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Isolated Head Tremor: Part of the Clinical Spectrum of Essential Tremor? Data from Population-Based and Clinic-Based Case Samples

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Abstract

Essential tremor (ET) still remains a clinical diagnosis. Nonetheless, it is misdiagnosed in 30 - 50% of cases. There are a number of areas of diagnostic uncertainty. One of these is isolated head tremor, on which published data are limited and at variance. We studied the prevalence of isolated head (i.e., neck) tremor in ET in two population-based studies (Turkey and New York) and a large clinical sample (New York); these 583 ET cases all received the same detailed tremor examination. Head tremor with mild arm tremor occurred in a very small percentage of cases in each sample (1.9% - 3.1%, overall 2.7%). Nearly all of these were women. Head tremor in the complete absence of arm tremor was not observed in any cases (0.0%). These clinical data may be of value to clinicians in practice settings and researchers in phenotyping efforts in the emerging field of ET genetics.

Keywords

Essential tremor; clinical; head tremor

Introduction

Head (i.e., neck) tremor is among the most commonly-noted clinical features of essential tremor (ET),¹⁻⁴ in contrast to Parkinson's disease (PD), in which it is rare.⁵⁻⁶ While tremor typically

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Author Contributions

Elan D. Louis: Research project conception, organization and execution; statistical analyses design and execution; manuscript writing (writing the first draft and making subsequent revisions).

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spreads from the hands to the head,^{2, 3} patients on occasion complain of isolated head tremor. ET remains a clinical diagnosis. Yet it is misdiagnosed in 30 – 50% of cases,⁷ suggesting that it could be the most commonly misdiagnosed movement disorder. One area of diagnostic ambiguity is that of isolated head tremor. Unfortunately, published data are limited to brief comments in case series that are broadly focused on other issues.⁸⁻¹⁷ Many of these series are small (30 - 40 cases)⁸⁻¹¹ and none⁸⁻¹⁷ provide detailed individual-level case data on the relative severity of arm vs. head tremor. Furthermore, results are discrepant. While several series imply that isolated head tremor is not part of the phenotypic spectrum of ET (0% of cases),⁸⁻¹⁰ others suggest that it is a feature of every fifth or tenth ET case.¹²⁻¹⁷ Hence, clinicians are left to wonder whether isolated head tremor is part of the phenotypic spectrum of ET? In this study, we examined the prevalence of isolated head tremor in ET in two population-based studies and a large clinical series. Each study provided sufficient individual case data to evaluate whether head tremor occurred in the absence of other tremors. We also conducted a critical review of published data. It is hoped that these efforts will assist in clinical diagnosis, thereby lessening diagnostic mis-classification. We also anticipate that these data will be of use in phenotyping efforts in emerging genetic studies of ET.¹⁸

Methods

Overview

Three ET case samples were used.¹⁹⁻²¹ In each set of cases, the same neurological examination was performed by study neurologists. A tremor examination included one test for postural tremor and five for kinetic tremor (pouring, using spoon, drinking, finger-nose-finger, spiral) performed with each arm (12 tests total, with tremor ratings of 0 [none], 1 [mild], 2 [moderate], 3 [severe]), along with evaluations of head (i.e., neck), voice and jaw tremors. Neck tremor in ET was coded as present or absent and was distinguished from dystonic tremor by the absence of twisting or tilting movements of the neck, jerk-like or sustained neck deviation, or hypertrophy of neck muscles. Study neurologists assigned ET diagnoses using the same published diagnostic criteria (kinetic tremor rated ≥ 2 during at least 3 tests or head tremor, in the absence of PD or dystonia); diagnostic criteria have been published in detail for each study,¹⁹⁻²¹ and diagnostic gradations of definite, probable and possible ET were applied in each case.²² Cases signed written informed consent upon enrollment.

Description of population-based study in Turkey

A population-based study of the prevalence of ET was conducted in Mersin, Turkey.²⁰ As described previously,²⁰ the target study population consisted of 2,500 adults who represented 0.65% of the Mersin population ≥ 40 years old. The epidemiological survey used door-to-door interviews and examinations. Study neurologists performed the evaluations, as described above. The neurologists visited the 2,500 residents in their homes between July and December, 2002. There were 89 prevalent ET cases (47 [52.8%] definite ET, 17 [19.1%] probable ET, and 25 [28.1%] possible ET).²⁰

Description of population-based study in New York

The Washington Heights-Inwood Genetic Study of Essential Tremor (WHIGET) was a family study of ET in the Washington Heights-Inwood community in northern Manhattan, New York. WHIGET enrollment began in 1995 and was completed in 2000. The design of this population-based study has been described in detail.²¹ After enrollment, each participant (age ≥ 18 years) was examined, as described above. There were 106 ET cases (59 probands with ET, 33 of their relatives with ET, and 14 affected relatives of control probands),²¹ whose diagnoses were definite ET (37 [34.9%]), probable ET (44 [41.5%]), and possible ET (25 [23.6%]).

