due to a combination of twisting, flexing, and extending movements of the neck. While some patients with cervical dystonia have a more sustained (tonic) contraction of the neck muscles, others have more dynamic (phasic) dystonia associated with neck/head tremor. When a patient with cervical dystonia attempts to correct the abnormal posture and bring the head into primary (or neutral) position, it causes the oscillatory movement, or tremor, to occur or even get worse.

It is not known why only some patients with ET have head/neck tremor. ET-associated head tremor appears to be more common in women with ET, and in those patients who have a more aggressive growth of ET, often with a less favorable prognosis than those without such head tremor. Head tremor by itself is very rare in patients with ET. Some clinicians have suggested that head tremor in patients with ET is almost always due to them also having cervical dystonia.

Dystonic tremor

Some head tremors, however, are quite irregular in rhythm and may be associated with subjective “pulling,” “spasm,” or “pain” in the neck. This type of tremor would suggest the possibility of dystonic tremor. This typically subsides, or even completely disappears, when the patient stops resisting the pulling and allows the head to move into the position of maximum pull (called the “null point”).

Other causes of head tremor

Head tremor is almost never seen in patients with Parkinson’s disease unless they have an associated ET. High amplitude head oscillation, also called titubation, may be seen in patients with damage to the part of the brain involved with coordination of movement, called the cerebellum. Cerebellar titubation is typically seen in patients with multiple sclerosis or stroke involving the cerebellum, brainstem, or both. In addition to the ET-related head tremor, other generic disorders such as Wilson’s disease and fragile X–associated tremor/ataxia syndrome (FXTAS) should also be considered in patients with head tremor.

Another head oscillatory movement referred to as bobble-headed syndrome, can be seen in patients with cysts or tumors in or around the third ventricle. Rarely, head oscillation can be seen in patients with aortic valve insufficiency.

Treatment options

Medications conventionally used in the treatment of ET, such as propranolol, primidone, and topiramate, may be helpful in patients with ET-related head tremor. In one study, zonisamide was found to be more effective than propranolol in the treatment of head tremor.

If medications are not effective, or cause intolerable side effects, then injection of botulinum toxin (Botox®) into the affected neck muscles may be the most effective and safest treatment, particularly if there is associated cervical dystonia. When used by an experienced clinician, botulinum toxin treatment is generally well tolerated, although some patients may experience transient neck weakness and swallowing difficulties.

As a last resort, when medications and botulinum toxin fail to adequately control the head tremor, deep brain stimulation (DBS) targeting the thalamus or adjacent brain nuclei may be considered. This procedure has been used successfully in the treatment of ET, Parkinson's disease, dystonia, and other movement disorders.

Orthostatic Tremor: ET or Not?

Tremor Talk

Several times a year, the IETF receives a phone call from a frustrated person saying that they have visited several doctors, including neurologists, and can't get a diagnosis. They generally describe their symptoms as an unsteady feeling in their legs while standing, a fear of falling, and relief upon sitting or walking. They ask us if they have ET.

Not being doctors, we can't say, but often we ask if any of the doctors they had visited ever mentioned orthostatic tremor (OT). They almost always answer with a question, "What's that?" That's a good question and is one many physicians cannot answer.

According to The Movement Disorder Society's 1998 consensus statement, three criteria must be met for a diagnosis of OT: a subjective feeling of unsteadiness when standing; a visible or palpable rippling of the leg muscles while standing; and an EMG recording of a 13-18Hz (cycles per second) tremor in the legs while standing.

OT is rare and misunderstood. Part of this misunderstanding is because some researchers and clinicians believe that OT is a form of ET. In fact, some medical websites, such as the Cleveland Clinic site, describe OT as being a "variant of essential tremor."

The main reason why some believe OT is a form of ET is because many patients with OT have a weight bearing arm tremor. According to research compiled by Peter Bain, MD, Imperial College, London, England, arm tremor exists in 90 percent of all cases of primary OT.

In addition, three medications commonly prescribed for ET – primidone, clonazepam and gabapentin – are also prescribed for OT, but with less treatment success than with ET. Other drugs used to treat OT include levodopa or a dopaminergic agonist, which are primarily Parkinson's medications, and sodium valproate and phenobarbital, which are anti-seizure medications.

According to OT patient, Gloria Nelson MacWright of New Jersey, the neat diagnostic description of OT minimizes the experience of living with the condition.

Gloria describes her OT as "a sudden, unique, disabling event" in her legs while attempting to do anything that requires remaining standing.

"The muscles in my legs become hard, my ankles feel weak and my toes curl under. In addition, I'm overwhelmed with a feeling of panic and the need to find a place to sit or to walk to relieve my symptoms."

According to Gloria, when she went searching for a diagnosis 28 years ago, she encountered disbelief that anything was physically wrong, and at one point she was misdiagnosed with Parkinson's disease. Finally, in 2000, she was diagnosed as having OT.

Peggy Whitta, Kenora, Ontario, Canada also has OT. She first noticed unsteadiness in her legs while trying to peg clothes to a clothesline. Sometime later, she attended a funeral service and was unable to hold her hymnal with both hands because she needed to steady herself by hanging on to the

bench in front of her.

Peggy was diagnosed fairly quickly because she had an EMG that showed she had the high frequency tremor of OT.

“I know I was lucky. I have heard of so many horror stories about people having OT and going to numerous neurologists before they finally get one that knows about OT. Some were even sent to psychiatrists, and you can imagine how that must have felt.”

Because of the challenges and humiliations endured in public because of hand, head or voice tremor, many people with ET can empathize with the frustrations and public embarrassments experienced by individuals with OT. Both Gloria and Peggy speak of preplanning details of social encounters and of the anxiety of encountering the “unfamiliar and unexpected.”

“Even happy social events can produce stress. The unfamiliar always brings up the stress level because I’m constantly looking for places to lean or sit,” explains Gloria. “Sometimes, if I’m forced to stand over my time limit, I freeze in position.”

Peggy recalls the difficulties of crossing the street because of OT. “As soon as I stop at the crosswalk for traffic — because I am standing — the tremor starts. If it is more than a minute or so before the light allows me to cross, it is not easy to get going again.”

Issues with standing also disrupt walking in unexpected ways such as needing to sit or lean against something for a few seconds before changing direction. Even a slight hesitation creates problems.

“The instant my brain thinks I am stopping — say someone calls my name and I momentarily pause — I get unsteady,” explains Peggy. “As another person with OT once said, walking with OT is like walking on a floor covered with marbles.”

Then there is the fear and reality of falling due to OT. “I have fallen twice — once I had my husband’s arm at night while we were walking home and he kind of side-stepped, and I wasn’t able to keep up with him.” says Peggy. “Once I have his arm he has to keep a steady pace, or if he is going to deviate he has to let me know. The other time I was sitting at a table painting a door, and I had to keep moving my chair. I am standing up and sitting down like a yo-yo. I forgot to move the chair one time and sat down heavily on the floor.”

A lack of awareness of OT creates emotional conflict for Peggy at times.

“My mother was in a nursing home and used a walker. There was no way I could possibly walk with her when we were going to lunch or tea down the hall. I had to go ahead. I always felt so self-conscious because I was sure the staff or people were thinking ‘Why doesn’t Peggy walk with her mother?’”

Returning to the question of whether OT is a form of ET, we asked members of the IETF Medical Advisory Board for their opinions.

According to Rodger Elble, M.D., Ph.D., Professor and Chair of Neurology, Southern Illinois University School of Medicine, Springfield, IL, “although ET affects the legs of 30 to 45 percent of

ET patients, the unusually high frequency (14-18 Hz) of OT and the marked synchrony among ipsilateral and contralateral muscles of the legs are never seen in ET. OT is a distinct entity, not a variant of ET.”

On the other hand, Joseph Jankovic, M.D., Professor of Neurology, Director of the Parkinson’s Disease Center and Movement Disorders Clinic, Baylor College of Medicine, Houston, TX, says that “while there are differences in clinical phenomenology (e.g. legs/trunk and high frequency in OT) and response to drugs, at least 50 percent of our OT patients have associated ET.”

Gloria believes that OT is a distinct condition, and created a website to raise awareness about OT, and to give individuals with OT a place to share their stories and find understanding. For her efforts, Gloria was awarded the Governor’s Award for Volunteerism from the state of New Jersey last year.

To learn more about OT, visit www.orthostatictremor.org.

Orthostatic Tremor

By Daniel Tarsy, M.D.

Orthostatic tremor is a relatively rare and typically misunderstood condition in which individuals experience marked trembling of their legs which begins immediately after they stand up. Tremor gradually increases to the point that they are unable to remain standing for more than 10-20 seconds at a time before needing to sit down or lean against a wall for support. Despite the strong feeling of shakiness, which is felt by the patient (hence the term “shaky legs syndrome”), there is little or no obvious tremor to be seen. Remarkably, the tremor disappears as soon as the affected individual begins to walk and is also absent when he or she is seated or lying down. For these reasons, the diagnosis is usually missed by general physicians as well as neurologists. If the patient remains standing, tremors become more obvious, begin to involve the trunk and spinal muscles, and cause the patient to feel he is at risk for falling, although falls are relatively uncommon in this condition.

The diagnosis can be made if the physician simply places his hand on the patient’s legs while they are standing and feels the trembling of leg muscles. If muscle activity is recorded by surface electrodes connected to an electromyography (EMG) machine, an unusually high frequency pattern of muscle tremor can be identified in the range of 14-18 Hz (cycles/second). Interestingly, simply being vertical is not enough to bring on the tremor since it is absent while walking or if the affected individual is suspended by a harness with his feet off the floor.

Orthostatic tremor therefore appears to be a special type of postural tremor, which is activated by weight bearing. However, its relationship to more common but much lower frequency postural tremor, such as essential tremor, is uncertain.

Most patients with orthostatic tremor do not have tremor of the hands, head, or voice and the family history is negative. Unlike essential tremor, propranolol (Inderal®), primidone (Mysoline®), and alcohol are ineffective for orthostatic tremor. On the other hand, clonazepam (Klonopin®) is remarkably effective in relatively low doses and is considered the treatment of choice for this condition. Recently gabapentin (Neurontin®) has also been shown to be effective in a controlled study and, if necessary, may be given in combination with clonazepam to achieve maximal therapeutic response.

The cause of orthostatic tremor is unknown. It affects men and women equally, usually in middle to late life. Unsteady gait and fear of falling is a common problem in elderly individuals. Orthostatic tremor is therefore a rare but potentially treatable cause of unsteadiness, which is very important to identify and treat in this patient population.

Task Specific Tremor

By Peter Bain, M.D.

The IETF Tremor Investigation Group (TRIG) has defined task specific tremor as tremor that only occurs to any significant extent during the performance of highly skilled activities such as writing, playing a musical instrument or using a jeweler's screwdriver. The tasks affected are usually those that require a high degree of precision and practice and take several years to acquire. Task specific tremor is a rare phenomenon.

The concept of task specific tremor was first introduced by Dr. John Rothwell in 1979. He described a young man with progressive difficulty in writing, caused by sudden bursts of tremor that occurred whenever the patient's right forearm was put in the position normally used for writing. Dr. Rothwell called this condition primary writing tremor, and this is by far the most common form of task specific tremor. Subsequently, a number of patients with similar difficulties have been reported from around the world.

The cause of primary writing tremor is the subject of many debates amongst neurologists with a special interest in tremor research. There are two main schools of thought: a) that primary writing tremor is a form of ET which has not expressed itself fully (in other words that it is a frustrated form of ET) and b) that it is a variant of writer's cramp (a localized condition resulting from a disease known as torsion dystonia).

