Essential tremor: a nuanced approach to the clinical features

Elan D Louis^{1,2,3}

ABSTRACT

Essential tremor is the most common form of tremor in humans. Given neurologists' high exposure to this condition, and its seemingly straightforward phenotype, it might seem easy to diagnose. However, 30%-50 % of patients labelled as having 'essential tremor' have other diagnoses, mostly Parkinson's disease and dystonia. The tremor of essential tremor is neither non-descript nor featureless but is multifaceted and highly patterned. This review focuses on its clinical features, beginning with a discussion of tremors and then briefly discussing its additional motor features, and presents several aids to help distinguish essential tremor from Parkinson's disease and dystonia. Careful attention to certain clinical nuances will aid the diagnosis and care of patients with essential tremor.

INTRODUCTION

Essential tremor is a chronic, progressive neurological disease (or perhaps family of diseases)¹ whose primary clinical feature is kinetic tremor (ie, tremor during voluntary movement) (table 1).² It is the most common form of tremor in humans^{3–5} and among most prevalent of movement disorders. Given neurologists' high exposure to this common entity as well as its seemingly straightforward phenotype, one might think it is easy to diagnose. However, this is often not the case⁶ and its diagnosis is often quite problematic.⁷ Some 30%–50% of 'essential tremor' cases have other diagnoses, particularly Parkinson's disease or dystonia.⁸⁹ The tremor of essential tremor is neither non-descript nor undifferentiated but is rich in attributes, multifaceted and highly patterned. Careful attention to its subtle clinical features will aid the diagnosis and ultimately the care of patients with essential tremor. This review focuses on the clinical features of essential tremor, beginning with a discussion of tremor(s) (see discussion points 1-11, table 2) and its additional motor features (see discussion points 12 and 13, table 2), and then reviewing the sometimes-difficult distinctions between essential tremor and either Parkinson's disease or dystonia (table 3).

ESSENTIAL TREMOR: CREATING A REFINED MAP OF THE TREMOR Discussion point 1: Its central clinical feature is kinetic tremor of the arms and hands

Although the upper limb tremor of essential tremor is commonly (and somewhat loosely) described as either postural or kinetic, closer inspection shows that kinetic tremor nearly always has a greater amplitude than postural tremor.²¹⁰ Thus, the central clinical feature of essential tremor is kinetic tremor of the arms and hands.² This tremor can be observed during numerous commonly performed activities of daily living, ranging from eating to drinking to writing, and it may be elicited on neurological examination during various manoeuvres (eg, finger-nose-finger manoeuvre, drinking from a cup, pouring water between two cups and writing) (figure 1).⁷ This kinetic tremor may range in severity from mild-almost negligible and difficult to distinguish from normal physiological tremor (figure 2)-to one that is severe and functionally disabling (figure 3).¹¹ Furthermore, key features of the tremor are that it is both rhythmical (ie, regularly recurrent) and oscillatory (ie, the movements rotate symmetrically around a central plane).¹²

Discussion point 2: The upper limb tremor is generally slightly asymmetric

Rather than being strictly symmetric, the upper limb tremor in essential tremor is generally mildly asymmetric (figure 4).^{13 14} The tremor generally has a slightly greater amplitude in one arm than in the other. In a study of 54 patients with essential tremor, the mean side-to-side difference in clinical ratings for each of six tasks (sustained arm extension, pouring water, drinking water, using a spoon, finger-to-nose movements and drawing spirals with each arm) was 0.54 of 3 points, a 1.3-fold difference

► Additional material is published online only. To view please visit the journal online (http://dx.doi.org/ 10.1136practneurol-2018-002183)

¹Division of Movement Disorders, Department of Neurology, Yale School of Medicine, Yale University, New Haven, CT, USA ²Center for Neuroepidemiology and Clinical Neurological Research, Yale School of Medicine, Yale University, New Haven, CT, USA ³Department of Chronic Disease Epidemiology, Yale School of Public Health, Yale University, New Haven, CT, USA

Correspondence to

Dr Elan D Louis, Yale Neurology, Division of Movement Disorders, New Haven, CT, USA; elan. Iouis@yale.edu

Accepted 26 April 2019



© Author(s) (or their employer(s)) 2019. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Louis ED. Pract Neurol Epub ahead of print: [please include Day Month Year]. doi:10.1136/ practneurol-2018-002183

Table 1	Tremor types, based on activation condition	
Type of tremor	Definition	
Action tremor	Tremor that occurs during a voluntary contraction of skeletal muscle. This includes isometric, kinetic and postural tremors	
lsometric tremor	Tremor that occurs when a muscle contracts against a rigid, stationary object (eg, while making a fist)	
Kinetic tremor	Tremor that occurs during a voluntary movement (eg, while writing or eating)	
Postural tremor	Tremor that occurs when the body part is held motionless against the force of gravity (eg, while arms are outstretched in front of the body)	
Rest tremor	Tremor that occurs in a body part that is supported against gravity and inactive (eg, while lying down)	

between sides.¹³ In another study of quantified computerised tremor analysis, there was a 1.71-fold mean difference between tremor amplitudes in the dominant and non-dominant sides.¹³ In about 5% of people with essential tremor, this tremor is markedly asymmetric or unilateral.¹⁵

