Essential Tremor Patient Handbook

Your reference guide for the most common movement disorder.

What is essential tremor?

Essential tremor (ET) is one of the most common neurological conditions and the most common cause of tremor. Tremor is an involuntary, rhythmic shaking of any part of the body. The hands are most commonly affected in ET, but the head, voice, legs, and trunk can also be affected.

The term essential, when used in a medical context, refers to a symptom that is isolated and does not have a specific underlying cause. Thus, ET refers to a disorder that displays the primary symptom of tremor, with no known cause.

The tremor of ET is an action tremor and most commonly occurs while performing activities such as eating, drinking, writing, typing, brushing teeth, shaving, etc. (kinetic tremor) or when the hands are in an outstretched position (postural tremor). ET can therefore make it difficult to complete everyday tasks and can lead to significant disability.

Some patients may present with a combination of tremors affecting different body parts. The severity of the tremor can vary from a barely noticeable tremor only present in situations of stress or anxiety, to severe tremor that has a significant impact on activities of daily living. Tremor severity can vary based on the activity being performed, the position of the body part, and the presence of stress or fatigue. The tremor may worsen over time and may spread to parts of the body not previously affected.

Who develops ET?

ET is estimated to affect up to 10 million people in the United States and many more worldwide. Men and women are equally affected and it occurs in people of all races. Although ET is most com-

Although commonly seen in older adults, essential tremor can occur at any age. Ryan Brown was diagnosed at four years old.

monly seen in older adults, it may occur at any age from childhood to late adulthood. ET is hereditary in more than 50 percent of cases. It appears to be an autosomal dominant disease, which means that a child of a person with ET has a 50 percent chance of inheriting the disorder. Consequently, ET has been referred to as familial tremor. ET has also been called benign tremor, which is misleading, as ET is commonly a source of frustration or social embarrassment, and in some cases can be disabling.

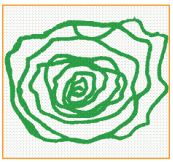
What causes ET?

The cause of ET is currently unknown. It is thought to be related to abnormal functioning of the part of the brain called the cerebellum or a chemical in the brain called GABA; however, no specific brain abnormalities have been confirmed. Although a large percentage of persons with ET have a family history of the disorder, a specific gene for ET has not been identified and therefore no genetic test for ET is currently available. In fact, not everyone who has a family member with ET develops ET. This suggests the possible influence of some not yet identified environmental factors.

How is ET diagnosed?

To avoid a misdiagnosis, it is important that you see a doctor who has been trained to diagnose and treat tremor. It is best to see a neurologist who specializes in movement disorders. There are no medical tests to confirm the diagnosis of ET. ET is diagnosed by medical history and neurological evaluation. The neurological exam generally includes an examination of tremor in various body parts in different positions as well as completion of writing samples, pouring, and drawing Archimedes spirals, after which the physician evaluates symptoms and rules out other potential causes of tremor such as:

- medication side effects,
- thyroid disease,
- excessive stress or trauma,
- excessive use of caffeine or other stimulants,
- presence of other movement disorders such as Parkinson's disease (PD) and dystonia.



Archimedies spiral drawn by a patient receiving no medication.

Archimedies spiral drawn by a patient receiving a trial dose of a medication.

Archimedies spiral drawn by a patient receiving a more effective dose of the same medication.

ET is diagnosed by medical history and neurological evaluation.

How do I prepare for an ET neurological evaluation?

During the neurological evaluation, the doctor will ask you for complete medical and family history and specific information about your tremor. Here are some questions the doctor may ask during your visit:

- Do you have a family history of tremor?
- Does anyone in your family shake for any reason?
- When did you first notice any type of shaking (tremor) and did it begin after a particular incident?
- What parts of your body are affected?
- Did the tremor start suddenly?
- Has it worsened over time?
- Are there specific factors that make your tremor worse such as stress, caffeine, or alcohol intake?
- What effect does alcohol have on your tremor within a few minutes after ingestion, and the following morning?
- Have you ever had a head injury?
- Does your tremor occur during action or while you are at rest?