The evidence supporting the hypothesis that primary writing tremor is a frustrated form of essential tremor is that both conditions have similar frequencies (between four and eight hertz). In addition, both types of tremor can be relieved by alcohol in about 30% and 50% of cases respectively. Alcohol suppresses these tremors for a period of about two to four hours.

The alternative view that primary writing tremor is a form of writer's cramp is supported by the following observations:

- Patients with writer's cramp develop abnormal posturing of the fingers, wrist and arm during writing but not while performing other actions. Therefore, it is a task-specific condition, but in general, it is one in which abnormal posturing rather than tremor occurs. A similar problem can affect musicians, typists, jewelers and sports persons (i.e., dart players), only while they are performing their specialized activities. These are known as occupational cramps.
- In some cases of writer's cramp, tremor is also evident in the arm that is writing in addition to abnormal posturing.
- The disease (hereditary torsion dystonia) underlying some cases of writer's and occupational cramps have been reported in some instances to cause tremor without any other abnormalities.
- There has been a report of one family in which writer's cramp, writing tremor, and non-task specific tremor were all seen in different members of the family. This finding might appear to indicate that writing tremor was caused by hereditary torsion dystonia. However, careful analysis of the pedigree of this family showed that the affected offspring were the products of a marriage between an individual with torsion dystonia and another with essential tremor. Consequently, I do not think that

any definitive conclusions can be drawn from their descendants, as they may have inherited either or both conditions.

A third alternative is that some cases of primary writing tremor are caused by torsion dystonia and others by essential tremor. Perhaps in some cases neither of these conditions is responsible. My own view is that the term primary writing tremor has been used by different neurologists to encompass three categories of patients:

1. Those with positional dependent tremor (as originally described by Dr. Rothwell) in whom tremulousness occurs (without any abnormal posturing) whenever the arm is put into a specific position such as the posture used for writing.
2. Those in who tremor only appears (without any abnormal posturing) once the skilled task, i.e., writing, has actually begun and not whenever the arm is placed in a specific position.
3. Those patients in whom both involuntary abnormal posturing (dystonia) and tremor occur together whenever the individual starts writing. This last group of patients should in fact be classified as having tremulous writer's cramp, and the exact relationship between this category and the two types of primary writing tremor described above (under categories a and b) requires further study.

It will be evident that there are many difficulties involved in the classification of task specific tremor, and that the mechanisms that induce are not understood. There is the fundamental question of whether it is a type of repetitive strain injury or whether an individual is simply predisposed to develop the problem. I personally suspect that task specific tremor results from the interaction of an innate predisposition with the frequent practice of a particular skill. Perhaps one day we shall know the answer.

In my own practice, I have noticed that primary writing tremor has an average age of onset of about 55 years and is more common among men. I have wondered whether this preponderance of males reflects a bias in British society, as traditionally men of this age group may have had more access to clerical careers.

One peculiar observation has intrigued me, notably that some patients with writer's cramp have found that when writing with either the right or the left hand abnormal posturing and aching developed in the right hand only, irrespective of which hand was actually writing! What is particularly interesting is that I have witnessed exactly the same phenomenon occurring in a patient with primary writing tremor, i.e., tremor appeared in the right hand irrespective of whether the right or left hand was writing.

Fortunately, most cases of primary writing tremor are treatable. I have found propranolol (Inderal®) to be the most useful medication. If propranolol is not effective or tolerated by my patients, I have found that either primidone (Mysoline®) or benzhexol (trihexyphenidyl or Artane®) may be beneficial in some patients. Alternatively, botulinum toxin (Botox®) injections into the appropriate muscles of the forearm can be very effective. The benefit from the injection can last for up to five months before the effect wears off after which the treatment will need to be repeated. Finally, stereotactic surgery to a part of the brain known as the thalamus can often abolish refractory primary writing tremor. This involves passing a fine needle into the substance of the brain using coordinates that have been predetermined radiologically and has a complication rate of about 0.3%

when the operation is performed in a neurosurgical center whose personnel specialize in the procedure.

One other option is to try and learn to perform the affected task with one's non-dominant hand. However, this is difficult for most adults to accomplish. In any case, experience among patients with writer's cramp has shown that in about 10% to 20% of cases the same problem will develop in the second hand. A similar statistic probably applies to patients with primary writing tremor. Nevertheless, this tactic is well worth trying.

Dystonic Tremor

By Christopher F. O'Brien, M.D.

Introduction - Tremor of a body part may be due to one of many neurological conditions. Most physicians are aware of the clinical distinction between resting and action tremor (and the neurological conditions that may cause either). There is, however, a bewildering array of other tremor types, many that do not fit neatly into simple classifications currently in use. One of the most important tremor types not due to ET or Parkinson's disease (PD) is dystonic tremor.

Dystonia is an involuntary muscle contraction resulting in postural abnormalities of the limbs, face, trunk or other body parts. The term dystonia may refer to a diagnosis, a syndrome, or simply a description of muscle contraction.

It is important to know that dystonia from various causes may produce tremor. Proper diagnosis allows for more accurate prognosis and appropriate therapy. Some patients have two conditions simultaneously such as ET and dystonia. The medications and surgery for dystonia are often quite different then for ET or PD. What follows are descriptions of some of the more frequently encountered dystonic tremors.

Dystonic Head and Neck Tremor

Involuntary shaking of the head occurs when neck muscles intermittently contract resulting in jerking movements. If the contractions are rhythmic and balanced on the right and left sides, a tremor typical of ET may result. If the neck muscles get their "faulty signals" from a different region of the brain, an irregular jerking may occur. Often this is unbalanced and the head and neck will twist or jerk more to one side than the other. This type of dystonic tremor is usually part of spasmodic torticollis (also known as cervical dystonia or CD).

Dystonic tremor can be quite variable and may diminish with simple "sensory tricks" such as a light touch or placement of the head against a headrest. This tremor is more common in women than men, begins in young-adult or midlife and is often associated with twisting of the head and neck into abnormal positions. There may be prominent enlargement of one or more neck muscles, and pain may be present. Treatment of dystonic tremor may include oral medications known to help dystonia such as anticholinergics (e.g., trihexylphenidyl (Artane®) or clonazepam (Klonopin®)) or botulinum toxin injections (e.g., Botox®).

The injections have gained wide spread use due to the high success rate. When injected into the correct muscles at the right dose, 75%-85% of patients achieve significant benefit. The injection works by temporarily weakening the spasming muscles thereby reducing tremor severity. The benefit lasts for two to six months on average and must be repeated usually two or three times per year. Horizontal tremor (i.e., "no-no") responds better then vertical tremor (i.e., "yes-yes") due to the arrangement of the neck muscles.

Dystonic Limb Tremor

This diagnosis may be quite challenging to make and treat. Many conditions result in jerking of the limbs. Most dystonic limb tremors occur with action (such as writing) and also involve twisting of the arm or fingers. Some individuals have tremor in association with writer's cramp. For mild dystonic hand tremor, mechanical writing aids or oral medication may be beneficial.

In a selected minority of patients with dopa-responsive dystonia, the tremor may respond to low doses of levodopa (Sinemet®). Botulinum toxin injections are quite useful if a specific muscle group can be identified as the source of jerking. The treating physician must be skilled in muscle localization with EMG guidance. For severe dystonic arm or leg tremor, stereotactic brain surgery may be the only source of relief, either by thalamotomy or thalamic stimulation (DBS).

Dystonic Vocal Tremor

Individuals with spasmodic dysphonia (SD) often go for years without proper diagnosis or are told their condition is due to vocal overuse or psychological problems. SD may cause a strangled sound (due to involuntary spasm of the adductor muscles), a breathy sound (due to overactive abductor muscles) or some chaotic mix of laryngeal muscle spasms. Diagnosis is possible with inspection of vocal cord movement using a laryngoscope by an experienced physician knowledgeable about SD. Treatment with EMG guided injections of botulinum toxin is helpful in approximately 80% of the adductor type, 50% of the abductor type and 40% of the mixed type. Oral medications are not as useful, but clonazepam is the most useful.

Future Research

Much work remains to be done in order to better define the cause and treatment of dystonic tremor. Patients with tremor who are not responsive to ET or PD therapy should consider an evaluation by a specialist familiar with diagnosis and treatment of dystonia. Most movement disorder centers offer the specialized assessment and treatment required.

Resources: Dystonia Medical Research Foundation, (800) 377-3978, the National Spasmodic Torticollis Association, (800) 487-8385, the National Spasmodic Dysphonia Association, (800) 795-6732.

Post-Traumatic Tremor

By Francisco E.C. Cardoso, M.D., and Joseph Jankovic, M.D.

Tremor may occur as a consequence of trauma to the central or peripheral nervous systems. While brain and nerve injuries are relatively common, post-traumatic tremor has been reported infrequently.

One reason for the apparent rarity of post-traumatic tremor may be that the occurrence of tremor after trauma may require genetic, chemical or other “pre-disposition.” Another reason may be that a relationship between trauma and subsequent tremor is not reported or recognized by patients or their physicians. Uncertainty about the maximum latency period allowed between trauma and the onset of tremor for the two to be considered related contributes to probable under-diagnosis of post-traumatic tremor. Intuitively, the shorter the latency between injury and tremor onset, the more likely the two are related. Similarly, more severe injury is more likely to produce tremor.

Closed or, more probably, open head injury may cause damage to any part of the brain; therefore, tremor may be only one component of the post-traumatic neurologic syndrome. Tremor following brain trauma is usually associated with lesions of the cerebellum. This posterior portion of the brain is normally responsible for coordination of movements. Cerebellar tremor is typically classified as “kinetic” tremor, which means that it occurs primarily during movement. The term “intentional” is sometimes used to describe this form of cerebellar tremor.

Kinetic tremor is most evident during eating; the patient often spills liquids and has difficulty using utensils, particularly when eating soup. During neurologic examination, kinetic tremor is best elicited by the “finger-to-nose” test. During this goal-directed movement, the tremor increases in amplitude when the patient’s index finger moves from his nose to the examiner’s finger. Another example of cerebellar tremor typically seen after head injury is “titubation”, an oscillatory (swinging to and fro) movement of the head and trunk.

Another example of post-traumatic tremor is the so-called “midbrain” or “rubral” tremor, resulting from damage to the pathways connecting the cerebellum to the brainstem, including midbrain structures such as the red (rubral) nucleus. Typically, this tremor is present during a voluntary maintenance of posture (i.e., when the arms are outstretched or held in the “wing-beating” position). The midbrain also contains the substantia nigra, damage to which can cause parkinsonian tremor. This tremor is present chiefly when the affected body part is at rest, typically producing a pill-rolling movement of the fingers and hands. Legs, lips, jaw and tongue may also be involved. Some physicians estimate that parkinsonism may be a consequence in up to 1.5% of all brain injuries.

Post-traumatic tremor may also be a consequence of peripheral trauma. Injuries to the peripheral nerves can result in all three types of tremor: rest, postural and kinetic. Although these tremors may remain restricted to the site of the lesion, they sometimes spread to involve other body regions. In a series of patients with peripheral post-traumatic tremor we studied at Baylor College of Medicine, 60% had some evidence of predisposition to tremor such as a family history of ET and/or a prior exposure to certain tranquilizers resulting in tremor as a side effect. Dystonic movements often accompany this type of tremor. In addition, pain, changes in the color and temperature of the skin, abnormal sweating, and atrophy of the bones and nails in the injured part may also appear. The cause of this tremor is uncertain, but it has been hypothesized that peripheral nerve lesions

somehow lead to abnormal activity in the predisposed central nervous system.