Discussion point 3: While drawing a spiral, the waveforms often align along a single predominant axis rather than several axes

Kinetic tremor in essential tremor is often present during writing tasks, for example, drawing an Archimedes

spiral or writing sentences.¹⁶⁻¹⁸ While drawing a spiral, the wave forms of the tremor often align along a single predominant axis (figure 5, figures 1 and 2); however, in patients with dystonia there may be several axes rather than a single axis.^{19 20} As the wave forms align, the tremor in spirals drawn with the right hand is most severe at 1-2 o'clock and 7-8 o'clock (ie, upper right and lower left quadrants), whereas those drawn with the left hand have a single identifiable tremor orientation axis that is 90° to that on the right (ie, tremor is most severe at 10-11 o'clock and 4-5 o'clock (lower right and upper left quadrants)). In a study of 120 people with essential tremor and 15 people with dystonic tremor, there was a single, clearly identifiable axis on one-half or more of spirals in 67.5% of essential tremor cases and only 40% of dystonia cases; in dystonia cases there were often several axes visible, some of them indistinct (figure 6).²⁰ Hence, while this finding does not completely distinguish the two diagnoses, it can suggest the more likely diagnosis.

Discussion point 4: In many patients the upper limb tremor has an intentional component

In about half of people with essential tremor, the upper limb tremor has an intentional component, with a slight or greater worsening (ie, increase in tremor amplitude) as the patient approaches the target during the finger-nosefinger manoeuvre (figures 4 and 7).²¹ Moreover, the

Discussion point number	Brief synopsis of discussion point	Relevant figure visually illustrating the point
1	The central clinical feature of essential tremor is kinetic tremor of the arms and hands	figure 1, figure 3
2	The upper limb tremor in essential tremor is generally slightly asymmetric	figure 4
3	While drawing a spiral, the wave forms often align along a single predominant axis rather than several axes	figure 5, figure 1, figure 2
4	In many patients, the upper limb tremor has a clear intentional component, with a slight or greater worsening as the patient approaches the target	figure 4, figure 7
5	The amplitude of postural tremor is generally less than that of the kinetic tremor; the postural tremor in the two arms is often out of phase	figure 8, figure 9.
6	The postural tremor is generally of greatest amplitude at the wrist or elbow and generally comprises a wrist flexion–extension movement rather than rotation–supination	figure 10
7	Rest tremor may develop in the arm in severe and longstanding essential tremor cases; it does not develop in the leg	figure 11
8	The tremor of essential tremor is not static; in most people with essential tremor, it gradually worsens over time	figure 12
9	Over time, the tremor in essential tremor tends to spread beyond the upper limbs to cranial structures; this occurs more often in women than men	figure 4
10	Unless it is particularly severe, this neck tremor, which is a postural tremor, should resolve while the patient lies on their back with their head fully at rest	
11	In contrast to Parkinson's disease, in which the jaw tremor develops when the mouth is closed, in essential tremor, the jaw tremor more often develops when the mouth is open (eg, during speech or when asked to open their mouths)	figure 13
12	Another motor feature of essential tremor is gait ataxia, which may be brought out by asking patients to walk tandem	figure 14
13	Some people with essential tremor may show mild dystonic posturing in an arm that has had longstanding and severe tremor	figure 9

		REVIEW			
Table 3 Aids to the differential diagnosis of essential tremor					
Diagnostic aid	Caveat	Essential tremor versus Parkinson's disease or essential tremor versus dystonia?			
Patients with essential tremor do not have rigidity	Cogwheeling (without rigidity) may occur in patients with essential tremor	Essential tremor versus Parkinson's disease			
Patients with essential tremor do not have hypomimia		Essential tremor versus Parkinson's disease			
Patients with essential tremor do not have bradykinesia accompanied by decrement	As many essential tremor patients are elderly, rapid alternating movements may be slow; however, true decrement should not be present nor should it be lateralising (ie, present on one side only) unless the patient is developing essential tremor–Parkinson's disease	Essential tremor versus Parkinson's disease			
Isolated rest tremor is not a feature of essential tremor		Essential tremor versus Parkinson's disease			
Isolated postural tremor is not a feature of essential tremor		Essential tremor versus Parkinson's disease			
Postural tremor predominantly involving the metacarpophalangeal joints rather than the wrist is not a feature of essential tremor		Essential tremor versus Parkinson's disease			
Postural tremor characterised by greater wrist rotation than wrist flexion and extension is not a feature of essential tremor		Essential tremor versus Parkinson's disease			
Re-emergent tremor (ie, tremor emerging after a latency) is not a feature of essential tremor; it is a feature of Parkinson's disease	Postural tremor of essential tremor may vary from moment to moment, which means that it may be absent or of low amplitude initially and then emerge after some time	Essential tremor versus Parkinson's disease			
Aside from a mild dystonic posture in patients with severe and longstanding essential tremor, dystonic postures, movements or tremor are not currently accepted as features of essential tremor		Essential tremor versus dystonia			
Dystonic tremor itself is often neither rhythmic nor oscillatory		Essential tremor versus dystonia			
The presence of moderate or marked head tremor in the absence of limb tremor is highly suggestive of an underlying diagnosis of dystonia		Essential tremor versus dystonia			
Unless it is particularly severe, the neck tremor of essential tremor, which is a postural tremor, should resolve while the patient lies on their back with their head fully at rest; in contrast, persistence of such tremor is often a sign of an underlying diagnosis of dystonia		Essential tremor versus dystonia			
Patients with neck tremor and underlying diagnoses of dystonia may have signs of dystonia, including head tilt or		Essential tremor versus dystonia			

