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Triggering Essential Head Tremor with Sustained Phonation: A Clinical Phenomenon with Potential Diagnostic Value

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Abstract

Here we report a clinical phenomenon that we have observed repeatedly in clinical research settings; namely, a triggering and/or exacerbation of head tremor during or immediately following sustained phonation. To our knowledge, it has not been reported previously nor has it been the subject of study. Here we: 1) report the phenomenon, 2) provide several visual illustrations, 3) estimate its prevalence, and 4) analyze its clinical correlates. Head tremor was assessed qualitatively, and scores were assigned pre-, during, and post-sustained phonation using the Tremor Research Group Essential Tremor Rating Assessment Scale tremor rating scale. Seventy (68.6%) of 102 essential tremor (ET) patients exhibited a qualitative increase in head tremor amplitude during and/or immediately after sustained phonation; in 5 (4.9%), head tremor would not have been detected without the voice activation maneuver (i.e., it was not visible at any other point in the videotape aside from the period during/immediately following sustained phonation). Women were more likely than men to exhibit this phenomenon ($p=0.05$), whereas age, age at onset, duration of tremor, and total tremor score did not predict responsiveness of head tremor to sustained phonation. Sustained voice activation is a useful examination maneuver that may elicit or amplify head tremor in ET. Head tremor is not reported to occur in patients with enhanced physiological tremor. Thus, this maneuver, by triggering head tremor, may be a useful diagnostic supplement, particularly in research/clinical settings where arm tremor is mild and the diagnosis (mild ET vs. enhanced physiological tremor) would otherwise be ambiguous.

Keywords

Essential tremor; clinical; head tremor; examination

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Introduction

Head tremor, resulting from oscillating contractions of the neck muscles, is one of the most common clinical manifestations of essential tremor (ET), with reported prevalence as high as 39–50% in clinical studies [1–5]. In individuals with marginal levels of limb tremor, the presence of head tremor is usually a clear indicator that the correct diagnosis is ET or dystonia rather than enhanced physiological tremor; the absence of dystonic features favors ET in such patients. In research settings, and especially genetic studies, the presence of this clinical feature (head tremor) is of particular diagnostic utility among young diagnostically-ambiguous relatives with only mild/marginal upper extremity tremor.

Given the potential importance of recognizing head tremor, an examination maneuver that brings out an otherwise unappreciated or subclinical head tremor may have utility in both clinical and research settings. Here we report a clinical phenomenon that we have observed repeatedly in ET patients in clinical research settings; namely, a triggering and/or exacerbation of head tremor during or immediately after sustained phonation. To our knowledge it has not been reported previously nor has it been the subject of clinical study. Here we: 1) first report the phenomenon, 2) furnish several visual illustrations, 3) estimate its prevalence, and 4) analyze its clinical correlates.

Methods

As described [6], ET patients were screened and selected for enrollment in a clinical-epidemiologic study at Columbia University Medical Center (CUMC). By design, ET patients were recruited from two sources: patients from the clinical practice at the Center for Parkinson's Disease and Other Movement Disorders at CUMC or those who responded to a study advertisement in the International Essential Tremor Foundation newsletter. We excluded 58 potential enrollees whose clinical chart could not be located, 57 without full contact information, 17 who were deceased. In addition, 410 were excluded prior to enrollment because medical record review revealed the presence of other neurological diagnoses aside from or in addition to ET (e.g., Parkinson's disease, dystonia). Four hundred fifty-five screened patients were enrolled. Upon enrollment, each patient signed informed consent approved by the CUMC Institutional Review Board, including signed consent for their videotaped examination to be published. Three hundred eighty-six of 455 enrolled patients qualified for a diagnosis of ET based on previously published diagnostic criteria [6]; 69 patients (12 with Parkinson's disease, 16 with dystonia, 3 with myoclonus, 1 with psychogenic tremor, and 37 with tremor too mild to fulfill criteria for ET) were excluded. After enrollment, each was evaluated in person by a trained tester who administered several structural clinical questionnaires that elicited demographic and clinical data. Each patient also underwent a 20-minute standardized videotaped neurological examination, which included a detailed assessment of head (i.e., neck), jaw, voice, and limb tremors. Each patient had previously been assigned a total tremor score, a measure of action tremor severity in the arms (range = 0 – 36), according to methods described [6]. For head tremor, patients first were assessed while seated and facing the camera ("baseline"), then during sustained phonation ("ahh" and "eee" for 10–15 seconds each), and then immediately following sustained phonation. Head tremor was often also visible during much of the remainder of the 20-minute videotaped assessment as well (e.g. while drinking water from a cup, while using a spoon, while touching finger-to-nose). Comprehensive assessments for Parkinsonism and dystonia were also performed, and such patients were excluded. Videotaped examinations of all patients were reviewed by a senior movement disorders neurologist (E.D.L.), who noted the presence or absence of head tremor at any point during the videotape. There were 102 ET patients with head tremor.