Description of clinical sample in New York

ET cases are being enrolled in an ongoing study of the environmental epidemiology of ET (2000 – present) at the Neurological Institute of New York, Columbia University Medical Center; the design of this study has been described in detail previously.¹⁹ Prior to enrollment, all cases were diagnosed with ET by their neurologist. After enrollment, each case (age ≥ 18 years) was examined, as described above.¹⁹ There are 388 ET cases (110 [28.4%] definite ET, 181 [46.6%] probable ET, and 97 [25.0%] possible ET).

Statistical Analyses

Chi-square tests were used to compare proportions across ET case samples.

Results

There were 583 ET cases across the three study samples. In each of the two population-based studies, the proportion of ET cases with head tremor was approximately 18% (Table 1); in the clinical sample it was higher (37.1%, Chi-square = 22.4, $p < 0.001$). The majority (62.5% - 73.7%) of ET cases with head tremor were women (Table 1). A smaller proportion of cases (12.3% – 21.1%, Table 1) had head tremor in the absence of voice or jaw tremor.

The prevalence of isolated head tremor depended on the definition that was used (Table 1). When defined initially as head tremor in the absence of either (1) voice or jaw tremor or (2) moderate arm tremor (i.e., arm tremor of at least moderate amplitude during one of twelve tasks), the prevalence was 2/89 (2.3%) in Turkey, 2/106 (1.9%) in the population-based study in New York, and 12/388 (3.1%) in the clinical sample (overall = 16/583, 2.7%). These 16 cases were nearly all women (14/16, 87.5%). Each of the 16 cases had mild kinetic arm tremor on one or more test, with most having mild tremor on several tests (mean = 5.3, median = 5, range = 1 – 10 tests, Table 2). When isolated head tremor was defined more stringently as head tremor in the absence of voice, jaw or any arm tremor (i.e., tremor score = 0 on all 12 tests), then the prevalence was 0.0% in each of the three case samples.

There were 192 ET cases who were taking daily anti-tremor medication (3 in Turkey, 5 in the population-based study in New York, and 184 in the clinical sample in New York). We repeated the analyses, excluding these 192 cases, and the results were similar in the remaining 391 cases (overall initial prevalence of isolated head tremor = 11/391 [2.8%], and 0/391 [0.0%] when more stringently defined).

Discussion

When defined most stringently as head tremor in the complete absence of other clinically-observable tremor, the prevalence of isolated head tremor in ET was 0.0%. While no cases had head tremor in the complete absence of arm tremor, 2.7% had head tremor in the presence of mild arm tremor, with most of these cases having mild arm tremor on multiple tests.

Critchley²³ listed the 13 most common groupings of muscular involvement in ET; isolated head tremor was not among them. He also described the typical pattern of spread of tremor (arms to head), but not the converse. Similarly, in a study on “essential tremor variants”, Koller et al.⁹ described isolated voice, chin or tongue tremors, but not isolated head tremor.

Published data are limited to brief remarks in case series that are broadly focused on other issues. In this literature, (1) isolated head tremor was either not observed at all, (2) its presence in a very small subsample of cases was likely due to torticollis, or (3) it is not clear whether head tremor occurred with complete absence of arm tremor. In treatment-based settings, the proportion of ET cases without arm tremor ranges from 0.0% - 6.9%. Thus, in a study in Benin,

Nigeria, 0/35 (0.0%) patients had isolated head tremor.¹⁰ Two studies in England indicated that 0/34 (0.0%)⁸ and 0/20 (0.0%)¹⁶ index cases and 0/135 (0.0%)¹⁶ affected relatives had isolated head tremor. Similarly, in studies of children, isolated head tremor occurred in 0/3924 and 0/19.²⁵ Another English study indicated that 1/42 ET cases had mild titubation without arm tremor; however, that patient also had torsion spasm (dystonia),¹¹ indicating that the diagnosis was dystonia rather than ET. Similarly, a study in the United States¹³ noted 24/350 (6.9%) with isolated head tremor,¹³ yet 165 (47.1%) cases also had dystonia,¹³ and the overlap between the two is not indicated. In a series in Bulgaria, approximately 4 – 5% had isolated head tremor; however, no data on torticollis or mild arm tremor were provided.¹⁴ In each of these studies, individual case data were not presented, so it is not clear whether head tremor occurred with complete absence of arm tremor.

There are few population-based data. In Papua, New Guinea, isolated head tremor occurred in as many as one in ten ET cases (16/175, 9.1%).¹² However, these cases had sustained contractions of the “sternomastoid and neck muscles” and head displacement (i.e., “simulated very closely the appearance of ... spasmodic torticollis”), indicating that the diagnosis was likely dystonia rather than ET.¹² In a study in rural Sweden, 3/210 (1.4%) cases had isolated head tremor, but none of these three were examined (i.e., examinations were reported by affected relatives).¹⁵ A study in rural Tanzania reported 13/65 (20.0%) had isolated head tremor; however, individual case data were not presented and it is not clear whether head tremor occurred with complete absence of mild arm tremor.¹⁷ In the population-based study in New York, we previously reported head tremor in 37 ET cases,²⁶ but included jaw tremor in our estimate of head tremor and did not report separately the proportion with isolated head tremor.