Pa dystonia may have signs of dystonia, including head tilt or rotation, hypertrophy of the sternocleidomastoid or other neck muscles, the presence of a tremor null-point, or a sensory trick by history (ie, a manoeuvre such as touching the chin or back of the head that lessens the tremor)

intention tremor is not limited to the upper limbs; 10% of patients have such tremor in their neck when their head approaches a target, for example, while drinking from a cup, the head may move downwards towards the cup (target), and there may be an intention tremor in the head as it approaches the target.²² Some people also have intention tremor in the legs, although this is generally not tested for on neurological examination.²³

Discussion point 5: The amplitude of postural tremor is generally less than that of the kinetic tremor; the postural tremor in the two arms is often out of phase In addition to the upper limb kinetic tremor,^{2 10} people with essential tremor often have an upper limb postural

tremor, although its amplitude is almost always less than that of the kinetic tremor (figure 8).^{2 10} In a study of 369 essential tremor cases, 95% showed a kinetic tremor that was more severe than postural tremor.² In nearly one-third of cases (32.8%), the kinetic tremor score was ≥ 1 points higher than the postural tremor score.² Conversely, only rarely (\sim 5%) was the postural tremor even marginally (<1 point) more severe than kinetic tremor, and in no case was the postural tremor score ≥ 1 point higher than the kinetic tremor score.² The postural tremor of essential tremor has other distinguishing features.² Although subtle, tremor amplitude can increase when switching from the arms-outstretched position to the arms in a wing-beat position.²⁴

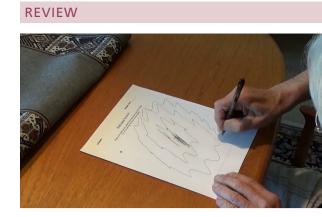


Figure 1 The kinetic tremor of essential tremor can be observed during numerous commonly performed activities of daily living; this patient develops severe tremor while drawing a spiral. As the wave forms align, the tremor in spirals drawn with the right hand is most severe at 1–2 o'clock and 7–8 o'clock (ie, upper right and lower left quadrants) (see discussion point 3 as well). See (online supplementary file 1) for video.

The postural tremor in the two arms is often out of phase, sometimes creating seesaw effect with the arms held in a wing-beat position (figure 9).^{7 12} Interestingly, this lack of phasic synchrony often improves functionality when the patient uses two hands rather than one to hold a glass or cup, because the tremors in each arm, being out of phase, cancel one another out to some degree.^{7 12}

Discussion point 6: The postural tremor is of greatest amplitude at the wrist or elbow and comprises a wrist flexion–extension movement

The postural tremor of essential tremor does not affect all upper limb segments equally. Its greatest amplitude is generally at the wrist or elbow, rather than metacarpophalangeal joints, and it usually comprises a wrist flexion–extension movement rather than rotation– supination, although not always (figure 10).^{7 12 25}



Figure 3 This patient's tremor is severe and functionally disabling. Attempting to use a spoon to bring water to the mouth proves nearly impossible. See (online supplementary file 3) for video.

Discussion point 7: Rest tremor may develop in the arm in severe and longstanding cases; it does not develop in the leg

Tremor at rest, without other cardinal features of parkinsonism such as bradykinesia or rigidity, develops in 1%-35% of patients with essential tremor, depending on the method of case ascertainment (eg, less among cases ascertained from a population and more among cases ascertained from a brain repository). However, in contrast to that of Parkinson's disease, rest tremor is a late feature among people with essential tremor who have marked kinetic tremor, and only ever develops in the arm (ie, not the leg) (figure 11).^{7 12 26-29} When assessing people with essential tremor for the presence of rest tremor, it is important that the arm is truly fully supported and at rest while it lies in their lap, otherwise kinetic tremor may be mistaken for rest tremor. The examiner may need to lie the patient down on their back to achieve a proper rest condition. As a final point, rest tremor in essential tremor does not occur in limbs in the absence of kinetic tremor.



Figure 2 The kinetic tremor, when initially present in essential tremor, is mild and almost negligible. This patient is drawing a spiral with the left hand and tremor emerges at several points while drawing the spiral, but primarily in the 10–11 o'clock and at 4–5 o'clock positions (see discussion point 3 as well). See (online supplementary file 2) for video.