Pharmacologic management of post-traumatic tremors is unsatisfactory in most cases. Drugs prescribed for PD or ET, only rarely improve post-traumatic tremors. Thalamotomy or deep brain stimulation of the thalamus may be helpful in some patients, though it is associated with risks including weakness, numbness and possible speech problems. Finally, physical therapy may be useful. For example, using a two to three pound wrist weight may enable a patient with severe kinetic tremor to eat or write.

Psychogenic/Functional Tremor

By *Anthony E. Lang, M.D., F.R.C.P.(C)*

The vast majority of patients suffering from tremor have a primary neurological disorder (such as Parkinson's disease or essential tremor) causing tremor or have the accentuation of a normal or "physiologic" tremor due to a variety of causes such as fatigue, certain drugs or a systemic illness, such as hyperthyroidism. A small proportion of patients presenting to neurologists have a type of tremor that is predominantly caused by psychological or psychiatric factors termed "psychogenic" or "functional" tremor. The terminology is currently under discussion. Some experts prefer the "psychogenic" term because most patients are believed to have underlying psychological or psychiatric causes. However, many of us now prefer the term "functional" because not all patients have underlying psychological causes and many patients find the term "psychogenic" demeaning or stigmatizing. The term "functional" can be used more readily in discussions with patients at the time of presenting the diagnosis when it is critical to get the patient working *with* the healthcare team rather than resenting them.

The occurrence of psychogenic/functional tremor has been recognized for over a century. These types of tremors can involve any part of the body. Psychogenic/functional tremors are often quite prominent and patients with these types of tremors can experience substantial disability. There are a number of clues that the neurologist uses to support the diagnosis. The onset is often quite abrupt and frequently triggered or precipitated by a specific event such as an injury. In the latter situation, litigation or compensation factors are not uncommon.

Like a parkinsonian tremor, psychogenic/functional tremor is frequently present at rest, however, it behaves differently from tremors known to be caused by established neurological diseases in that the same tremor persists when a new position is taken up and continues through a voluntary activity often without changing in severity (whereas parkinsonian tremors typically lessen and ET (when present at rest) worsens. The speed or frequency of a psychogenic/functional tremor varies considerably from time to time in contrast to most tremors due to known neurologic diseases. One feature that physicians search for when examining such patients is fact that when the patient is distracted and forced to concentrate on other things the tremor often goes away or changes its rhythmicity considerably.

Patients with psychogenic/functional tremors commonly have a wide variety of other complaints that are difficult to reconcile with either established neurological disease or with changes found on examination. In addition, the physical examination frequently demonstrates a number of unusual or bizarre features that are not compatible with known patterns of neurological dysfunction (e.g., weakness or changes in sensation that don't behave like anything seen with known neurological disease). Investigations are typically unrewarding or normal, although it is possible for a patient to have both psychogenic/functional tremor and some other neurological problem that may or may not be defined through laboratory investigations (this combination is a great challenge for diagnosis and management).

The course of psychogenic/functional tremor varies greatly from patient to patient. Sometimes only a brief transient period of tremor is seen once in a lifetime usually triggered by a specific stressful event. On the other hand, the symptoms may be variably present for years or even decades with little change.

Underlying psychological factors or psychiatric disturbances in patients suffering from psychogenic/functional tremor also vary considerably. Traumatic past life events may be important, such as sexual abuse in young women. Depression is a very common associated or causative factor. Many patients suffer from so-called anxiety disorders or have very poor abilities to cope with stress. In some patients, even careful psychological evaluation fails to define the specific underlying psychological disturbances.

Importantly, a diagnosis of psychogenic/functional tremor does not imply that the patient is “crazy.” Tremor can be a symptom of primary psychological disturbances just as stress or emotional disruption and can cause numerous other physical complaints such as chest pain, high blood pressure, stomach ulcers, change in bowel habits, etc. Alternatively, this diagnosis does not imply that the patient is purposefully causing the tremor to occur. It is exceedingly rare for patients to actually purposefully contrive their tremor. This is known as “malingering,” a condition not due to primary psychiatric disturbances, but related to an individual’s attempt to “fool the system” for secondary gain such as financial, attention, etc. Even when financial factors are playing a role, such as in the case of litigation or compensation, it is uncommon for overt malingering to be the cause of this type of tremor. Psychogenic/functional tremor is often a very difficult disorder to diagnose. Usually it requires the expertise of a neurologist with considerable experience in diagnosing and managing all other forms of tremor (since it is only with this experience that one can discern the unusual or atypical features that confirm a diagnosis of psychogenic/functional tremor).

Management is even more of a challenge. Where obvious precipitants or psychological factors are playing a role, one may attempt to address these issues directly. However, in many cases, one is faced with a complex interaction of longstanding psychological factors with more recent triggering or contributing factors. Here, management can be very difficult, and many patients retain long-term disability. This disability often forces retirement or substantially interferes with daily activities.

Some time ago, we carried out a 10 year review of 70 patients with a diagnosis of established psychogenic/functional tremor seen in my clinic at the Toronto Hospital. Seventy-three percent had the tremor begin abruptly. Almost 50% experienced maximal disability at onset (in contrast to the usual slowly progressive nature of most tremors of primary neurologic origin). Forty-six percent had a static or nonprogressive course (again atypical for tremors with neurologic origin) while 17% had a fluctuating course. Tremors usually started in one limb and spread rapidly to involve the entire body or to demonstrate a mixed or atypical distribution. Fifty-one percent of the patients had a precipitant for the psychogenic/functional tremor; most commonly some form of physical injury such as motor vehicle accident, head, neck, limb or back injury.

Spontaneous resolution and recurrence were common, as were distractibility and “entrainment” which refers to a change of the original tremor frequency to match the frequency of a requested repetitive task in another limb. The tremor had been present for an average of over four years in these 70 patients. There was nothing about the severity or duration of tremor that excluded the possibility of a psychogenic cause.

When presented with the diagnosis in a supportive and careful fashion, some patients are quite receptive and willing to work with a psychologist or psychiatrist recognize and hopefully successfully deal with the causative psychological factors.

On the other hand, when presented in the most supportive and understanding fashion, some patients are extremely resistant to the suggestion that psychological factors could be primary and continue to go from doctor to doctor searching for an alternative explanation. Unfortunately, many doctors either do not present the diagnosis appropriately or tell the patient that there is nothing wrong and that they will get better. Neither of these approaches is very helpful or productive.

In summary, psychogenic/functional tremor is an uncommon but well-established form of tremor. The diagnosis usually can be made with certainty by the careful (sometimes extended) assessment of an experienced physician. This accurate diagnosis allows avoidance of subsequent, unnecessary, expensive and sometimes risky investigations. Proper diagnosis also allows the avoidance of unnecessary trials of various drugs designed to suppress tremor, all of which may have substantial side effects.

Management of psychogenic/functional tremor can be difficult and despite the fact that these patients do not suffer from underlying primary neurological dysfunction, prognosis, especially in well-established cases, must be guarded. Some patients can be taught to entrain their tremor to lower and lower speeds/frequencies and eventually stop the tremor altogether. How often and for how long this approach will work remains uncertain.

Tremor of Psychiatric Origin

By Stewart A. Factor, D.O.

Tremor is the most common of movement disorders. It is characterized by involuntary rhythmic oscillating movements of almost any body part due to contractions of muscles, and is generally a feature of physical or organic disease. There are many neurologic and non-neurologic disorders that have tremor as a symptom.

Tremor may be the primary feature, as in ET, or it may be one of several signs of disease, as in Parkinson's disease, multiple sclerosis or stroke. Tremor is generally classified according to its relationship to movement as it may occur at rest, with maintaining posture or with goal-related action. Most tremors caused by organic disease can be classified in this manner. In addition, the tremor is usually fairly consistent in amplitude and frequency, as well as direction of movement.

There is one cause of tremor that is frequently overlooked and that is tremor of psychiatric origin (psychogenic). It may be related to psychiatric disease in a number of ways. Tremor occurs in patients with disorders such as depression or anxiety, and may represent a manifestation of these. Tremor may also occur as a manifestation of conversion hysteria, as a substitute for an emotional problem. In this case, the tremor is not under voluntary control, and the patient is unaware of the source of the problem. In both situations, psychotherapy may be helpful. Tremor may also occur in malingerers, people who intentionally try to mislead physicians.

The diagnosis in these three situations is not always easy to determine. Frequently, a psychiatric diagnosis cannot be made. In general, a neurologist who specializes in caring for patients with tremor can recognize that tremor is psychogenic in nature.

Psychogenic tremor has a number of unusual features which may alert the suspicions of the evaluating physician. Psychogenic tremor often comes on abruptly and reaches peak severity almost immediately. It does not appear to follow a progressive course, but may come and go in an unpredictable manner. The tremor may occur in the arms and legs, on one side or both sides of the body, or these patients may have head tremors. It is often present in equal severity at rest, with maintaining a posture and with action. This makes it extremely difficult to classify.

The tremor can be quite variable in frequency and amplitude with a noticeable increase caused by attention to the affected limb and decrease with distractions. These variations can be quite dramatic. Even the direction of tremor changes from examination to examination.

There can be selective disabilities with psychogenic tremors. An example of this is a patient who can write properly, but has difficulty drawing. In some cases, very bizarre characteristics may be present. In one case described by Dr. Koller and his colleagues, hand tremor disappeared when the hand was held, but then occurred in another limb. In addition, if patients are asked to perform certain tasks such as finger tapping or toe tapping, the tremor may take on the frequency of that movement. Patients may otherwise have a totally normal neurologic examination or they may have other abnormalities that would fall under the rubric of non-neurologic or psychogenic. These include give-way weakness, strange non-physiologic sensory losses, pain or strange abnormalities of gait. There may also be a past history of other psychogenic disorders or undiagnosed strange syndromes.

In addition, psychogenic tremor can occur in patients with organic movement disorders. These patients often respond poorly to medications that typically improve tremor. If the tremor stops, it often does so abruptly. Many patients with psychogenic tremor are involved in lawsuits related to the cause. Because of the unwillingness of physicians to diagnose a psychiatric cause, patients consult multiple physicians.

Making the diagnosis of psychogenic tremor may be difficult. The clinical features should alert suspicion that the tremor may be of psychiatric origin; however, other causes must be kept in mind. In some disorders the tremor may be unusual. An example of this is Wilson's disease. This is a disease of abnormal copper metabolism. It may be the cause of a variety of involuntary movements. A blood and urine test can lead to the diagnosis of Wilson's disease, and these tests should be done. Tests for other causes of tremor should also be performed so that one will be certain that an organic cause is detected if one exists.

Once testing is complete and all other possibilities are examined, there are some aspects that can be very helpful in making the diagnosis. First, the opportunity to see the patient without the tremor, in a situation in which the patient does not know he is being watched, can be helpful. Some physicians use a placebo test. Although a disappearance of symptoms with placebo can be used to diagnose psychogenic tremor, a negative test (meaning that the tremor persists) does not mean that the cause is not psychiatric. Advice from a psychiatrist can also be helpful.

The patient's reaction to the diagnosis varies depending on the underlying psychiatric diagnosis. For the most part, patients with conversion hysteria or depression will be receptive to this diagnosis and will be willing to see a psychiatrist as part of the diagnosis and treatment. Some people, however, will become angry and will never return.

Two directions in therapy can be taken. Some physicians would rather not tell the patient that they suspect a psychiatric diagnosis. Instead they will suggest to the patient that the illness is not permanent and should clear up over a certain period of time. This gives the patient an opportunity to show improvement on his own. Some patients are very susceptible to suggestion. When the symptoms do not go away, the patient ends up seeking another medical consultation. The other possibility is to inform the patient that it is believed that there is an underlying psychiatric problem. This would lead to psychotherapy, which includes counseling and sometimes medications. As previously noted, response to this is varied.

