# TREMOR IN DYSTONIA

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Dystonia has been classically defined as involuntary muscle contractions that cause sustained, twisted or abnormal postures (1). A 2013 Consensus Statement improved the previous dystonia classification system and now recommends defining dystonia under two axes: clinical features and etiology (2). The clinical features include age of onset, body distribution, temporal pattern, coexistence of other movement disorders, and other neurological manifestations. A review of this statement is strongly recommended.

A dystonic tremor is an action tremor that may be postural, kinetic, or both. Rarely, a dystonic tremor occurs at rest. The tremor is often asymmetric, jerky and irregular in amplitude and periodicity. At times, it is possible to identify the net direction in which the neck or a limb is twisting. When the limb or neck is positioned in this direction, the dystonic tremor can be diminished or abolished. This is known as the tremor's **"null point"**. Positioning the neck or limb in an opposing direction, in contrast, will cause the tremor amplitude to increase. Patients with dystonic tremor may also have sensory tricks or "**geste antagoniste**". These are maneuvers that can be voluntarily performed to minimize the dystonia, like touching one's face, head or hand.

Tremor is often found in patients with dystonia. A prospective study of adult-onset primary dystonia (defined by the previous classification system) found 262 of 473 patients (55.4%) were identified as tremulous. Arm tremor was seen in approximately 30% of patients and head tremor in 40% of patients. Almost one-third of patients had both (3). In a study of 427 subjects with primary adult-onset dystonia, those who had tremor often had it since the time of dystonia onset (4). Interestingly, dystonia had a greater tendency to spread in patients with tremor.
It is important to remember that tremor can occur in patients with dystonia in areas that are unaffected by dystonia. This is known as tremor associated with dystonia, and should be distinguished from dystonic tremor. This tremor tends to be postural and kinetic, and may resemble essential tremor. Similarly, essential tremor patients may have associated dystonia (5).

<u>Dystonic tremor can be misdiagnosed as PD tremor or ET tremor. A few pearls to help distinguish are as follows:</u>

1. Isolated neck tremor is most suggestive of dystonia, and early diagnosis with ET should be avoided (6). A detailed tremor examination of 583 essential tremor patients reported that none of them had head tremor in the absence of arm tremor (7).
2. Isolated voice tremor is most suggestive of laryngeal dystonia (known as spasmodic dysphonia). Patients with dystonic vocal tremors cannot change pitch during vocalization, and performing this maneuver can aide in the bedside diagnosis (8, 9). A sustained phonatory vowel task with fibreoptic nasolaryngoscopic examination of pharyngeal and laryngeal musculature, supplemented by acoustic measures can help to characterize and confirm the nature of the tremor (10).
3. In patients with neck tremor, the presence of neck pain accompanying the neck tremor is suggestive of dystonia (11, 12). About 80% of patients with cervical dystonia have associated neck pain (13). Tightness and cramping in a tremulous arm is also associated with dystonia.
4. In patients with neck tremor, the presence of hypertrophy in neck musculature, especially if there is asymmetry, is suggestive of dystonia. Muscle hypertrophy is present in nearly all patients with cervical dystonia (13).

**PEARLS TO EXAMINING DYSTONIA**

Ask the patient to vocally sustain “ahh” or “eee” for 7 seconds. Listen for breaks in the voice.

Ask the patient to write. It is best to ask the patient to write a sentence in script. When evaluating the writing task, make sure shirt sleeves are pulled up. Ideally feet are exposed as well. Ask the patient to write a sentence repetitively like, “Today is a sunny day in Southern California.” In an affected limb, look for tight grip of the pen, unusual posturing of the hand that may progress as the patient continues to write, dystonic mirroring/overflow in another limb, and involvement of more proximal muscles. Ask the patient if (s)he feels tightness in the limb. Look for involuntary toe flexion, extension or foot inversion that would suggest dystonic overflow.

Look for a “null point”, the position where the dystonic tremor is reduced. For example, a patient with a right torticollis manifesting as a tremor may have complete cessation of tremor when turned to the right, and exacerbation of tremor when turned to the left. Limbs may be passively positioned by the examiner in search for a “null point” as well. Common positions include extreme pronation, supination, flexion and extension of the wrists.

The approach to treating dystonic tremor varies depending on the location of the tremor and the age of the patient. Botulinum toxin is often used for focal tremors in the neck or limb. Trihexyphenidyl, baclofen, and clonazepam may be helpful in some cases. Tetrabenazine is used off-label for dystonia as well. Rarely, carbidopa/levodopa can improve dystonic symptoms. In severe or refractory cases of dystonia, deep brain stimulation is used. The globus pallidus internus is usually the target.

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