Figure 4 The upper limb tremor in essential tremor is generally slightly asymmetric, affecting one arm slightly more than the other. In this patient, tremor during the finger–nose–finger manoeuvre is more marked on the left than right side. There is an intentional component to the tremor, particularly as she approaches her nose, and more evident on the left than right side. See (online supplementary file 4) for video.

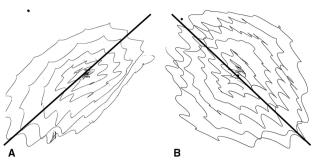


Figure 5 While drawing a spiral, the wave in essential tremor may align along a single predominant axis rather than several axes. As the wave forms align, the tremor in spirals drawn with the right hand (patient A) is most severe at 1–2 o'clock and 7–8 o'clock (ie, upper right and lower left quadrants), whereas those drawn with the left hand (patient B) have a single identifiable tremor orientation axis that is 90° to that on the right (ie, tremor is most severe at 10–11 o'clock and 4–5 o'clock (lower right and upper left quadrants)). Lines indicate axes.

Discussion point 8: The tremor usually gradually worsens over time

In most people with essential tremor, the tremor gradually worsens over time (figure 12).^{30 31} There are several patterns of clinical progression, the two most common being¹ late-life onset (after aged 60) with steady progression and² early-life onset (before aged 40) with mild, stable tremor for many years, which then, in the 60 s and onwards, progresses steadily. The least common pattern is of early-life onset with marked worsening over the ensuing decade. There are surprisingly few prospective, longitudinal natural history studies of essential tremor; however, the best estimates

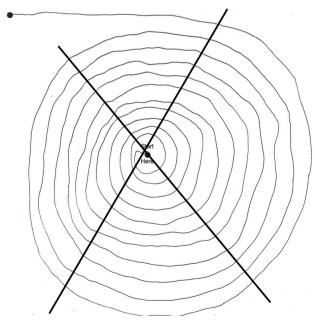


Figure 6 While drawing a spiral, the wave in a patient with dystonia may align along a several axes, and these may be indistinct. Lines indicate possible axes.



Figure 7 Intentional tremor, with a slight or greater worsening (ie, increase in tremor amplitude) develops as the patient approaches the target during the finger–nose–finger manoeuvre. This patient's intention tremor is most evident when he approaches his nose. See (online supplementary file 5) for video.

indicate that arm tremor worsens at an annual rate of 2%-5%.³¹

Discussion point 9: Over time the tremor spreads beyond the upper limbs to cranial structures

The tremor in essential tremor tends over time to spread beyond the upper limbs, with patients developing cranial tremors, involving the neck (most commonly involved), voice, or jaw.^{32–35} Early, marked head tremor without limb tremor is a marker of dystonia.^{7 12} Cranial tremors are particularly common in women with essential tremor: neck tremor is several times more likely in women with essential tremor than in men.^{7 12 35 36}

Discussion point 10: Unless it is particularly severe, the neck tremor, which is a postural tremor, should resolve while the patient lies on their back with their head fully at rest

The neck tremor of essential tremor often begins as a unidirectional tremor, either as a 'no–no' (ie, horizontal) or 'yes–yes' (ie, vertical) tremor.³⁷ Over many years, this can both increase in severity and evolve into

Clinical case 1

A 74-year-old man had noticed a tremor when holding a spoon to eat soup and when signing cheques at the bank. It had been present for 6 years but had been more noticeable for three. On examination, there was a moderate amplitude tremor during the finger—nose—finger manoeuvre, slightly worse on the right than the left. There was also terminal worsening of tremor as his finger approached the examiner's finger. There was a similar degree of tremor while pouring water between two cups. When drawing a spiral, the wave forms of the tremor lined up to form a single axis. Based on these physical examination findings, the physician diagnosed essential tremor. Additional history indicated that the patient's father and brother had a similar condition.

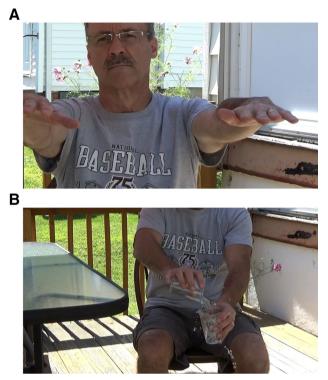


Figure 8 The postural tremor of essential tremor is generally less severe than the kinetic tremor. (A) This patient has mild postural tremor, more evident in the left than the right arm. (B) His kinetic tremor, evident while pouring, is clearly more severe than his postural tremor. See (online supplementary files 6; 7) for video.

a more complex tremor that is multidirectional tremor and more difficult to characterise.³⁷ Unless it is particularly severe, this neck tremor—which is a postural tremor—should resolve while the patient lies on their back with their head fully at rest.³⁸ In one study of people with essential tremor who had a head tremor while seated, the tremor resolved in 92% when supine. Among the small percentage in whom head tremor persisted while supine, the head tremor was severe.³⁸



Figure 9 The postural tremor in the two arms is often out of phase, sometimes creating a seesaw effect with the arms held in a wing-beat position. This patient has severe postural tremor. Some patients with essential tremor may show mild dystonic posturing in an arm that has had longstanding and severe tremor. This patient has flexion of the right thumb, which is likely dystonic. See (online supplementary file 8) for video.