Despite these options and possibilities, many patients can go on indefinitely with these tremors. For those patients who are seen early after the onset of tremor, there is a better possibility of obtaining a cure. However, for those who have continued on for very long period, the chance for improvement becomes less.

How common is psychogenic tremor? The bulk of the literature on psychogenic tremor is based on two reports of 26 patients. This small number suggests that this disorder is rare. One group evaluated the incidence of psychogenic disorders in a neurology clinic. Of 4,470 patients seen, 405 had psychogenic disorders (9%). Of the 405 patients, 11 had tremor (.24% of the total number of patients). This supports the notion that psychogenic tremor is rare.

However, since the clinical features of psychogenic tremor have been more clearly delineated, it's become obvious to movement disorders specialists that psychogenic tremor is probably more

common than previously thought. An example of this comes from my own clinic. Out of 450 patients seen in the Movement Disorders Clinic at Albany Medical College from 1988 to 1991, seven had psychogenic tremors (1.5%), or 14 percent of tremor patients referred. Psychiatric disease is an important cause of tremor, one which we are only beginning to diagnose, understand and, in a multidisciplinary research center, learn to treat.

Tremor and Myoclonus

By Mark Hallett, M.D.

Tremor is an involuntary movement characterized by rhythmicity. Ordinarily the clinical diagnosis of an involuntary movement as a tremor is fairly easy. The more difficult aspect is deciding what type of tremor it is. However, on some occasions the diagnosis is not straightforward. Another type of involuntary movement that can be rhythmic is myoclonus, which is typically characterized by the quickness of the individual movements. Ordinarily the movements come irregularly, but rarely can be rhythmic.

There are many types of myoclonus. Indeed, the classification of myoclonus has been somewhat difficult and, at times, controversial. Personally, I divide the myoclonias into those that are fragments of epilepsy and those that are not. The former would be called epileptic myoclonus and the latter non-epileptic myoclonus. Epileptic myoclonus is seen in patients with (1) epilepsy, (2) brain damage caused by lack of oxygen, (3) metabolic disturbances such as kidney failure and (4) other brain diseases such as viral encephalitis.

The causes of non-epileptic myoclonus are varied. One is essential (meaning of unknown origin) myoclonus; this is typically a genetically determined disorder without obvious brain disease that is limited to the movement problem. Another is nocturnal myoclonus; myoclonus at night. One form of this is described as periodic movements in sleep in which movements of the legs occur about once every 20 seconds. This has rhythmicity, but the frequency is so low that there is no confusion with tremor. Another form is a type of movement in patients with dystonia, called dystonic myoclonus. Dystonia is ordinarily characterized by prolonged abnormal postures, but sometimes the movements are quicker.

Most of these types of myoclonus can be quick and rhythmic. Physicians can be confused about the correct diagnosis. In most cases, electrodiagnostic studies can clarify the confusion. One can measure the electrical activity of the muscles, the electromyographic (EMG) activity, using electrodes pasted on the surface of the skin, but needles inserted into the muscles may be required.

Measurement of the associated brain electrical activity, the EEG, may also be useful. The distinction is crucial because the therapy for tremor and myoclonus usually differs. A rare type of myoclonus, ordinarily rhythmic, is called spinal or segmental myoclonus. This occurs when a part of the spinal cord develops its own abnormal generator producing movements of a limb or part of the trunk.

One disorder that has been difficult to classify has been called palatal myoclonus. This is the abnormal movement of the soft palate in a quasi-rhythmic pattern at a frequency of approximately 1 - 3 Hz (movements per second). The disorder can arise in isolation from unknown cause or can result from a lesion in the brain. Sometimes there is a click with each movement of the palate, audible even several feet from the patient. At the International Congress of Movement Disorders in Washington, D.C. in 1990, it was decided that a more accurate classification would be palatal tremor.

Tremor and myoclonus are genetically related. Both ET and essential myoclonus can run in families as autosomal dominant traits (meaning that children of an affected parent have a one in two chance of inheriting the disorder). In some families with genetically determined tremor, some individuals also have ET or only tremor. In some families with genetically determined tremor, some individuals

also have myoclonus or only myoclonus. Both disorders in these families can be improved with alcohol, providing another link between the two. The pathophysiology by which a similar genetic background can produce tremor in one person and myoclonus in another is not at all clear, yet clearly these movement disorders do have important similarities. In at least some of these families, the genetic abnormality is in the gene leading to the disorder called myoclonus dystonia.

Neurotoxins and Tremor

By Anthony E. Lang, M.D., F.R.C.P. (C)

A neurotoxin is an agent capable of eliciting irreversible dysfunction and damage to the nervous system due to death of nerve cells. Sometimes exposure is so severe and damage so great, death is the result. Toxic exposure may be divided into that occurring with acute high-level exposure and chronic low-level exposure. If patients survive the acute form, chronic and permanent neurologic deficits commonly persist.

Symptoms of neurotoxic exposure are broad and sometimes nonspecific. Questions arise as to whether an individual has had sufficient exposure to a possible causative neurotoxin to result in neurologic dysfunction. This uncertainty causes conflicts in compensation or litigation procedures. There has been increasing public awareness and concern regarding the potentially harmful effects of exposure to low levels of a number of environmental toxins, for example, those originating in industrial waste. Patients commonly express concerns that past exposure to a possible toxin contributed to the development of a neurodegenerative disorder such as Alzheimer's disease (AD), Parkinson's disease (PD) or motor neuron disease (Amyotrophic Lateral Sclerosis - ALS). However, it must be emphasized that the role of modern industrial pollution in the causation of such common neurologic diseases is largely unknown.

More often, unequivocal toxic exposure occurs in workers involved in specific industries that utilize the potential toxin in a manufacturing process. Routes of exposure vary depending on the nature of the toxin and the type of work involved. Individuals may be exposed by inhalation, direct skin contact and accidental oral ingestion.

Not typically included in the category of "neurotoxins" are illicit chemicals utilized by drug abusers through various routes including intravenous injection. One exception, mentioned later, is the neurotoxin MPTP has caused permanent Parkinsonism in drug abusers. Neurotoxins can be subdivided under the classifications of metals, solvents, pesticides and other agents.

Metals

Our discussion will be limited to the consequences of environmental exposure to excessive amounts of heavy metals. It is noted there is considerable interest in the role of metals in causing neurologic diseases due to the body's metabolism and handling of metals in the absence of overexposure. Examples of this include iron in Parkinson's disease and the rare disorder, Hallervorden-Spatz disease, and aluminum in Alzheimer's disease. Another well-known example is Wilson's disease, which is a genetic disorder of abnormal copper metabolism.

Most acute heavy-metal exposures result in a syndrome known as encephalopathy which manifests confusion, poor memory, mood changes, headache and possibly coma and seizures. A non-specific fine postural tremor of the arms may be present in this acute stage. Chronic exposure to certain heavy metals, including lead and arsenic, results in damage to peripheral nerves usually causing weakness in the hands and feet and tingling and loss of sensation in these areas. A peripheral neuropathy (damage to the peripheral as opposed to central nerves) may also result in a postural tremor of the limbs, but typically this is not a prominent feature of the neuropathy of chronic metal exposure.

Manganese has a selective ability to damage the basal ganglia region of the brain causing a syndrome very much like Parkinson's disease. This may be accompanied by varying types of tremor, including a tremor at rest and/or postural tremor. This syndrome has been seen in manganese ore miners in Chile as well as those involved in manganese smelting and individuals exposed to manganese oxide in a selected number of occupations such as the manufacture of paints and fireworks and possibly welding. Although this disorder is rare, insights into the mechanism of action of manganese and its ability to cause selective toxic effects in this region of the brain may eventually aid our understanding of naturally-occurring diseases.

Acute exposure to mercury may result in kidney failure, bowel hemorrhaging and death. Neurologic problems typically occur in survivors over the next 24 hours. Acute exposure to inorganic mercury may occur in the manufacture of scientific and electronic instruments, and work with dental amalgam. Symptoms include clumsiness and tremor.

The results of acute exposure to methyl mercury were seen between 1953 and 1956 when inhabitants of the Minamata Bay region of Kyushu Island in Japan ingested fish tainted with large levels of this toxin ("Minamata disease"). Severe neurologic consequences included numbness of the limbs, lips and tongue followed by clumsiness, slurred speech, deafness, blindness, spasticity and intellectual impairment.

In contrast, chronic, low-level mercury exposure is possible in a variety of occupations including workers exposed to mercury vapors or mercury in dental amalgam (it should be emphasized that there is no evidence to support the concern that neurologic disease may be caused by tooth fillings containing dental amalgam). Chronic exposure typically results in postural tremor, deterioration of intellect and changes in emotional state. Historically, nitrates of mercury were used in felt making and hat makers were particularly at risk. This accounts for the Mad Hatter character in Lewis Carroll's "Through the Looking Glass" as well as the term "Hatter's Shakes" to describe tremors seen in these individuals.

Organic solvents

Organic Solvents are volatile chemical compounds used industrially to extract, dissolve or suspend materials that are poorly soluble in water. As with heavy metals, exposure to organic solvents may result in a variety of both systemic and neurologic signs and symptoms. Rarely is tremor a major or predominant component. Certain organic solvents may be more likely to cause tremor.

Carbon disulfide used in insecticides, the rubber industry, the preparation of rayon viscose fibers, and in grain storage may selectively damage the basal ganglia as well as other regions of the brain. Parkinsonism's resting and postural tremors may occur as a result of toxic exposure to carbon disulfide. But it is usually the slowness and stiffness of this disorder which predominate rather than the tremor.

Toluene is an organic solvent used in the production of adhesives, glues, lacquers, paint thinners, rubbers and other substances. Toxic work-related exposure can be seen, for example, in rotogravure printers. It is more common now to see the consequences of acute and chronic exposure in glue sniffers. Tremor may be one syndrome component comprising mental changes, clumsiness, incoordination, unsteady gait and damage to peripheral nerves.

Pesticides

There is considerable interest in the possible role of rural living and pesticide or fungicide exposure in causing Parkinson's disease. Support for this comes from several sources, including the chemical similarity between the pesticide paraquat and the neurotoxin MPTP. Despite intense research in this field, the role of environmental exposure in PD remains uncertain.

The largest group of pesticides is the organophosphate insecticides. These affect transmission between nerves and muscles as well as causing dysfunction in the autonomic nervous system (which controls such faculties as pupillary function, blood pressure, and bowel and bladder control) and mental changes. There are numerous other types of pesticides, most with the same profile of toxicity.

Carbaryl has been used commercially on cotton and corn to control grasshoppers and gypsy moths and is also found in some household gardening sprays. This is less toxic than the organophosphates; so much greater exposure is required to produce neurologic symptoms. Dermal (through the skin) and oral exposures have resulted in a variety of symptoms similar to those of the organophosphates. Interestingly, tremor may occur as a complication of chronic exposure. This may relate in part to central effects on the neurotransmitter dopamine.

Finally, chlordecone (Kepone®) used to control weevils, ants and roaches, has caused tremor ("Kepone shakes") and other symptoms in heavily-exposed industrial workers in Virginia.

Other Neurotoxins

As mentioned earlier, this article will not review the consequences of substance abuse. In addition to abusing chemicals that transiently affect neurologic function, drug addicts and alcoholics also may be exposed to toxins that permanently damage the nervous system. One example already discussed is toluene. Alcoholics may imbibe methyl alcohol (sometimes used by others in suicide attempts). Methyl alcohol exposure can also selectively damage the basal ganglia resulting in Parkinsonism, dystonia and tremor.

