



Essential Tremor (ET)

Dystonic tremor

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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 than 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 to 85 percent 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 than 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 percent of the adductor-type, 50 percent of the abductor type and 40 percent of the mixed-type. Oral medications are not as useful but clonazepam is the most useful.

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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 (www.dystonia-foundation.org); National Spasmodic Torticollis Association (www.torticollis.org); and National Spasmodic Dysphonia Association (<http://www.dysphonia.org/>).



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.