- What medications—over-the-counter vitamins, herbal supplements, and prescriptions—are you taking and what medications have you taken in the past? (Certain medications may cause tremor, so it's a good idea to bring a list of the medications you are taking or the pill containers themselves. It is also helpful to list any medications you have taken in the past.)

What are other causes of tremor?

Although ET is the most common cause of tremor, there are many other potential causes. Some additional causes of tremor include:

- **Enhanced physiologic tremor** – Everyone has a mild, barely detectable tremor referred to as a physiologic tremor. During situations causing stress, fear, anger, anxiety, or fatigue this tremor may become noticeable and is referred to as enhanced physiologic tremor.
- **Drug-induced tremor** – Tremor can be a side effect of prescription medications, such as beta-adrenoceptor agonists, valproic acid, thyroxine, tricyclic antidepressants, selective serotonin reuptake inhibitors, amiodarone, procainamide, methylxanthines, antipsychotics, and lithium among others. Some over-the-counter medications, supplements, herbal remedies, and illegal drugs may also cause tremor.
- **Post-traumatic tremor** – An injury to the head can cause tremor.

- **Other neurological disorders** – Other than ET, the most common neurological disorders causing tremor are Parkinson's disease and dystonia (involuntary muscle contractions), but tremor may also occur with multiple sclerosis or after a stroke.

Do I have ET or Parkinson's disease?

It is often difficult to differentiate ET from Parkinson's disease (PD), particularly during the early stages of the disease. Consequently, many people are initially misdiagnosed. And to complicate the situation, it is possible to have more than one neurological disorder. Here are some basic differences between ET and PD:

- **Type of tremor** – The tremor in ET generally occurs with action, either while performing a task (kinetic tremor) or while holding the body part against gravity such as having the arms outstretched in front of the body (postural tremor). The tremor of PD most commonly occurs when the limb or other body part is at rest (resting tremor).

Dr. Vincent DePinto retired from vascular surgery because his tremor interfered with his ability to preform delicate surgeries.

- **Additional symptoms** – The primary symptom of ET is tremor. Although tremor is a primary symptom of PD, additional features, such as slowness, stiffness, balance problems, and shuffling gait are also generally present.
- **Body parts affected by tremor** – ET and PD both involve tremor of the arms/hands, but head and voice tremor most commonly appear in ET and almost never in PD.
- **Handwriting** – In ET handwriting is often large and shaky, whereas in PD, handwriting is often very small (micrographia).
- **Family history** – Persons with ET often have a family history of tremor, whereas in PD, family history of PD or tremor is much less common.
- **Alcohol** – ET is often reduced by drinking alcohol whereas in PD, alcohol generally does not improve tremor.
- **Medications** – ET generally does not respond to medications used for PD, and PD generally does not respond to medications used for ET.

What is DaTscan® and how can it help me?

DaTscan® is a neuroimaging tool (brain scan) that can assist in differentiating ET and PD, but it does not provide a definitive diagnosis for either disease. In cases where neurological examination is not sufficient to make a diagnosis of either ET or PD, DaTscan may be helpful.

PD is related to a decrease in a chemical in the brain called dopamine. DaTscan® involves a picture of the brain using single photon emission computed tomography (SPECT), to measure dopamine transporter levels in the brain. In PD, an abnormal scan representing a reduction in dopamine transporter levels is seen, whereas in ET the scan is normal.