In the early 1980's, a small number of heroin addicts in California developed acute, severe and permanent Parkinsonism after they injected a "bad batch" of homemade "heroin" which contained the neurotoxin MPTP. Both postural and resting tremors have been seen as part of this syndrome. The discovery of the selective ability of MPTP to damage the part of the brain most prominently affected in classic Parkinson's disease (the substantia nigra pars compacta) revolutionized research in almost every aspect of PD, including the cause (possible environmental exposure), specific mechanisms of cell damage and death (e.g., mitochondrial dysfunction), the nature of symptoms (e.g., tremor as a manifestation of substantia nigra damage) and possible neuroprotective therapies (e.g., antioxidant drugs).

Research is extremely active in the area of one further category of neurotoxins, the excitatory amino acids. This is a large and important topic, which is beyond the scope of this discussion. With respect to disorders associated with tremor, it had been proposed (but not proven) that exposure to the excitatory amino acid betamethylamino-L-alanine (BMAA) contained in the cycad plant may cause a form of Parkinsonism associated with dementia and motor neuron disease seen in the Chamorro inhabitants of Guam. Excitatory amino acids may cause neurologic damage through chronic exposure in the environment (most often via oral intake). They are also normal constituents of the brain, and there is increasing evidence that they play a role in acute neurologic dysfunction in disorders such as stroke, brain anoxia (the loss of sufficient oxygen for brain function) and seizures.

Finally, excitatory amino acid toxicity may also play a role in slowly progressive neurodegenerative disorders such as Huntington's disease and PD.

As one can gather from this brief review, tremor is rarely a consequence of toxin exposure, and then it hardly ever occurs as an isolated symptom. More often than not, when an individual consults a physician complaining of tremor and claims an association with an environmental exposure, the association is no more than coincidental, and the tremor is due to some other much more common neurologic problem.

However, each claim must be considered carefully, since there is still a great deal that we do not know about neurotoxins. On the other hand, we also see patients feigning neurologic diseases attempting to obtain compensation, and in these cases the tremor would relate to primary psychiatric factors such as malingering or "compensation neuroses."

A major challenge to industrialized nations in the 21st century will be the elimination of toxic waste. The importance of this goal to the preservation of our environment is well established. How low-level toxic exposure affects our health is still not certain. Further research is required to establish the possible roles of exogenous (coming from the environment) and endogenous (formed within the body) toxins in the causes of "naturally" occurring neurologic diseases such as essential tremor and Parkinson's disease.

Leg Tremor versus Restless Legs Syndrome

By Joseph Jankovic, M.D.

There are many conditions that can cause tremor in the legs, including Parkinson's disease (PD), ET, and orthostatic tremor. At least half of all patients with PD have tremor involving their feet or legs, usually present when they are sitting or lying. Typically the "rest" tremor associated with PD is usually accompanied by other PD symptoms such as slowness of movement (bradykinesia), stiffness of muscles (rigidity) and gait, and postural difficulties.

ET chiefly involves the hands, but may also involve the head, voice and other body parts, including the legs. Among 350 ET patients, 13.7% had leg tremor. Leg tremor associated with ET is rarely troublesome for the patient, although it can be a source of marked embarrassment. Both PD and ET tremors can interfere with driving because of the rhythmic pressure applied to the gas pedal or brake.

Orthostatic tremor is probably the most disabling form of leg tremor. Hardly visible to the naked eye, it can be easily palpated or recorded by EMG. The tremor is typically present when the patient is standing, and is often perceived by the patient not as a tremor, but as an uncomfortable sensation or leg cramps while standing. The sensation is relieved immediately after sitting or lying down.

Leg tremor can lead to a perception of leg restlessness and it often confused with "restless legs syndrome." This disorder is relatively common, but it is frequently not recognized (even by physicians), and it is often wrongly attributed to simple anxiety. Restless legs syndrome may occur alone as a familial (hereditary) disorder and as a complication of PD or peripheral neuropathies.

Although other body parts may be involved, the symptoms of restless legs syndrome are generally confined to the lower limbs. In addition to the sensation of restlessness, patients often complain of a "creepy-crawly" sensation and feelings of "pins and needles" in the legs. These disagreeable sensations, called "paresthesia," usually occur at night, but they may also be present during daytime hours. As a result of the daytime restlessness, the patients are unable to sit still and may not be able to travel long distances in the car or sit still on an airplane.

Nearly all patients with restless legs syndrome have associated "periodic limb movements in sleep", manifested by rhythmic leg twitches occurring one to four times per second. These "periodic limb movements in sleep" may produce kicking movements and as such, are usually more bothersome to the bed partner than to the patient.

The feelings of restlessness and leg discomfort can be temporarily relieved by walking about, massaging or stretching muscles, and doing leg exercises. Levodopa, dopamine agonists, benzodiazepines and narcotic analgesics often relieve the symptoms of restless legs syndrome.

To receive more information, contact the Restless Legs Syndrome Foundation, 1610 14th Street NW, Suite 300, Rochester, MN 55901, (507) 287-6465, www.rls.org.

Truncal Tremor

By Eduardo S. Tolosa, M.D., and Pilar Sanz, M.D.

Introduction

The term tremor refers to a rhythmic oscillatory movement of a body part. Tremor can occur at rest when muscles are not voluntarily activated, or during action when muscles are voluntarily contracted. Types of action tremors include postural, kinetic and isometric tremors.

Although hands are the most frequently affected body part in tremor, other areas can also be involved such as the larynx in vocal tremor, the cervical muscles in patients with head tremor or the midline paravertebral and abdominal muscles in truncal tremor. The locations and characteristics of tremor depend in part on the cause of the involuntary movements.

ET typically involves the hands and less commonly can, in addition, involve the vocal cords, the head or the legs. In Parkinson's disease patients, it is not uncommon for tremor to occur in the legs or in the jaw, for example, in addition to the arms. Involvement of the trunk occurs very rarely in patients with either of these two common disorders. On the other hand, cerebellar tremor frequently also affects the limbs, but unlike ET or PD, can also involve the trunk. One other condition, orthostatic tremor, also causes truncal tremor.

Truncal Tremor in PD or ET

While uncommon in both disorders, in PD the trunk can tremble as an extension of a prominent arm or leg tremor; in ET it can be encountered in patients with severe generalized tremor. Also, in a minority of ET patients, truncal tremor occurs as consequences of leg and trunk tremor when standing. Such patients are considered to have orthostatic tremor secondary to ET. In patients with typically moderate ET involving the hands, external influencing factors such as intense fatigue or stress can trigger tremor in body areas other than the hands such as the legs and trunk. Under such circumstances, ET patients may complain of disabling tremor on standing that subsides when the aggravating factors remit.

Truncal Tremor in Cerebellar Disorders

Cerebellar disease results in three types of tremor: postural tremor, kinetic tremor and titubation. Postural tremor can be demonstrated by asking the patient to hold the arms horizontally or to elevate the legs while in a supine position. Kinetic tremor can be demonstrated by performing the finger-to-nose or the heel-to-shin test. Titubation is the word used to describe a postural action tremor of the trunk that occurs when the patient is either standing or sitting. It consists of a rhythmic oscillation of the head or trunk forward or backward or side to-side. Titubation can appear in isolation or associated with kinetic tremor of the limbs. Truncal titubation is always associated with head titubation, but head titubation can occur alone. Truncal tremor in cerebellar disorders can frequently be disabling, interfering with sitting or standing. Titubation of the trunk is frequently thought of as a sign of midline cerebellar lesions but probably has less localizing value than has traditionally been taught.

Cerebellar truncal tremor is encountered in patients with multiple sclerosis or with damage to the cerebellum following head trauma. Other causes of cerebellar truncal tremor are structural (e.g., tumors) and hereditary degenerative disorders.

Truncal Tremor as a Manifestation of Dystonia

Abnormal movements due to sustained muscle contractions, frequently causing twisting and repetitive movements of abnormal postures are described as dystonic. Dystonic movements can affect any part of the body and are very frequently aggravated with voluntary movements. Depending on the location of the dystonia, several clinical syndromes have been described. When it involves one arm and interferes with writing it is called writer's cramp. If the dystonia involves the neck, patients experience involuntary neck twisting called spasmodic torticollis. The paravertebral muscles of the trunk, the pelvic and the abdominal muscles can be sites of dystonic spasms both generalized dystonia of childhood and in adult-onset focal dystonia, causing repetitive back-and-forth truncal and pelvic movements. The spasms can appear while sitting, standing and walking; that is, when muscles are activated, and generally disappear when the patient is lying down and the muscles are relaxed.

In patients with dystonia, it is not infrequent to observe a tremor in the part of the body affected by the dystonic spasms. This type of tremor is called dystonic tremor and is most noticeable when patients are voluntarily trying to counteract the involuntary spasms. At times, dystonic tremor is more prominent than the actual dystonic spasms and may even be the only clinical manifestations. In such patients, it may be difficult to decide whether they are suffering from a tremor disorder, e.g., ET or primary writing tremor or from a focal dystonia. In some instances, truncal tremor can occur in a patient without other signs and symptoms of dystonia. In such cases it could represent an isolated manifestation of dystonia.

Orthostatic Tremor

The most typical tremor affecting the trunk is that known as orthostatic tremor (OT), which consists of involuntary shaking of the legs and the trunk on standing. The American neurologist, Dr. Kenneth Heilman, first described OT in 1984. OT is not present when the patient walks, sits or lies down. Generally, a primary or idiopathic form of OT is recognized. In these patients, no other identifiable neurologic disorder is present and electromyographic (EMG) recordings identify a frequency of 14-16 Hz.

In patients with primary OT, the legs and truncal tremors can vary from quivering sensations in the legs to shaking of the whole body. The presenting symptoms are frequently not of tremor, but rather of unsteadiness on standing or leg stiffness. One striking finding is that the tremor is abolished when the patient is lifted off the floor. Arm tremors in patients with primary OT are uncommon, but if present, consist of 14-16 cycle (14-16 Hz) tremor of the outstretched hands and are similar clinically to ET of the hands. Neurophysiologic studies in OT patients suggest a central origin and that the tremor is generated in structures concerned with the maintenance of posture.

In some ET patients, orthostatic tremor occurs, and there may be some overlap between primary OT and ET in some cases. A positive family history of tremor identical to ET, for example, has been described in some OT individuals. There are thought to be clinical and physiologic as well as pharmacologic differences between these types of tremors.

Other Movement Disorders That Affect the Trunk and Are Not Tremor

Segmental myoclonus is a rhythmic movement generally of low frequency (around 1 Hz) which presents at rest and on walking and even during sleep. The movement has its origin in focal lesions of the spinal cord caused by problems such as cervical myelopathy, demyelination, infections or

spinal anesthesia and often is restricted to one somatic level, for example, one limb and the adjacent trunk. Myoclonus can also involve the trunk in the disorder called propriospinal myoclonus that occurs also in patients with lesions of the spinal cord. These patients have repetitive axial myoclonic muscle spasms causing symmetric flexions of the neck, trunk, hips and knees. The movements, though, are not rhythmic.

Skeletal myoclonus, or myorhythmia, is also a very slow (2-4 Hz) rhythmic movement that can affect the trunk, alone or with palatal myoclonus. The limbs are very frequently involved. This type of movement disorder can occur in patients with cerebellar degeneration or with brainstem infarction (tissues dying due to lack of blood supply). Other conditions that may produce involuntary movements of the legs or trunk when standing are clonus at the ankle or knee in patients with spasticity generally secondary to spinal cord lesions, and action myoclonus (muscle jerks that occur during movements) of the legs that can be seen in a variety of disorders and are typical of postanoxic (loss of oxygen supply) states or induced by certain toxins or drugs, (e.g., bismuth salts) or due to some degenerative disorders of the nervous system.