Figure 10 When held outstretched and forward, the postural tremor of essential tremor is generally of greatest amplitude at the wrist (sometimes elbow) rather than metacarpophalangeal joints, and generally comprises a wrist flexion–extension movement rather than rotation–supination. See (online supplementary file 9) for video.

An interesting feature of the neck tremor in essential tremor is that patients are often unaware of its presence (ie, they have an agnosia for it), particularly early on when it is mild.^{39 40} In one study, the authors asked people with essential tremor whether they were aware of head tremor while it was occurring on examination, thereby allowing them to gauge real-time awareness of their involuntary movement.³⁹ Half of those with visible head tremor on examination reported having no head tremor at that moment. Even among those with examination evidence of moderate or severe head tremor, 45% were unaware of head tremor.³⁹

Discussion point 11: Jaw tremor may occur with the mouth closed in Parkinson's disease, and with the mouth open in essential tremor (eg, during speech or when asked to open their mouths)

Jaw tremor may develop in people with essential tremor when they develop cranial tremors. Between 7.5% and 18.0% of people with essential tremor develop jaw tremor, the lowest prevalence being in a population-based sample. People with essential tremor

Clinical case 2

A 69-year-old man had noticed tremor for 2–3 years. This developed when using his hands to eat and also while typing, causing him to double-click the letters on the key pad and to make mistakes. His work involved a lot of typing and so this caused him considerable frustration and consternation. On examination, he developed tremor during various tasks (eg, drinking from a cup, drawing a spiral) and also, although much less so, during arm extension, mainly at the wrist, which shook up and down. When asked to hold a cup of water in the midline with both hands, the cup shook less than when holding the cup with either of the two hands. Based on this examination, the physician diagnosed essential tremor.

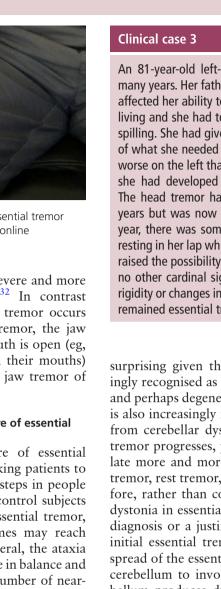


Figure 11 This patient with longstanding essential tremor shows mild tremor at rest in both hands. See (online supplementary file 10) for video.

and jaw tremor had a more clinically severe and more topographically widespread disorder.³² In contrast to Parkinson's disease, where the jaw tremor occurs with the mouth closed, in essential tremor, the jaw tremor is more common when the mouth is open (eg, during speech or when asked to open their mouths) (figure 13). In this sense, it is more a jaw tremor of activity than rest.^{7 12 29 32-34}

Discussion point 12: Gait ataxia is a feature of essential tremor

Gait ataxia is another motor feature of essential tremor, and may be brought out by asking patients to walk tandem.^{41–44} The number of mis-steps in people with essential tremor exceeds that in control subjects of similar age. In most people with essential tremor, this ataxia is mild, although sometimes may reach moderate severity (figure 14).⁴⁵ In general, the ataxia leads people to have reduced confidence in balance and a mild but significant increase in the number of near-falls and falls compared with age-matched controls.⁴⁶ It is not so severe that patients become wheelchair restricted.

Discussion point 13: Some people may show mild dystonic posturing in an arm that has had longstanding and severe tremor

Some patients with essential tremor may show mild dystonic posturing in an arm that has had longstanding and severe tremor (figure 9). This is not An 81-year-old left-handed woman had had tremor for many years. Her father and sister had a similar tremor. This affected her ability to perform numerous activities of daily living and she had to drink from a cup with a lid to avoid spilling. She had given up writing and now dictated much of what she needed to write. The tremor had always been worse on the left than right side. About 5 years previously. she had developed a head tremor and a voice tremor. The head tremor had been barely noticeable for several years but was now present nearly all the time. This past year, there was some mild tremor in her left hand when resting in her lap while watching television; her doctor had raised the possibility of Parkinson's disease but there were no other cardinal signs of that disorder (ie, bradykinesia, rigidity or changes in gait or balance). Hence, the diagnosis remained essential tremor.

surprising given that (1) essential tremor is increasingly recognised as a disease arising from an abnormal and perhaps degenerative cerebellum;^{47–52} (2) dystonia is also increasingly regarded as a disease arising partly from cerebellar dysfunction^{53 54}; and (3) as essential tremor progresses, patients tend to layer and accumulate more and more neurological signs (eg, intention tremor, rest tremor, cranial tremors).^{21 26 27 34 55} Therefore, rather than conceptualising the development of dystonia in essential tremor as either a second clinical diagnosis or a justification or rationale to retract the initial essential tremor diagnosis, it may represent a spread of the essential tremor-related pathology in the cerebellum to involve whichever system in the cerebellum produces dystonic postures/movements.⁵⁶ Yet at the moment, there is no consensus on how much dystonia is acceptable within the definition of essential tremor.⁵⁷ If the dystonia is in a body part that is not tremulous (eg, in the neck in a patient with upper limb tremor), the addition of a second diagnosis (dystonia) is more justifiable than in a patient with longstanding, severe and otherwise phenomenologically typical essential tremor who has developed mild posturing in a limb that shakes.