The Tremor Research Group Essential Tremor Rating Assessment Scale (TETRAS®) was used to rate the severity of head tremor (0 – 3). This scale has been shown to have high inter- and intra-rater reliability, both overall and for head tremor [7]. For the current study, the 102 videos were reviewed by a movement disorders neurologist in training (B.A.W.), and a random subsample of 20% were co-reviewed by the senior neurologist. The inter-rater reliability was high (Pearson's correlation coefficient = 0.95, $p < 0.001$ for TETRAS® scores assigned by the two raters).

In each patient, head tremor was assigned a TETRAS® score at baseline (i.e., prior to sustained phonation), during sustained phonation, and immediately following sustained phonation. As the TETRAS® scores were sometimes insensitive to small levels of amplitude change, the rater also assigned a qualitative assessment of change (no change, increase in head tremor, decrease in head tremor) comparing baseline to period during and after immediately following sustained phonation. Statistical analyses were carried out using SPSS (version 19.0; Chicago, Illinois).

Results

Of 386 ET patients, 102 (26.4%) had head tremor at some point during the videotaped examination. Demographic and clinical characteristics of these 102 ET patients are shown (Table 1). Seventy (68.6%) of 102 patients exhibited a qualitative increase in head tremor amplitude during or immediately following sustained phonation. In 5 (4.9%) patients, head tremor would not have been detected without the voice activation maneuver, (i.e., it was not visible at any other point in the videotape aside from the period during/immediately following sustained phonation).

Table 2 shows results stratified according to baseline TETRAS® head tremor score. The more subtle the head tremor at baseline, the greater the likelihood that it would worsen in response to sustained phonation.

The mean increase in the TETRAS® head tremor score was 0.6 ± 0.5 points during sustained phonation and 0.5 ± 0.5 points immediately following sustained phonation. There were no instances in which TETRAS® score or qualitative head tremor amplitude decreased as a consequence of sustained phonation.

We examined the clinical correlates of this phenomenon. Head tremor qualitatively worsened in response to sustained phonation in 58 (73.4%) of 79 women vs. 12 (52.2%) of 23 men ($\chi^2 = 3.73$, $p = 0.05$). However, age, age of tremor onset, duration of tremor, and total tremor score did not predict responsiveness of head tremor to sustained phonation (Table 2).

Discussion

These results suggest that vocalization maneuvers, originally intended to evaluate voice tremor, can be useful to assess/release head tremor in ET patients. In a high proportion of patients in our cohort, head tremor was either triggered or amplified by sustained phonation. In 5%, head tremor would not have been detected without the voice activation maneuver (i.e., it was not visible at any other point in the videotape aside from the period during/immediately following sustained phonation). Whether the phenomenon we observed occurs in normal controls is an interesting question. As part of the same parent study, we enrolled 406 controls and all were videotaped, including 45 with no visible arm tremor (total tremor score = 0) and 361 with mild tremor that was within the range of normal (total tremor score in these 361 = 4.3 ± 2.7 , range = 1 – 15). In none of these was tremor noted to be triggered by sustained phonation. Head tremor is not reported to occur in patients with normal forms

of tremor, which is consistent with this finding. This maneuver, by triggering a head tremor, may be a useful diagnostic supplement in research/clinical settings where arm tremor is mild and the diagnosis (mild ET vs. enhanced physiological tremor) is otherwise ambiguous. Clinicians and clinical researchers may wish to incorporate this maneuver into their examination regimen. One setting in which this maneuver may be helpful is genetic studies; in these studies, one is often examining younger family members with mild tremor of short duration. The detection of head tremor is usually a clear indicator that the correct diagnosis is mild/early ET rather than enhanced physiological tremor.

The head tremor of ET is thought to be postural. A recent study of patients with head tremor found that >90% of ET patients had head tremor that resolved in a supine (resting) position, while head tremor persisted at rest in 68% of spasmodic torticollis patients [9]. It would be interesting to observe ET patients with head tremor performing the voice activation maneuvers while supine to see if sustained phonation can trigger head tremor in an otherwise resting state. Perhaps the condition of sustained vocalization can be thought of as a further departure from the resting state, providing one possible mechanism for this phenomenon. Other possible mechanisms include alteration of sensory feedback loops during and/or after phonation, as well as head tremor being elicited or worsened as an overflow phenomenon.

It is notable that women in this study were 1.4 times more likely than men to exhibit a worsening of head tremor in response to sustained phonation (with 73% [women] and 52% [men] worsening, respectively). It is known that head tremor is more prevalent in women than men with ET. Hubble et al. [1] reported that the prevalence of head and/or voice tremor was twice as high in women than men with ET, although the outcome measure included either type of tremor. In community-based studies of ET patients, we reported that the prevalence of head tremor was four-fold [8] to six-fold [5] higher in women relative to men. Reasons for this are unclear; differential influence on ET phenotype by sex hormones is possible. The 1.4-fold gender difference in the prevalence of voice-activated head tremor is not sufficient to explain the overall 2 – 6 fold gender discrepancy in the prevalence of head tremor.