Treatment of Truncal Tremor

The treatment of a truncal tremor depends on its cause or origin. In patients with PD or ET who complain of truncal tremor, an uncommon situation, treatment should be with the drugs usually employed for tremor reductions in these conditions. Primary OT is known to improve with clonazepam, and several cases have been described that have responded to primidone and to phenobarbital. Beta-blockers and alcohol that are useful in ET are rarely useful in primary OT.

Dystonic truncal tremor can be treated with anticholinergic drugs such as trihexyphenidyl (a common antiparkinson drug which is also used in the treatment of dystonic syndromes). In segmental myoclonus, treatment should be directed to the cause, for example, the surgery for spinal tumors, but good results with clonazepam have also been reported in symptomatic treatment. Other drugs useful in the treatment of the various myoclonic disorders that resemble tremor include piracetam, sodium valproate, and 5-hydroxytryptophan with carbidopa.

Treatment of truncal titubation in cerebellar disorders is most difficult. The cause of the cerebellar syndrome should be identified, and special care should be taken to identify treatable causes, e.g., drug toxicity. When possible, specific treatment should be instituted. There are presently no effective medications for the treatment of cerebellar truncal tremor, but isoniazid, administered with care, can be effective in an occasional patient.

The Future: Prospects for Better Treatments and a Cure

The Drug Development and Approval Process

By U.S. Food and Drug Administration, FDA.gov, Reprinted with permission

The path a drug travels from a lab to your medicine cabinet is usually long, and every drug takes a unique route. Often, a drug is developed to treat a specific disease. An important use of a drug may also be discovered by accident.

For example, Retrovir (zidovudine, also known as AZT) was first studied as an anti-cancer drug in the 1960s with disappointing results. Twenty years later, researchers discovered the drug could treat AIDS, and Food and Drug Administration approved the drug, manufactured by GlaxoSmithKline, for that purpose in 1987.

Most drugs that undergo preclinical (animal) testing never even make it to human testing and review by the FDA. The drugs that do must undergo the agency's rigorous evaluation process, which scrutinizes everything about the drug--from the design of clinical trials to the severity of side effects to the conditions under which the drug is manufactured.

Stages of Drug Development and Review

Stage 1

Investigational New Drug Application (IND) - The pharmaceutical industry sometimes seeks advice from the FDA prior to submission of an IND.

Sponsors - Companies, research institutions, and other organizations that take responsibility for developing a drug. They must show the FDA results of preclinical testing in laboratory animals and what they propose to do for human testing. At this stage, the FDA decides whether it is reasonably safe for the company to move forward with testing the drug in humans.

Stage 2

Clinical Trials - Drug studies in humans can begin only after an IND is reviewed by the FDA and a local institutional review board (IRB). The board is a panel of scientists and non-scientists in hospitals and research institutions that oversees clinical research.

IRBs approve the clinical trial protocols, which describe the type of people who may participate in the clinical trial, the schedule of tests and procedures, the medications and dosages to be studied, the length of the study, the study's objectives, and other details. IRBs make sure the study is acceptable, that participants have given consent and are fully informed of their risks, and that researchers take appropriate steps to protect patients from harm.

Stage 3

Phase 1 studies are usually conducted in healthy volunteers. The goal here is to determine what the drug's most frequent side effects are and, often, how the drug is metabolized and excreted. The number of subjects typically ranges from 20 to 80.

Stage 4

Phase 2 studies begin if Phase 1 studies don't reveal unacceptable toxicity. While the emphasis in Phase 1 is on safety, the emphasis in Phase 2 is on effectiveness. This phase aims to obtain preliminary data on whether the drug works in people who have a certain disease or condition. For

controlled trials, patients receiving the drug are compared with similar patients receiving a different treatment--usually an inactive substance (placebo), or a different drug. Safety continues to be evaluated, and short-term side effects are studied. Typically, the number of subjects in Phase 2 studies ranges from a few dozen to about 300.

Stage 5

At the end of Phase 2, the FDA and sponsors try to come to an agreement on how large-scale studies in Phase 3 should be done. How often the FDA meets with a sponsor varies, but this is one of two most common meeting points prior to submission of a new drug application. The other most common time is pre-NDA, right before a new drug application is submitted.

Phase 3 studies begin if evidence of effectiveness is shown in Phase 2. These studies gather more information about safety and effectiveness, studying different populations and different dosages and using the drug in combination with other drugs. The number of subjects usually ranges from several hundred to about 3,000 people.

Stage 6

Post-market requirement and commitment studies are required of or agreed to by a sponsor, and are conducted after the FDA has approved a product for marketing. The FDA uses postmarket requirement and commitment studies to gather additional information about a product's safety, efficacy, or optimal use.

Stage 7

New Drug Application (NDA) - This is the formal step a drug sponsor takes to ask that the FDA consider approving a new drug for marketing in the United States. An NDA includes all animal and human data and analyses of the data, as well as information about how the drug behaves in the body and how it is manufactured.

Stage 8 & Approval

When an NDA comes in, the FDA has 60 days to decide whether to file it so that it can be reviewed. The FDA can refuse to file an application that is incomplete. For example, some required studies may be missing. In accordance with the Prescription Drug User Fee Act (PDUFA), the FDA's Center for Drug Evaluation and Research (CDER) expects to review and act on at least 90 percent of NDAs for standard drugs no later than 10 months after the applications are received. The review goal is six months for priority drugs.

Clinical Trials

By Ira Shoulson, M.D.

Without clinical trials, we would have little confidence in the reported benefits of medical procedures, therapies and devices. The clinical trial has become the definitive method for scientific assessment of medical value and is the hallmark of evaluation by regulatory agencies such as the Food and Drug Administration (FDA). As one indication of importance, the National Institutes of Health (NIH) supports approximately 1,000 clinical trials each year at a cost exceeding \$136 million. This reflects the importance that our society places on a dispassionate analysis of promising health interventions.

One of the earliest clinical trials evaluated several treatments for scurvy in 12 sailors on board ship. In this 18th century trial, one of the sailors given oranges and lemons recovered dramatically. This observation in a controlled setting provided the first substantive therapy for scurvy, a disorder determined eventually to result from vitamin C deficiency. Clinical trials have since become more rigorous, sophisticated and expensive.

A clinical trial is an unbiased study in humans comparing the value of one or more therapeutic interventions with a control. The clinical trial has well-defined points of starting and terminating the study. The design of the trial is controlled so as to minimize potential bias. The control group is similar or matched in relevant respects (such as age and, in the case of ET patients, tremor severity) to the group which receives the experimental intervention. This control feature is the basis for reasonably attributing differences in outcome to the intervention under examination.

Modern clinical trials involve considerable planning and effort with respect to design, implementation, analysis and interpretation of the results. In practice, these aspects of a trial take several years to complete and usually involve many investigators and subjects, often in the setting of multiple research sites. The design of a clinical trial is planned and carried out by experienced investigators with a variety of collaborators and consultants including nurses, pharmacists, research scientists, biostatisticians, analysts/programmers and lay persons.

Clinical trials are sponsored by a variety of sources including pharmaceutical companies, foundations and governmental agencies. In the United States (U.S.), the NIH is a major sponsor of clinical trials. Regulation and supervision of clinical trials in the U.S. is carried out at several levels including local review by the institutional entity responsible for the trial (Institutional Review Board or IRB) and federal review by a number of agencies including the FDA or NIH.

The design includes a protocol giving the rationale and primary aim of the study, the primary response variable, a definition of who is to be studied, an estimate of subjects required, the nature of the control group, the mechanics of allocation to experimental treatments, the ways for maintaining blindness or minimizing bias, the procedures for evaluating and monitoring potential adverse effects, a timetable of investigation and the methods for collecting, analyzing and interpreting data.

The rationale states the scientific and clinical justification for the study. The primary response variable defines major outcome that is to be measured and analyzed. The primary response variable may be a subjective measure such as the extent a specific sign or symptom of illness is influenced by treatment. Alternatively, the primary response variable takes the form of a more objective measure

such as a clearly defined event (e.g., heart attack, death) or the measurable change of a relevant biologic index (e.g., tumor size). Although a clinical trial may address several questions, it is designed to answer one major question.

The study population defines eligibility for participation of research subjects. Eligibility criteria set forth characteristics required for inclusion in the study. For example, a study of a therapy in Parkinson's disease (PD) may need to focus on patients of a certain age and particular stage of illness who are receiving levodopa therapy. Other criteria define conditions or features which are intended to exclude certain subjects such as those with serious medical illness, intellectual decline (dementia) or active potential for childbearing. These inclusion and exclusion criteria are not meant as prejudices against certain persons but rather to clearly define who is to be studied with respect to the primary aim and major response variable. The estimate of how many subjects to study or sample size is usually derived from statistical assessment of the size of the effect expected from the intervention and the likelihood of detecting it if indeed it occurs.

Effects expected to be robust and uniform can be detected with a smaller population of subjects than subtle and variable effects. In drug trials for ET patients, IETF Medical Advisor Dr. Leslie J. Findley found that a minimum of 15 patients is required so that day-to-day variability in patients' tremors will not prejudice the results of the trial.

Definition of the control group is fundamental to the design of clinical trials. The control group provides a formal comparison. If an intervention is remarkably prompt and dramatic, as levodopa therapy was in treating signs of PD, then a control group may not be as critically required. On the other hand, a control group is essential for adequate comparison when the effect is expected to be less or to develop slowly over months or years.

Clinical trials may be conducted by administering a well-accepted standard drug to the control group for comparison with the drug to be tested in the experimental group. In many instances, there is no standard drug and it is necessary to administer placebos to the control group.

Placebos are substances that contain no active ingredients, but are prepared to appear and taste like the active substance. Placebos are often referred to as "sugar pills" because most contain lactose sugar. Lactose is used commonly as filler in the preparation of tablets and capsules. Saline or salt water is another placebo that is used as the inert ingredient in testing new substances that must be given by vein or injection such as Botox®.

The use of placebos in clinical trials prompts concerns regarding ethics in administering inert substances for comparison with an active compound. Where there is no accepted standard drug to administer, it can be argued that it is unethical not to give placebos to the control group. Without placebos, one might erroneously conclude the new substance is better than nothing when in fact it is no different than nothing or may even be worse than nothing. Placebos serve as a critical point of reference in assessing the value of an experimental therapy.

The potential research subject must be informed of the protocol for the trial, including the possibility that he or she might receive a placebo rather than the study drug, in an informed consent statement. This is a written statement of the purposes, procedures, potential risks and possible benefits of the study. A potential subject must review the informed consent statement with the investigator and indicate his or her understanding and willingness to participate in the research

study. This process of informed consent must be completed prior to entry into a clinical trial.

The informed consent statement is not a contract that binds the subject to participate, but a method of insuring that essential aspects of the study are well-known and understood in advance. Subjects may end their participation in the study for whatever reason without risking prejudice to their ongoing clinical care. Informed consent statements are reviewed and approved by IRBs before clinical trials begin.

Assignment of experimental and control treatments is carried out by a formal process of randomization so that subjects are equally likely to be assigned to the experimental or to the control group. Randomization removes the potential bias in assigning treatments and helps ensure that the experimental and control groups are reasonably comparable or matched at the beginning of the trial. Randomization also helps guarantee the validity of the statistical analyses assessing the final results of the trial.

The randomized trial is usually a double-blind study in which neither subject nor investigator knows of treatment assignments. Since both subjects and investigators have vested interests in the successful outcome of the trial, the double-blind design minimizes the possibility that value will be ascribed erroneously to the experimental treatment.