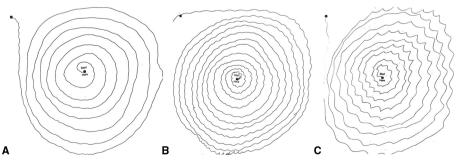


Figure 12 The tremor of essential tremor is not static. In most people with essential tremor, the tremor gradually worsens over time. This patient drew spirals at age 49 (A), 61 (B) and 74 (C).

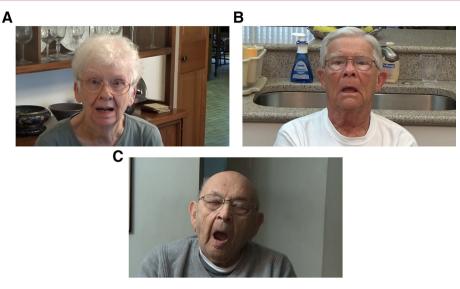


Figure 13 In contrast to Parkinson's disease, in which there is jaw tremor when the mouth is closed, the jaw tremor in essential tremor is more likely when the mouth is open (eg, during phonation or when asked to open their mouths), as in these three people with essential tremor. See (online supplementary file 11) for video.

AIDS TO THE DIFFERENTIAL DIAGNOSIS Essential tremor versus Parkinson's disease

The diagnosis of essential tremor is often quite problematic,⁷ and several studies have shown that 30%–50% of 'essential tremor' cases have other diagnoses, particularly Parkinson's disease and dystonia.⁸

There are several distinguishing points. Patients with essential tremor do not have rigidity, hypomimia and bradykinesia accompanied by decrement, for example, no sequential decrement in amplitude during finger taps. There are several caveats. First, people with essential tremor may have cogwheeling (without rigidity). Second, as many people with essential tremor are elderly, rapid alternating movements may be slow; however, there should not be true decrement nor should it be lateralising (ie, present on one side only) unless the patient is developing essential tremor– Parkinson's disease.

The characteristics of the tremor can also help to distinguish essential tremor from Parkinson's disease. Parkinson's disease is more likely if there is an isolated



Figure 14 This patient with essential tremor has moderately severe difficulty with tandem gait. See (online supplementary file 12) for video.

rest tremor (ie, rest tremor without kinetic tremor), isolated postural tremor (ie, postural tremor with no or minimal kinetic tremor), postural tremor predominantly involving the metacarpophalangeal joints rather than the wrist, or postural tremor characterised by greater wrist rotation than wrist flexion and extension.⁷ ¹² ²⁵ ²⁹ Another feature of Parkinson's disease is re-emergent tremor, a type of postural tremor that starts after a brief latency of several seconds.⁵⁸ Note, however, that the postural tremor of essential tremor may vary from moment to moment, and so it may be absent or of low amplitude initially and then emerge after some time.

Essential tremor versus dystonia

Aside from a mild dystonic posture, dystonic postures, movements or tremor are not currently accepted as features of essential tremor. In addition, dystonic tremor itself is often neither rhythmic nor oscillatory.

Although it can be difficult to evaluate people with head tremor, it is worth considering several points. First, moderate or marked head tremor without limb tremor strongly suggests an underlying dystonia.⁷ ¹² Second, unless it is particularly severe, the neck tremor of essential tremor-which is a postural tremor-should resolve while the patient lies on their back with their head fully at rest³⁸; in contrast, persistence of such tremor suggests an underlying diagnosis of dystonia.³⁸ Third, patients with neck tremor and underlying diagnoses of dystonia may have signs of dystonia, including head tilt or rotation, hypertrophy of the sternocleidomastoid or other neck muscles, the presence of a tremor nullpoint, or a sensory trick by history (ie, a manoeuvre such as touching the chin or back of the head that lessens the tremor).

Key points

- Essential tremor is the most common form of tremor in humans.
- ► 30%–50% of 'essential tremor' cases have diagnoses other than essential tremor.
- The phenotype of essential tremor carries within it more nuance than previously thought; indeed, the tremor is multifaceted and highly patterned.
- Careful attention to these clinical nuances will aid in the diagnosis and care of patients with essential tremor.

Funding This study was funded by NINDS (R01NS073872, R01NS086736, R01NS088257, R01NS094607).

Competing interests None declared.

Patient consent for publication Not required.