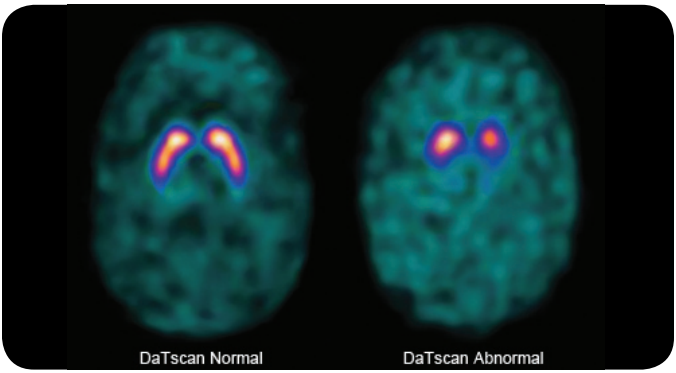


Figure 1

Figure 1 compares the normal comma-shaped appearance on both sides of the brain of the DaTscan in ET (left) with the period-shaped appearance of the DaTscan in PD (right) due to reduced dopamine transporter levels. It is important to understand that a normal DaTscan does not confirm a diagnosis of ET; it only helps to rule out a diagnosis of PD.

What therapies are available for ET?

While a cure for ET has not yet been found, medications may help improve your quality of life. If your ET is mild and not bothersome to you, you may not need treatment. However, if ET interferes with your ability to work, perform daily activities, or interact socially, you may want to consider available therapies. The goal of treatment is to reduce

the severity of tremor to improve daily functioning such as eating, drinking, writing, and typing, as well as to reduce social embarrassment.

Medications are estimated to be effective for about 50 percent of patients and they rarely completely eliminate tremor; therefore, it is important for you to have realistic expectations about drug therapy. Each person responds differently to the various treatment options, so it may take time to find the treatment that is best for you. You may need to try more than one treatment or use a combination of treatments before you and your physician find what works best for you.

First-line therapies

Propranolol (Inderal®, Inderal-LA®)

Propranolol is currently the only medication approved by the Food and Drug Administration (FDA) for the treatment of ET. Propranolol is in a class of drugs called beta-blockers, which are used primarily for treating high blood pressure. These drugs block adrenaline in the nervous system; however, it is not clear exactly how propranolol works in treating ET. Propranolol is effective in reducing tremor in approximately 50 to 60 percent of persons with ET.

Your doctor may prescribe propranolol to be taken as needed, such as during particularly stressful situations, or it may be taken daily if your disability is persistent. Tremor reduction generally occurs one to two hours after a single 10 to 40 milligram (mg) dose, and the effect generally lasts about four hours. A once-daily, long-acting preparation is also available. Although propranolol is most effective for hand tremor, it may also be effective for head and voice tremor. Individual response varies, and complete tremor reduction is rare. Side effects of

Natalie Kusilek, IETF member since 1999, with her daughter Teri. Natalie has struggled to find a treatment option that helps control her head tremor.

propranolol are usually mild and are more frequent at higher doses (more than 120 mg/day). The main side effects are decreased pulse rate and low blood pressure. Less common side effects are fatigue, depression, impotence, nausea, weight gain, rash, and diarrhea. If you experience unpleasant side effects, be sure to talk with your doctor. If you have heart failure, diabetes mellitus, or asthma, you should talk with your general medical doctor before you take propranolol.

Propranolol is typically started in divided doses from 10 to 60 mg/day. While most patients benefit from doses less than 120 mg/day, the daily dose can be increased up to 320 mg if necessary. No additional benefit has been seen in doses greater than 320 mg/day. Older patients should start with 10 mg/day, and the dose should be slowly increased to 80 to100 mg/day. Propranolol long-acting should be started at 60 mg/day and can be slowly increased to 120 mg/day or higher as needed and tolerated. Do not abruptly stop this medication without first talking with your physician. Other beta-blockers such as atenolol, metoprolol, and nadolol may also be beneficial for ET.

Primidone (Mysoline®)

Primidone is an anti-seizure medicine that is often used to treat ET. Approximately 60 percent of people with ET are helped by primidone, and the benefit usually lasts 24 hours after each dose. When you first start taking primidone, you may experience nausea, poor balance, dizziness, fatigue, drowsiness, and flu-like symptoms that generally subside after a couple of days. You can reduce the possibility for these symptoms by starting with a small dose at bedtime and gradually increasing the dose until tremor is suppressed. If you experience more serious side effects, you should contact your doctor. Although primidone may have initial side effects, there are few long-term problems.