Treatment assignment codes are maintained by persons not involved in the evaluation of subjects or of the major response variable. In the event of a medical emergency, treatment assignment can be disclosed for the patient and appropriate action taken.

The safety monitoring committee, composed of individuals not involved in the evaluation of subjects, monitors the benefits and adverse effects of the experimental treatment with respect to the control. If this committee finds inordinate benefit or risk, it may recommend early termination of the trial.

It is normal for subjects and investigators to develop their own notions regarding which treatments have been assigned to individuals. It is not uncommon for subjects receiving placebos to develop “side effects” which they attribute to treatment. The beneficial and adverse effects reported by subjects taking placebos are usually real, and they help to place in perspective those effects reported by subjects who are taking the experimental treatment.

The recruitment of subjects for a large clinical trial is no trivial issue. Information publicizing such a trial must be conveyed accurately and widely. The cooperation of referral sources including family physicians, specialists and lay organizations is essential in order to achieve recruitment goals. Patients may not be eligible because of designed inclusion and exclusion criteria. Some eligible patients may have a change of mind or relocate and decide not to participate, or may elect to withdraw after enrollment.

We hope that the publication of this article helps you make an informed decision about participating in a clinical trial.

Localization of Genes for ET

By Daniel B. Mirel, Ph.D., and Kirk C. Wilhelmsen, M.D., Ph.D.

This report represents the progress of our research into the genetic components of ET, research that was funded in part by a grant from the IETF.

Two reasonably large North American families agreed to be the subjects of this study. The participation of such families is essential for research aimed at the identification of a gene or genes that predispose people to ET. Clinicians collected clinical information, family histories, and blood samples from as many family members as were available for study. Each of the two families has enough members who are affected with ET, and the members are closely related enough to each other to permit the obtaining of statistically significant evidence that ET is a genetic trait in these families (and probably in others as well).

The pattern of inheritance of ET in families is believed to be autosomal dominant. Autosomal means that both mothers and fathers can transmit to both sons and daughters a form (an allele) of the gene that causes disease. Dominant means that only one parent needs to pass this allele on to a child to cause disease in that child. Genetically, ET also seems to be partially penetrant, in that one may possess the ET gene causing allele but not have the symptoms of ET at all, or not until late in life.

To find a gene for ET, it is necessary to determine what chromosome segment(s) of all (or most) of the current ET sufferers in a family are identical and were inherited ultimately from the same common ancestor. Although the members of any one large family are descended from one mother and one father, presumably only the members with ET (and unaffected “carriers” of the gene form) share one particular chromosome segment in common that can be traced back to one of those two “founders.” A possibility is that one studied ET family is distantly related to the other, or to other families in which ET is found. In that case, current individuals with ET would have inherited the allele from one very distant ancestral founder.

DNA was extracted from the white blood cells sampled from members of these two families. Using molecular genetic techniques, the DNA was analyzed to allow the differentiation of naturally occurring inherited variations between individuals, which enable the chromosomes to be “labeled” and distinguished.

Using the information about the family relationships, a reconstruction of the probable passage of maternal and paternal chromosomes from generation to generation was deduced. Finally, using the knowledge of the family members who display clinical symptoms of ET, a single, relatively small section of one chromosome was identified as likely bearing the allele of the gene that is associated with ET. The location of this “shared inherited segment” (known as a haplotype) was the same for both families. The haplotype itself was not the same, however, implying that there was no common founder of the ET allele for the two families.

This finding of a location of an ET-predisposing gene is quite promising, but much work remains to be done. Although this chromosomal region contains many hundreds of genes of different known and yet-unknown functions, it also likely contains a gene with one or more alleles that predispose people to ET. There are several ways to narrow down the search for the causative gene. One

method involves combining the data from these two families with those from other families who also have an ET haplotype at the same place on the same chromosome.

A second method involves recruiting and studying more distantly related members of the families from this study, some of whom also have ET (if these people exist).

A third method entails looking specifically at genes residing in the chromosomal region whose known functions are consistent with the etiology of ET, and searching for alleles of those genes that could be causing the disease. All of these methods require more time and more study, and further participation of families like the two described here.

At this point, there is still no test that can identify people with a genetic risk for ET.

Further research will probably reveal that alleles of many different genes can be associated with ET in different individuals and families. The identification and characterization of these genes, and the understanding of how certain alleles predispose one to ET, will hopefully provide new insights into the etiology of tremor and the development of new therapeutic strategies.

Redefining ET into Three Sub-Categories

Reviewed and edited by Rodger Elble, M.D., Ph.D.

Researchers are divided as to whether ET is caused by a loss of nerve cells in the brain (neurodegeneration), say Drs. Günther Deuschl and Rodger Elble in a recently published paper in the journal *Movement Disorders* (Vol. 24, 2009, pp. 2033–2041).

There is a growing belief that genetics combine with environmental factors to produce the gradual destruction of certain parts of the nervous system that in turn causes ET. This possibility places ET within the same neurodegenerative category as Parkinson's disease and Alzheimer disease and are, for the most part, irreversible.

The second possibility, say the authors, is that genetics and possibly environmental factors cause networks of nerve cells (neurons) in the nervous system to oscillate abnormally (like a vibration in a car), that if left unchecked, could damage certain neurons leading to interference with the normal function of nerve networks resulting in symptoms of ET: tremor, clumsiness and possibly other problems. These problems, caused by oscillation, are reversible if the oscillation is stopped with drugs or surgery (e.g., deep brain stimulation (DBS)). Furthermore, oscillation-induced nerve damage is theoretically preventable if the tremor is treated early and effectively.

ET is a cerebellar disease

Classic ET, according to Drs. Deuschl and Elble “is a common action tremor that occurs when patients voluntarily attempt to maintain a steady posture or move. ET affects the upper limbs in most cases and less commonly affects the head, voice, face/jaw, tongue, trunk and lower limbs.” Neurologic examination in patients with classic ET reveals no other abnormalities except an unsteady tandem gait in some patients. Tandem gait is walking a straight line, heel to toe, as in the state trooper test for sobriety. It is a very sensitive test for disturbances of balance.

Patients with advanced ET usually have a disabling tremor that occurs during movement toward a target destination, such as when reaching for an object or drinking from a cup. There is little or no tremor at the beginning of the movement, but there is a dramatic increase in tremor as the hand approaches the target. This tremor is indistinguishable from the so-called intention tremor that occurs in cerebellar disease. Cerebellar disease produces intention tremor, unsteady gait, and uncoordinated movement of the limbs, and these problems are notoriously resistant to available therapies.

By contrast, the intention tremor and unsteady tandem gait are completely reversible in many ET patients, suggesting that ET is not a neurodegenerative disease. The consumption of alcohol temporarily suppresses intention tremor and reverses the unsteadiness during tandem walking, which is not what one would expect if ET were caused by cerebellar degeneration. The intention tremor and unsteady tandem gait also are reversed by DBS, which is believed to work by disrupting the abnormal oscillation within nerve networks. Therefore, Deuschl and Elble propose that while some neuronal degeneration may occur in ET, abnormal oscillation, not neurodegeneration, is the primary problem.

Redefining ET as a syndrome with subtypes

Senile ET

Dr. Elan Louis and colleagues have found a small but statistically significant association between ET and two common neurodegenerative diseases: Parkinson's disease and dementia of Alzheimer type. However, the association with dementia was found only in patients who developed ET after age 65, and the association with Parkinson disease was found only in ET patients age 65 and older. Furthermore, the published autopsies showing signs of neurodegeneration were all performed on ET patients older than 70.

Deuschl and Elble argue that the neuronal oscillation of ET could simply worsen the symptoms of any already existing neurodegenerative disease, increasing the likelihood of a clinically recognizable problem. On the other hand, a neurodegenerative disease could make ET worse or could simply produce tremors that resemble ET. Indeed, the peculiar association with neurodegenerative diseases in older persons suggests that many older persons have tremor resembling ET but may not have true ET. Therefore, Deuschl and Elble recommend that patients with classic ET beginning after age 65 be classified as having senile ET.

Hereditary ET

It is generally agreed that genetics play a major role in the development of ET. However, many large families and hundreds of other patients have been studied, and the only gene found to date is the LINGO1 gene. It is likely that ET is caused by a number of gene mutations and/or risk factor genes. It is unclear whether these genes cause the same condition or multiple conditions with similar symptoms. Regardless, studies of large families and twins have shown that familial ET almost always makes its appearance before age 65. Deuschl and Elble recommend that patients with ET, beginning before age 65, be classified as having hereditary ET if there is at least one other immediate family member (parent, sibling, or child, if spouse is unaffected) with classic ET beginning before age 65.

Sporadic ET

People within this classification have classic ET beginning before age 65, but do not have an immediate family member with ET.

Conclusions

A syndrome is the association of several clinically recognizable features that are produced by more than one disease. According to Deuschl and Elble, genetic data, signs and symptoms, neuroimaging findings, and results from post-mortem (after-death) examination of brain tissue indicate that classic ET is a syndrome, not a single disease.

Deuschl and Elble believe that the three proposed subtypes of classic ET (Hereditary, Sporadic and Senile) will facilitate the study of better-defined populations of persons with the ET syndrome and enable more accurate interpretation of research findings.

In particular, these sub-classifications will allow investigators to better control for the effects of heredity, environmental exposures, and the diseases of aging. Well-defined study groups of ET patients should improve the odds of finding the genes for ET, identifying non-genetic causes of ET, and deciphering age-associated disease that may cause or influence ET. Such study groups should also enhance the ability of researchers to find effective medications.

Finally, the neurodegeneration and oscillation hypotheses are not mutually exclusive. In the neurodegeneration hypothesis, the primary problem is nerve cell destruction, as in Parkinson disease

and Alzheimer disease. Ultimately, the destructive neurodegenerative process leads to symptoms of ET. In the oscillation hypothesis, symptoms are produced primarily by the abnormal behavior (oscillation) of nerve cells. In other words, the initial problem is a functional disturbance of nerve networks, not a destructive neurodegenerative process. However, the oscillation, if left unchecked, could ultimately produce nerve cell destruction, and this, in turn, could worsen the symptoms of ET and could enhance the effects of other neurologic illnesses, which are increasingly likely to occur as we grow older. Both hypotheses illustrate the importance of achieving a better understanding of the causes of ET.

Stem Cell Research Offers Promises, Few Realities for ET Research

By Peter Heder, M.D.

Stem cell research has generated a lot of controversy and excitement among both experts and the general public. Stem cells are non-differentiated cells that can theoretically develop into various specialized cell types such as nerve, liver, kidney, etc. The basic idea behind the use of these cells, which may be derived from embryonic or adult tissues, is that they will regenerate impaired or dead cells in damaged organs.

The most promising areas of stem cell use in neurology are for those disorders where certain types of nerve cells are selectively damaged and die. Examples of this include Parkinson's disease in which dopamine producing neurons die, and amyotrophic lateral sclerosis (ALS), also called Lou Gehrig's disease, in which spinal cord nerve cells controlling skeletal muscles are selectively lost.

Before we address the question of a potential role of stem cells for treatment of essential tremor, we need to emphasize that even though the potential use of stem cells is scientifically sound, it could take years before we see any practical use. Furthermore, given the complexity of the central nervous system, it is also possible that this exciting idea will simply never work in widespread clinical use. Undoubtedly, the knowledge derived from stem cell research, however, will help us better understand how the brain develops and how it may react to various injuries.

Is there any promise for therapeutic use in ET?

At this point, we cannot answer this question because our understanding of the causes of ET is very limited. First, we need to make more strides in our efforts to find a cause (or more likely multiple causes) of ET. We are not even certain about the degree of nerve cell loss in the brains of patients with ET and cannot definitely point to the types of nerve cells that cause ET. Before we can plan the strategy of stem cell research in ET, we need to have a more detailed understanding of these questions. The ET community can play a very important role in this quest because new clues into ET development will likely come through genetic studies and studies of brain tissue of patients with ET. Both kinds of studies are impossible without voluntary participation of patients affected by ET.