Provenance and peer review Commissioned. Externally peer reviewed by Peter Bain,London, UK

REFERENCES

- 1 Louis ED. 'Essential tremor' or 'the essential tremors': is this one disease or a family of diseases? *Neuroepidemiology* 2014;42:81–9.
- 2 Louis ED. The primary type of tremor in essential tremor is kinetic rather than postural: cross-sectional observation of tremor phenomenology in 369 cases. *Eur J Neurol* 2013;20:725–7.
- 3 Louis ED, Ferreira JJ. How common is the most common adult movement disorder? Update on the worldwide prevalence of essential tremor. *Mov. Disord.* 2010;25:534–41.
- 4 Benito-Leon J, Bermejo-Pareja F, Morales JM, *et al.* Prevalence of essential tremor in three elderly populations of central Spain. *Mov Disord* 2003;18:389–94.
- 5 Seijo-Martinez M, Del Rio MC, Alvarez JR, *et al.* Prevalence of essential tremor on Arosa Island, Spain: a community-based, door-to-door survey. *Tremor Other Hyperkinet Mov* 2013;3.
- 6 Nahab FB, Peckham E, Hallett M. Essential tremor, deceptively simple. *Pract Neurol* 2007;7:222–33.
- 7 Louis ED. Twelve clinical pearls to help distinguish essential tremor from other tremors. *Expert Review of Neurotherapeutics* 2014;14:1057–65.
- 8 Jain S, SE L, Louis ED. Common misdiagnosis of a common neurological disorder: how are we misdiagnosing essential tremor? *Arch Neurol* 2006;63:1100–4.
- 9 Schrag A, Munchau A, Bhatia KP, *et al.* Essential tremor: an overdiagnosed condition? *J Neurol* 2000;247:955–9.
- 10 Brennan KC, Jurewicz EC, Ford B, *et al.* Is essential tremor predominantly a kinetic or a postural tremor? A clinical and electrophysiological study. *Mov Disord* 2002;17:313–6.
- 11 Louis ED, Barnes L, Albert SM, et al. Correlates of functional disability in essential tremor. Mov Disord 2001;16:914–20.
- 12 Louis ED. Diagnosis and management of tremor. *Continuum* 2016;22:1143–58.
- 13 Louis ED, Wendt KJ, Pullman SL, *et al.* Is essential tremor symmetric? observational data from a community-based study of essential tremor. *Arch Neurol* 1998;55:1553–9.
- 14 Biary N, Koller W. Handedness and essential tremor. Arch Neurol 1985;42:1082–3.

- 15 Phibbs F, Fang JY, Cooper MK, et al. Prevalence of unilateral tremor in autosomal dominant essential tremor. Mov Disord 2009;24:108–11.
- 16 Louis ED, Ford B, Wendt KJ, et al. A comparison of different bedside tests for essential tremor. Mov Disord 1999;14:462–7.
- 17 Bain PG, Findley LJ, Atchison P, et al. Assessing tremor severity. J Neurol Neurosurg Psychiatry 1993;56:868–73.
- 18 Gutierrez J, Park J, Badejo O, et al. Worse and worse and worse: essential tremor patients' longitudinal perspectives on their condition. Front Neurol 2016;7.
- 19 Louis ED, Yu Q, Floyd AG, et al. Axis is a feature of handwritten spirals in essential tremor. Mov Disord 2006;21:1294–5.
- 20 Michalec M, Hernandez N, Clark LN, *et al*. The spiral axis as a clinical tool to distinguish essential tremor from dystonia cases. *Parkinsonism Relat Disord* 2014.
- 21 Louis ED, Frucht SJ, Rios E. Intention tremor in essential tremor: prevalence and association with disease duration. *Mov Disord* 2009.
- 22 Leegwater-Kim J, Louis ED, Pullman SL, *et al.* Intention tremor of the head in patients with essential tremor. *Mov Disord* 2006;21:2001–5.
- 23 Kestenbaum M, Michalec M, Yu Q, *et al.* Intention tremor of the legs in essential tremor: prevalence and clinical correlates. *Mov Disord Clin Pract* 2015;2:24–8.
- 24 Sanes JN, Hallett M. Limb positioning and magnitude of essential tremor and other pathological tremors. *Mov Disord* 1990;5:304–9.
- 25 Sternberg EJ, Alcalay RN, Levy OA, *et al.* And intention tremors: a detailed clinical study of essential tremor vs. Parkinson's disease. *Front Neurol* 2013;4.
- 26 Cohen O, Pullman S, Jurewicz E, *et al.* Rest tremor in patients with essential tremor: prevalence, clinical correlates, and electrophysiologic characteristics. *Arch Neurol* 2003;60:405–10.
- 27 Louis ED, Hernandez N, Michalec M. Prevalence and correlates of rest tremor in essential tremor: cross-sectional survey of 831 patients across four distinct cohorts. *Eur J Neurol* 2015;22:927–32.
- 28 Koller WC, Glatt S, Biary N, et al. Essential tremor variants: effect of treatment. Clin Neuropharmacol 1987;10:342–50.
- 29 Thenganatt MA, Louis ED. Distinguishing essential tremor from Parkinson's disease: bedside tests and laboratory evaluations. *Expert Rev Neurother* 2012;12:687–96.
- 30 Putzke JD, Whaley NR, Baba Y, et al. Essential tremor: predictors of disease progression in a clinical cohort. J Neurol Neurosurg Psychiatry 2006;77:1235–7.
- 31 Louis ED, Agnew A, Gillman A, et al. Estimating annual rate of decline: prospective, longitudinal data on arm tremor severity in two groups of essential tremor cases. J Neurol Neurosurg Psychiatry 2011;82:761–5.
- 32 Louis ED, Rios E, Applegate LM, *et al.* Jaw tremor: prevalence and clinical correlates in three essential tremor case samples. *Mov Disord* 2006;21:1872–8.
- 33 Hernandez NC, Louis ED. Jaw tremor resulting in broken teeth: on the essential tremor spectrum. *Tremor Other Hyperkinet Mov* 2015;5.
- 34 Louis ED, Gerbin M, Galecki M. Essential tremor 10, 20, 30,
 40: clinical snapshots of the disease by decade of duration. *Eur J Neurol* 2013;20:949–54.
- 35 Louis ED, Ford B, Frucht S. Factors associated with increased risk of head tremor in essential tremor: a community-based study in Northern Manhattan. *Mov Disord* 2003;18:432–6.