It is possible that the sustained phonation maneuvers cause physiologic or psychological stress in subjects, and that this stress contributes to the mechanism by which head tremor is amplified. However, we did not observe any discomfort or stress in study participants; many were in fact amused by performing voice activation. With regard to possible medication effect in this setting: 35 (34.3%) of the 102 head tremor patients were taking beta blockers and 67 (65.7%) were not at the time of examination. Head tremor worsened in 26 of the 35 patients (74.3%) on beta blockers, and in 44 of the 67 (65.7%) who were not, a difference which was not significant (Fisher's $p = 0.51$).

The effect of the maneuver was most prevalent in patients with the mildest head tremors at baseline, which is not surprising. A finite mechanism would be expected to have less of a relative effect on a severe head tremor, and even an equivalent increase in amplitude would be more difficult for the examiner to detect.

It is known that the majority of patients with ET do not seek medical attention, so the phenomenology that was seen may not be present to an equal extent in the overall ET population. This is a limitation of this study. Study strengths include the uniform approach to patient examinations and the attempt for the first time to systematically observe, quantify, and study the clinical correlates of this clinical phenomenon.

In summary, sustained voice activation is a useful examination maneuver that may elicit or amplify head tremor in a considerable percentage of ET patients. This observation may have

diagnostic value in some settings and could provide clues to underlying mechanisms of postural head tremor in this disease.

Patient 1 exhibited no head tremor at baseline (TETRAS® head tremor score: 0). Seven to eight seconds into sustained phonation, a mild head tremor of < 0.5 cm amplitude emerged (TETRAS® head tremor score: 1). Sustained phonation was the only maneuver in the 20-minute videotaped examination that elicited head tremor in this patient. Patient 2 had no head tremor at baseline (TETRAS® head tremor score: 0). Towards the very end of sustained phonation, a subtle head tremor emerged (TETRAS® head tremor score: 0). Sustained phonation was the only maneuver in the 20-minute videotaped examination that elicited head tremor in this patient. Patient 3 exhibited a baseline head tremor of < 0.5 cm amplitude (TETRAS® head tremor score: 1). Immediately following sustained phonation, the amplitude transiently increased to greater than 0.5 cm (TETRAS® head tremor score: 1). Hence, the first two patients' head tremor was triggered by sustained phonation; the third patient's head tremor was exacerbated by sustained phonation.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Table 1

Demographic and clinical characteristics of 102 ET patients with head tremor

Age (years)	70.9 ± 11.6
Women	79 (77.5)
Age at tremor onset (years) ^a	46.2 ± 21.3
Duration of tremor (years)	24.5 ± 19.5
Family history of ET ^a	29 (28.4)
Total tremor score (range 0 – 36)	18.3 ± 9.1
Take daily medication for tremor	52 (51.0)
Take daily or as-needed medication for tremor	57 (55.9)
Head tremor by history	78 (76.5)
Head tremor on examination	102 (100)
Voice tremor by history	37 (36.3)
Voice tremor on examination	49 (48.5)
Jaw tremor on examination	20 (19.6)

All values are number (percentage) or mean ± SD.

^aMissing data on one or more participants.

ET, essential tremor.

Table 2

Head tremor response to sustained phonation

	Head Tremor Worsened		
	Yes	No	
Continuous Variables			
Age (years)	72.1 ± 11.4	68.2 ± 11.9	t = 1.56, p = 0.12
Age at tremor onset (years)	48.2 ± 20.5	41.7 ± 22.8	t = 1.43, p = 0.16
Duration of tremor (years)	23.5 ± 18.6	26.5 ± 21.5	t = 0.71, p = 0.48
Total tremor score (range 0 – 36)	18.7 ± 8.8	17.3 ± 9.9	t = 0.72, p = 0.47
Categorical Variables			
Gender			
Women	58 (73.4)	21 (26.6)	$\chi^2 = 3.73$, p = 0.05
Men	12 (52.2)	11 (47.8)	
Voice tremor on examination			
Present	33 (67.3)	16 (32.7)	$\chi^2 = 0.17$, p = 0.68
Absent	37 (71.2)	15 (28.8)	
Jaw tremor on examination			
Present	16 (80.0)	4 (20.0)	$\chi^2 = 1.49$, p = 0.22
Absent	54 (65.9)	28 (34.1)	
Embarrassed by head tremor			
Yes	48 (69.6)	21 (30.4)	$\chi^2 = 0.09$, p = 0.77
No	22 (66.7)	11 (33.3)	
Baseline TETRAS® Head Tremor score ^a			
0	23 (100) ^b	0 (0)	$\chi^2 = 25.07$, p = 0.001
1	39 (70.9)	16 (29.1)	
2	8 (34.7)	15 (65.2)	
3	0 (0)	1 (100)	

All values are mean ± SD or number (row percentage).

^aTETRAS® head tremor subscore while sitting and inactive:

0 (no head tremor); 1 (amplitude < 0.5 cm); 2 (amplitude 0.5 – 2.5 cm); 3 (amplitude 2.5 – 5 cm).

^bTwenty-three patients had no baseline head tremor, but 18 of these has head tremor at some other point during the videotaped examination.