Scientists Identify Protein Linked to Essential Tremor

Tremor Talk

In early March 2014, a team of researchers from Université Laval and CHU de Québec, in Québec City, Canada identified unusually high levels of a certain protein in the brains of people suffering from essential tremor. The discovery, the details of which were published in the journal *Movement Disorders*, could lead to an effective treatment for essential tremor.

“Even though it’s not a lethal degenerative disease, essential tremor still poses a serious problem to sufferers, making it extremely difficult to perform basic everyday activities,” explains lead researcher Frédéric Calon, a professor at Université Laval’s Faculty of Pharmacy and affiliated with CHU de Québec Research Center.

Dr. Calon and his colleagues had access to a brain bank developed more than 40 years ago by University of Saskatchewan professor Ali Rajput to test their hypothesis that the brains of ET sufferers show an overabundance of certain proteins. The researchers focused on two proteins in particular—LINGO1 and LINGO2—which, according to some genetic studies, may be linked to the movement disorder. They measured the concentrations of these proteins in the cerebellums of nine subjects with essential tremor, 10 Parkinson’s subjects, and 16 healthy subjects.

Their analyses revealed that there was a higher concentration of LINGO1 in the cerebellar cortex of people suffering from essential tremor; twice that of healthy subjects. This increase was even more pronounced in people who had been living with the condition for more than 20 years. These differences were not observed in the subjects with Parkinson’s disease.

“Other studies have shown that LINGO1 slows neuroregeneration following damage to the brain or spinal cord,” points out Dr. Calon, “so we believe that inhibiting this protein could be a promising treatment avenue to explore for essential tremor. The drugs currently prescribed to people suffering from this neurological condition were developed 30 years ago and their effectiveness is limited.”

“This is very exciting news,” says IETF executive director Catherine Rice. “The largest obstacle to getting better treatment options is our lack of understanding of what causes essential tremor. This research is starting to unravel the mysteries and will hopefully lead to better more tailored treatments and medications in the future.”

Initial funding for Dr. Calon’s work was provided by a research grant from the IETF. In addition to Dr. Calon, this study’s coauthors are Charlotte Delay, Cyntia Tremblay, Élodie Brochu, Sarah Paris-Robidas, Vincent Émond et, Ali Rajput, and Alex Rajput.

The IETF will continue to follow Dr. Calon and his research team as they continue this important, groundbreaking work.

ET and Alcohol Sensitivity: Clinical Test Examines Alcohol Sensitivity of Essential Tremor

Tremor Talk

Researchers Karina Knudsen, M.D., Delia Lorenz, M.D., and Gunther Deuschl, M.D., Ph.D., Department of Neurology, Christian-Albrechts-University, Kiel, Germany, have conducted a study to develop a simple diagnostic test for alcohol sensitivity of essential tremor patients. Results were published in *Movement Disorder Society*.

The idea of this study is to develop a standardized test for the responsiveness of an individual patient to alcohol. The tremor-suppressing potential of alcohol on ET was first reported in 1975 and later confirmed by several studies. Meanwhile, alcohol sensitivity is part of the secondary criteria for the diagnosis of ET. Alcohol specifically reduces ET severity at low blood levels, with a reduction of tremor amplitude of 50–70%.

As a first step, the alcohol effect needed to be quantified. Both the time course and the amount of tremor suppression were systematically studied. Ten patients were tested for alcohol sensitivity under controlled conditions in the laboratory, and 15 patients were instructed to perform an alcohol test at home following an adapted dosage of alcohol.

For the control subjects, the dosage of alcohol was adapted for each individual according to weight and sex, to receive a target response of approximately 0.8% blood alcohol. Tremor testing using multiple ratings was conducted prior to and after consuming alcohol.

The time course of the anti-tremor effect showed significant improvement of up to 50% in both groups for all the outcome parameters. Tremor deteriorated after three hours. A quarter of the patients noticed the alcohol effect for the first time during the test.

The alcohol response of patients with ET has never been studied under controlled laboratory conditions. The study confirms the well-known qualitative effect of alcohol on tremor in ET patients with a quantifying approach and proposes a test for quantification of the alcohol response. The strong effect of alcohol within the first 90 minutes after alcohol ingestion is followed by a severe rebound effect. Its effect is only short-lived and exhibits a rebound after more than three hours and the next morning.

The response of the patients was highly consistent. The researchers conclude that the test qualifies for home testing and can be used as a screening tool for confirmation of the diagnosis.

Alcohol responsiveness may be a key physiological feature of ET. So far, it is unknown if the alcohol test can also be used as a differential diagnostic tool. About one in three patients with tremor have been found to be misdiagnosed as having ET, with the most frequent false diagnoses being Parkinson's disease and dystonia. Certainly more research is necessary to accept alcohol-sensitivity as a reliable differential diagnostic tool for ET against all other tremor entities.

ET, Exercise and Longevity

By Joseph Jankovic, M.D.

Any discussion of the management of a chronic disorder would not be complete without emphasizing the importance of physical activity. Exercise has been shown to lower blood pressure, reduce obesity, and prevent cardiovascular disease, but there is little data on the role of exercise in essential tremor (ET).

In fact, many patients with ET observe that their tremor markedly increases after strenuous physical activity or exercise, and they understandably wonder if they should avoid such activity.

As long as there is no cardiac, orthopedic or other condition that creates limitations, patients with ET should remain physically active. Be reassured that worsening of tremor after exercise is expected due to the increase of adrenaline (or epinephrine and norepinephrine) during exercise. Adrenaline, released during any physical or psychological stress, leads to increased muscle activity manifested as worsening of tremor. This effect, however, is temporary and the tremor usually returns to its previous state after a few minutes of rest.

Alcohol and propranolol, a beta adrenergic blocker, can reduce stress-induced worsening of ET symptoms; hence the two drugs are often used to “calm” the nervous system. Even professional actors often use propranolol to minimize the tremor effects of anxiety associated with stage fright.

Many studies have shown that exercise benefits not only the body, but also the brain. It has been shown to improve learning, memory, and depression. It also appears to protect the brain from neurodegeneration [Cotman et al, 2007].

Although Parkinson’s disease is different from ET (despite the occasional overlap of the two disorders), studies on exercise in Parkinson’s disease may be relevant to patients with ET. In a study of 48,574 men and 77,254 women, higher levels of physical activity were associated with lower risk of Parkinson’s disease [Chen et al, 2005]. Based on systematic literature review, the Practice Recommendations Development Group from The Netherlands concluded there is sufficient evidence to recommend physical therapy and exercise to improve balance, joint mobility, and muscle power and to improve physical capacity of patients with Parkinson disease [Keus et al, 2007].

There are many other studies that provide evidence that exercise may be helpful in improving motor function [Kwakkel et al, 2007], although firm evidence that exercise lowers the risk of Parkinson’s disease is still lacking [Logroscino et al, 2006].

Exercise may prolong life not only by preventing or reducing the risk of life-threatening disorders, but by slowing the aging process. One of the most compelling arguments in favor of exercise as an important anti-aging factor is the recent finding about leukocyte telomere length. The length of telomeres, caps on the end of chromosomes in human DNA, is a biological indicator of human aging. Longer telomere length correlates with exercise, as well as increased lifespan.

In a study of 2,401 twin volunteers, comprising 2,152 women and 249 men, who were asked to complete detailed questionnaires about their level of physical activity, smoking status, and socioeconomic status, the leukocyte telomere length was 200 nucleotides longer in the most active

subjects as compared to the least active subjects during their leisure time ($P<.001$), even when adjusted for age, sex, body mass index, smoking, socioeconomic status, and physical activity at work.

Long-held beliefs reflect that ET patients live longer than people without it. Russian neurologist Minor suggested in 1935 “that a factor for longevity was also contained in the tremor gamete.” In a study published in 1995, it was found that parents of ET patients who experienced tremor lived on the average 9.2 years longer than those parents who did not have tremor. Because the parents with tremor who lived longer probably had ET, it was concluded that ET confers some anti-aging influence and significantly increases longevity.

While there is no obvious explanation for this striking observation, it is possible that patients with ET have an underlying personality trait that encourages dietary, occupational, and physical habits that promote longevity. The small amounts of alcohol consumed to calm tremor might prolong life, or tremor itself might be viewed as a form of exercise that would have beneficial effects on general health and on longevity. Further studies are needed on the potential anti-aging effects of ET and whether exercise confers additional benefits by favorably modifying the course of the disease.

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More opinions on exercise and ET

Rodger Elble, M.D., Southern Illinois University School of Medicine

Bilodeau and coworkers (*Muscle Nerve* 2000; 23: 771-778) performed a small pilot study of the effect of exercise on ET and found that strength training reduced tremor, at least temporarily. Light resistance exercise did not help. The long term effects of exercise are not known. Some patients experience a transient increase in tremor during fatigue after exercise. The long-term effects of heavy resistance training should be studied further.

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To date, limited research has been completed in the role of exercise and individuals with ET. A small study in individuals with ET has demonstrated the benefit of strength training with improvement of hand steadiness.

The Driving Need for Essential Tremor Research

By Ludy Shih, M.D.

Essential tremor is a common but misunderstood disorder affecting the lives of millions of Americans. As a movement disorders neurologist, I see this firsthand in the patients that come to clinic looking for help in treating their tremor. Patients are affected in different ways and respond to treatments in varying degrees. Overall, tremor and tremor-related disability tend to get worse with time. As a clinician investigator, I see two things driving the need for essential tremor research: A) the need for more effective and better tolerated medications for tremor, and B) the ways that the common clinical features of ET should lend us clues to the understanding of tremor.

To illuminate on point A:

Tremor medications that are currently used are a byproduct of research and development on other neurological conditions, like epilepsy. Because little is known about how tremor works in humans and how it might be manipulated successfully, pharmaceutical companies have little incentive to develop a robust product development program for tremor.

To expand on point B:

ET cases are familial; giving us hope that genetic mutations can be identified that will reveal basic knowledge about how tremor works in humans, similar to what the discovery of genes causing Parkinson's disease has done for that field. If we are fortunate, genetic mutations may reveal a chemical target that can be manipulated with the right drug, or they may reveal a more complex mechanism rooted in nervous system development or even neurodegeneration although this latter point is quite controversial.

Our research at Beth Israel Deaconess Medical Center is focused on two areas. One goal is to try to identify patients who have subtle structural brain changes that may be associated with a more rapid progression of tremor related disability. Also, we are trying to identify whether there is any association with genetic variations in a gene called LINGO1 that is not seriously defective in people with ET but may contain slight variations that help contribute to the development of essential tremor in a patient.

The second goal is to work collaboratively with other centers through the North American Essential Tremor Consortium to try to build a robust, data-rich repository on a large group of patients in order to classify and identify biological markers of tremor that might help enrich our genetic studies. Genetic techniques have advanced significantly over the years, but there are many forms of tremor. Each may have their own mechanism and methods of treatment. Therefore, careful clinical characterization may still be necessary in order to make the proverbial "finding of a needle in a haystack" more productive.

Patients can help by volunteering to be a part of these types of studies and by raising awareness in their community about the need for essential tremor research. Like any difficult endeavor, persistence, teamwork, collaboration and organization among patients, their advocates, physicians, and scientists are often the key to success. This way we can be as prepared as possible for that next critical scientific and clinical breakthrough. Continued support for the IETF—from people like you—is crucial to sustaining our research efforts.

Resources

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