- 36 Hardesty DE, Maraganore DM, Matsumoto JY, et al. Increased risk of head tremor in women with essential tremor: longitudinal data from the Rochester epidemiology project. Mov Disord 2004;19:529–33.
- 37 Robakis D, Louis ED. Head tremor in essential tremor: "Yesyes", "no-no", or "round and round"? *Parkinsonism Relat Disord* 2016;22:98–101.
- 38 Agnew A, Frucht SJ, Louis ED. Supine head tremor: a clinical comparison of essential tremor and spasmodic torticollis patients. J Neurol Neurosurg Psychiatry 2012;83:179–81.
- 39 Eken HN, Louis ED. Agnosia for head tremor in essential tremor: prevalence and clinical correlates. J Clin Mov Disord 2016;3.
- 40 Louis ED, Pellegrino KM, Rios E. Unawareness of head tremor in essential tremor: a study of three samples of essential tremor patients. *Mov Disord* 2008.
- 41 Arkadir D, Louis ED. The balance and gait disorder of essential tremor: what does this mean for patients? *Ther Adv Neurol Disord* 2013;6:229–36.
- 42 Hoskovcova M, Ulmanova O, Sprdlik O, *et al.* Disorders of balance and gait in essential tremor are associated with midline tremor and age. *Cerebellum* 2012.
- 43 Hubble JP, Busenbark KL, Pahwa R, et al. Clinical expression of essential tremor: effects of gender and age. Mov Disord 1997;12:969–72.
- 44 Kronenbuerger M, Konczak J, Ziegler W, et al. Balance and motor speech impairment in essential tremor. Cerebellum 2009;8:389–98.
- 45 Louis ED, Galecki M, Rao AK. Four essential tremor cases with moderately impaired gait: how impaired can gait be in this disease? *Tremor Other Hyperkinet Mov* 2013;3.
- 46 Rao AK, Gilman A, Louis ED. Balance confidence and falls in nondemented essential tremor patients: the role of cognition. *Archives of physical medicine and rehabilitation* 2014;95:1832–7.

- 47 Benito-Leon J, Labiano-Fontcuberta A. Linking essential tremor to the cerebellum: Clinical evidence. *Cerebellum* 2016;15:253–62.
- 48 Marin-Lahoz J, Gironell A. Linking essential tremor to the cerebellum: neurochemical evidence. *Cerebellum* 2016;15:243–52.
- 49 Louis ED. Linking essential tremor to the cerebellum: neuropathological evidence. *Cerebellum* 2016;15:235–42.
- 50 Filip P, Lungu OV, Manto MU, et al. Linking essential tremor to the cerebellum: physiological evidence. Cerebellum 2016;15:774–80.
- 51 Handforth A. Linking essential tremor to the Cerebellum-Animal model evidence. *Cerebellum* 2016;15:285–98.
- 52 Cerasa A, Quattrone A. Linking essential tremor to the Cerebellum-Neuroimaging evidence. *Cerebellum* 2016;15:263–75.
- 53 Bologna M, Berardelli A. The cerebellum and dystonia. *Handb Clin Neurol* 2018;155:259–72.
- 54 Bares M, Filip P. Cerebellum and dystonia: the story continues.
 Will the patients benefit from New discoveries? *Clin Neurophysiol* 2018;129:282–3.
- 55 Louis ED, Asabere N, Agnew A, et al. Rest tremor in advanced essential tremor: a post-mortem study of nine cases. J Neurol Neurosurg Psychiatry 2011;82:261–5.
- 56 Louis ED. The evolving definition of essential tremor: what are we dealing with? *Parkinsonism Relat Disord* 2018;46 Suppl 1(Suppl 1):S87–S91.
- 57 Bhatia KP, Bain P, Bajaj N, *et al.* Consensus statement on the classification of tremors. from the task Force on tremor of the International Parkinson and movement disorder society. *Mov Disord* 2018;33:75–87.
- 58 Jankovic J, Schwartz KS, Ondo W. Re-emergent tremor of Parkinson's disease. J Neurol Neurosurg Psychiatry 1999;67:646–50.