Primidone dosage typically starts at 12.5 mg (one quarter of a 50 mg tablet) or 25 mg (half a 50 mg tablet) at bedtime. After one week, the dose can be increased to 50 mg at bedtime. The dose can then be increased by 50 mg a week, typically up to a dose of 250 mg/day or until adequate tremor control is achieved. Doses of up to 750 mg/day provide benefit in some patients. Primidone can be taken as a single dose at bedtime, particularly if sleepiness is a problem, or it can be taken in divided doses throughout the day.

Combination of Propranolol and Primidone

If your tremor is not well controlled by propranolol or by primidone alone, you may experience better results when you take both medicines together.

Benzodiazepines

Clonazepam (Klonopin®), diazepam (Valium®), lorazepam (Ativan®), and alprazolam (Xanax®) are frequently used to treat ET. These drugs are especially helpful in patients with associated anxiety. Although diazepam has been shown to improve

Take your medications exactly as prescribed by your physician.

tremor, it is typically not as effective as propranolol and primidone. Alprazolam has been shown to significantly reduce tremor as well as symptoms of anxiety. The effectiveness of alprazolam has been found to be equal to that of primidone.

If benzodiazepines are used for long periods in large dosages, they can become addictive. There is also risk of withdrawal symptoms if the drugs are stopped suddenly. These drugs may be particularly useful in patients who do not respond to other medications or who have associated anxiety. Side effects include sleepiness, dizziness, depression, fatigue, loss of coordination, memory loss, and confusion.

Second-line therapies

Gabapentin (Neurontin®)

Gabapentin is an anticonvulsant. It has a modest benefit in ET and is often tried when propranolol and primidone are not effective. Gabapentin is generally well tolerated. Side effects include fatigue, slurred speech, drowsiness, impaired balance, and nausea, especially when beginning treatment. The drug has a short duration of action and requires multiple doses a day. In older persons, gabapentin is typically started at 100 mg three times daily; in younger persons it is often started at 300 mg three times daily.

Topiramate (Topamax®)

Topiramate is an anticonvulsant that has been shown to be effective in controlling tremor in some

persons with ET. Starting with a low initial dosage and slowly increasing the dosage over time can minimize side effects. Dosages should be started at 25 mg or 50 mg at night for the first week, and increased by 25 to 50 mg/day each week, depending on side effects, up to a total dose of 300 to 400 mg/day. Side effects include numbness or tingling (paresthesia), memory loss, and weight loss. Topiramate should be avoided in patients with a history of kidney stones and should be used with caution in the elderly due to an increased risk of memory problems and confusion.

Mirtazapine (Remeron®)

Mirtazapine is an antidepressant. Due to its lack of effectiveness for the majority of persons with ET 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

If medications fail, you may consider therapy that involves injecting botulinum toxin into the affected muscles. Botulinum toxin injections have been useful in the treatment of some patients with hand, head, and voice tremor. Receiving optimal benefit from botulinum toxin requires a specialist skilled in the technique. The effects last about three to four months and, for continued benefit, must be repeated about three times a year. Transient weakness of the injected muscles is the most common side effect. Be sure to check with your insurance provider about coverage, as this treatment can be expensive.

Surgical Therapies

If medical treatment is not successful and your tremor continues to cause disability, surgical alternatives such as deep brain stimulation (DBS) of the thalamus or thalamotomy might be an option. You can learn more about surgical treatments for ET in “Surgical Treatment of Essential Tremor,” which is available from the IETF office.

Elizabeth Guthrie, support group leader and artist, opted for DBS surgery when medications no longer controlled her tremor.

What is the effect of alcohol on ET?