In the current study, a nearly identical proportion of cases in the two population-based studies had head tremor (18.0% in Turkey and 17.9% in New York) whereas the proportion of ET cases with head tremor in the clinical sample was approximately double that (37.1%). As can be seen from the data in Table 1, this difference is not due to age or gender. Rather, is likely due to the fact that clinical samples tend to ascertain a more self-selected group of cases with more severe and widespread tremor. Indeed, in some highly self-selected samples, the proportion of cases with head tremor can exceed 60%.²⁷

This study had limitations. We did not assess children,^{19–21} and one of our samples did not assess cases <40 years old.²⁰ A limitation of this study is that data on the severity and direction of tremor were not collected uniformly across studies and were not presented here. Yet strengths of this study are the use of both population-based and clinical data. Arm tremor in ET cases was assessed in detail in each study and individual level data were provided, thereby allowing us to both estimate the prevalence of isolated head tremor and examine the effects of using different definitions of arm tremor (e.g., none vs. mild) on this prevalence. Furthermore, patients with neck dystonia were systematically excluded.

In summary, head tremor with mild arm tremor occurred in a very small percentage of ET cases (1.9 - 3.1%, overall 2.7%), with most of these having mild arm tremor on multiple tests. Nearly all of these were women. However, head tremor in the complete absence of arm tremor was not observed in any of these cases (0.0%). Head tremor in the complete absence of clinically-observable arm tremor was not part of the clinical spectrum of ET in these three broadly-ascertained ET case samples nor is there compelling support for the presence of such patients in the published ET literature.

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Table 1

Data from three studies of essential tremor

	Population-based study in Turkey	Population-based study in New York	Clinical sample in New York
Total number of ET cases	89	106	388
Age	57.3 ± 11.6 yrs	69.8 ± 18.4 yrs	67.4 ± 15.3 yrs
Gender	42 (47.2%) women	63 (59.4%) women	200 (51.5%) women
ET cases with head tremor	16 (18.0% of 89)	19 (17.9% of 106)	144 (37.1% of 388)
Age	64.3 ± 11.3 yrs	69.6 ± 20.0 yrs	71.9 ± 12.1 yrs
Gender	10 (62.5%) women	14 (73.7%) women	101 (70.1%) women
ET cases with head tremor and no voice or jaw tremor	11 (12.3% of 89)	13 (12.3% of 106)	82 (21.1% of 388)
Age	65.7 ± 12.3 yrs	65.4 ± 22.2 yrs	70.2 ± 12.8 yrs
Gender	6 (54.5%) women	11 (84.6%) women	58 (70.7%) women
ET cases with head tremor, no voice or jaw tremor, and no arm tremor of moderate or greater amplitude during at least one of 12 tests	2 (2.3% of 89)	2 (1.9% of 106)	12 (3.1% of 388)
Age	52.5 ± 0.7 yrs	56.5 ± 26.2 yrs	63.3 ± 14.6 yrs
Gender	1 (50.0%) woman	2 (100%) women	11 (91.7%) women
ET cases with head tremor and no voice or arm tremor	0 (0% of 89)	0 (0% of 106)	0 (0% of 388)

In each cell, the number of cases (percentage of total cases) is provided, as well as their mean ± SD age and gender distribution.

Table 2

Kinetic tremor ratings in 16 ET cases who had head tremor in the absence of either (1) voice or jaw tremor or (2) moderate arm tremor

Case	Post D	Post ND	Pour D	Pour ND	Spoon D	Spoon ND	Drink D	Drink ND	FNF D	FNF ND	Spiral D	Spiral ND
Population-based study in Turkey												
1	1	1	1	1	1	1	1	1	1	1	1	1
2			1	1	1	1	1	1	1	1	1	1
Population-based study in New York												
1	1		1						1			
2	1	1								1		
Clinical sample from New York												
1						1				1	1	1
2										1		
3					1				1	1	1	1
4			1	1				1	1	1	1	1
5			1					1	1	1	1	1
6			1		1		1	1	1	1	1	1
7	1		1	1	1				1	1	1	1
8	1	1	1	1		1	1	1	1	1	1	1
9			1	1	1	1	1	1	1	1	1	1
10								1				
11			1	1				1	1	1	1	1
12				1		1		1	1	1	1	1

Post = postural tremor, Pour = pouring, Spoon = using spoon, Drink = drinking, FNF = finger-nose-finger, Spiral = drawing Archimedes spiral.

D = dominant arm, ND = nondominant arm

1 = Tremor rating of 1 (mild tremor).

Blank cells have tremor rating = 0 (no tremor).