Adults with ET often notice that drinking alcohol reduces tremor for one to two hours. When you use alcohol responsibly, it can be very effective in temporarily reducing tremor. It may even be helpful to have one or two drinks during social events to suppress tremor. There are, however, important issues to consider in using alcohol for ET. Rebound tremor may occur after excessive alcohol use, making tremor temporarily more severe the next day. If you use alcohol to reduce tremor, be sure to talk with your doctor about it. You should avoid excessive use of alcohol, and never consume alcohol if you plan to drive. The limitations of the use

of alcohol as a treatment for ET include the short duration of response, development of tolerance to the anti-tremor effect, increased tremor as alcohol is metabolized, and the risk of alcoholism.

What other tactics might help me manage my ET?

While the following do not have proven effectiveness for ET, some people with ET have found them to be beneficial:

- **Occupational therapy** – An occupational therapist can provide helpful techniques or introduce devices to make living with ET more manageable. Some examples include using heavier utensils, cups, and glasses; wrist weights; plate guards; heavier, wider writing instruments; and other adaptive devices such as a special computer mouse that compensates for tremor, or computer voice recognition programs that greatly reduce the need for typing.

Kate Larson is a horse trainer and riding instructor. For Kate, it isn't about how ET limits her; it's about what she can accomplish despite her tremor.

- **Alternative treatments** – Many people have tried treatments such as acupuncture, hypnosis, meditation, yoga, and massage therapy with unconfirmed benefit. These treatments may be particularly helpful in persons with anxiety. Biofeedback or behavioral therapy may also be helpful for persons dealing with high levels of stress or anxiety. There are currently no specific vitamins or supplements recommended to reduce tremor.

Practical suggestions

There are many ways to minimize the degree to which ET interferes with your life. Here are some practical suggestions:

- Learn as much as you can about ET. You are your own best advocate.
- ET is commonly misdiagnosed. Make sure your physician is knowledgeable about ET and is actively working with you to control your tremor. Our website provides a list of physicians with experience treating ET and other movement disorders.
- Don't hide your tremor. Talk about it to friends, relatives, and colleagues. Creating greater awareness about your tremor can greatly reduce your anxiety and may consequently reduce your tremor. Not only will you make yourself more comfortable around others, but they will be more comfortable around you! The more everyone learns about ET, the faster public awareness will increase. Greater awareness will bring attention to ET and facilitate research so additional treatments and a cure can be discovered.

- If your child has ET, you may want to meet with his/her teacher(s) to discuss. Order the free IETF booklet “Children with Essential Tremor: A guide for parents, teachers and other caring adults” and share it with your child’s teacher. “Tremor Disorders in Children: A Clinical Discussion” is an IETF booklet you may want to share with your physician.
- Find ways to reduce stress and learn some relaxation techniques. Anxiety and depression are common in ET.
- Avoid things that may worsen tremor such as caffeine, some herbal remedies, and certain prescription medications.
- You can find many more ideas by calling the IETF and asking for the flyer “Coping with Essential Tremor.”
- Join a support group or learn how to become a leader. The IETF provides information about joining or starting an ET support group. A list of groups is also available on the IETF website: www.essentialtremor.org.

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Resources

For more information about ET, call the IETF toll-free at 888-387-3667 or visit the website: www.essentialtremor.org

Join us on Facebook: https://www.facebook.com/InternationalEssentialTremorFoundation

Follow us on Twitter:
http://twitter.com/essentialtremor

Sign up for the monthly electronic newsletter, Tremor Gram, at www.essentialtremor.org

Participate in the forums of our online blog:
www.tremortalk.org

My Important Information

My doctor: _____

Phone: _____

My neurologist:_____

Phone: _____

My medication(s):_____

_____

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

International Essential Tremor Foundation (IETF)

The International Essential Tremor Foundation is a 501(c)(3) non-profit organization that funds research to find the cause of essential tremor 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.

International Essential Tremor Foundation

Your Voice for Essential Tremor

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