Task specific writing tremor: series and outcomes

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Abstract

Objective: To describe a large series of subjects with task specific writing tremor
Background: Task specific tremors remain a controversial area, possessing some features similar to essential tremor and some similar to dystonia. Task specific tremor can occur with many occupational and repetitive tasks but writing tremor is probably the best to study because the majority of people write.

Methods: We evaluated 40 subjects currently actively seen in the Baylor College of Medicine Movement Disorders Clinic with task specific writing tremor (WT). The diagnosis was made by observing tremor only elicited by writing, without more than trace action tremor, or by a robust history of tremor only during writing for at least three years before tremor in any other setting. Patients in whom the major feature was dystonia (abnormal posturing of the hand) were not included.

Results: The age of tremor onset was highly variable, 45±18 years, and 24 (68%) were male; 27 were Caucasian, 8 were African-American, 4 were Hispanic, and 1 was Asian Indian. A family history of any tremor in a first degree relative was reported by 18 (45%), whereas three more (7.5%) had tremor in a second or third degree relative. No patient reported a family history of dystonia. In those patients who developed additional less specific tremor, the next tremor provoking action was eating (10), brushing teeth/shaving (4), typing (2), suture removal (1) and drafting (1). Five subjects reported that the next tremor was a rest tremor. Three of these rest tremors first occurred in the contralateral hand. Pharmacologic treatments of writing tremor, including alcohol, were generally poor, whereas botulinum toxin and VM DBS were relatively successful.

Conclusions: Compared to patients with “classic” essential tremor in our clinic, writing tremor patients were more likely African, more likely male, more refractory to tremor medications, and more likely to develop rest tremor without other overt Parkinsonian signs.

Introduction

Since its initial description, task specific tremors (TST) have remained a controversial area, possessing some features similar to essential tremor and some similar to dystonia. Task specific tremor can occur with many occupational and repetitive tasks but writing tremor (WT) is probably the best to study because the majority of people write, whereas occupational task specific tremors (i.e. golf, musicians, etc.) evaluations are strongly biased by the necessity of pursuing that activity. Furthermore WT is probably the most common TST, although no formal epidemiology exists. Organic writers cramp may overlap with WT, as a large percentage of these subjects have some tremor. This tremor, however, is usually low frequency, irregular, and position dependent. In contrast WT is higher frequency, more regular, and not associated with dystonia.

Results

Forty subjects (28 male, mean age at final visit 61.8±13.7 years) are included. Ethnic background was Caucasian (27) African (8), Hispanic (4), Asian Indian (1). The age of WR onset was highly variable (mean 45±17.6 years). At the final visit, 12 remained exclusively writing tremor. In those patients who developed additional less specific tremor, the next tremor provoking scenario was eating/drinking (10), brushing teeth/shaving (4), typing (2), suture removal (1) and drafting (1). Five subjects reported that the next tremor was a rest tremor and in the remaining five, the next tremor scenario was unknown or mixed. A family history of any tremor in a first degree relative was reported by 18 (45%), whereas three more (7.5%) had tremor in a second or third degree relative. The tremor type in family members was WT (6), general action tremor (1), Parkinsonism/rest tremor (8). There appear to be multiple tremor types in their families. Only three subjects from all of these family members reported to have tremor were personally examined by us. No subject reported a family history of dystonia.

On examination, 23 subjects (43%) had some postural tremor, but in 15 of these the greatest tremor was rated as a 1 out of 4, which is not inconsistent with physiologic tremor. In those with postural tremor, 9 were symmetric, 11 were of greater amplitude in the dominant (writing) hand, and 2 had a larger tremor in the contralateral hand. Interestingly 9 subjects had a rest tremor, which was exclusively ipsilateral (3), ipsilateral more than contralateral (3), or mostly contralateral (3). Five of these also had a re-emergent tremor, including two with only tremor of postural repose without other rest tremor. 3

Although this study cannot evaluate the prevalence of WT, the ratio of ET to WT in our database was 21:3:1 and the ratio of organic writers cramp (task specific writing dystonia) to WT was 7.4:1. Therefore WT is likely much less common than these other conditions.

In general, pharmacological treatment of WT is less successful than with ET. We used a crude 0-4 scale (0=no benefit, 1=mild but not satisfactory benefit, 2=moderate benefit, 3=marked benefit, 4=complete tremor cessation). Beta-blocker scored 0.76±0.77 (n=21), primidone was 0.90±0.88 (n=10), topiramate was 1.10±1.00 (n=10) botulinum toxin was 2.0±0.94 (n=10) and deep brain stimulation of the Vim thalamic nucleus was 0.0±0.0 (n=2). Only 2/14 subjects felt they were alcohol responsive; the majority of others have not drank alcohol.

Discussion

This is the largest and most phenomenologically detailed report on WT. Novel observations include the higher prevalence in African-Americans, a male predominance, an association with the development of rest tremor, sometimes presenting on the side opposite to the WT, and the assessments of progression to less task specific tremors. Furthermore these are the only series documenting treatment of botulinum toxin and deep brain stimulation (DBS) along with oral medications. Our results generally support more association with ET dystonia and are a robust response VM DBS. However, the lack of response to oral ET medications and unique phenomenology and ethnic backgrounds suggest that this is a singular disease.

Most physiologic assessments tend to support some association with WT of regular action tremor, as opposed to dystonia. PET studies of WT show increased activity of the cerebellum similar to ET. By contrast, motor cortex activation in writing tremor also includes the contralateral premotor area (area 6) and ipsilateral prefrontal area. Only patients with writing tremor show bilateral activation of the parietal lobule (area 40) with a more pronounced activation on the contralateral side. Transcranial magnetic stimulation (TMS) studies are not consistent with dystonia 6, 7 and reciprocal inhibition patterns are also inconsistent with dystonia. 2

The observed progression from WT to rest tremor without other PD signs appears novel but is not surprising. Recent studies confirm that some patients with ET develop clinical PD 8, and dystonia (usually in the leg) is a relatively common presentation heralding PD in young patients.

Our subjects generally responded most robustly to botulinum toxin injections and DBS of the Vim, both of which were more effective than oral ET medications. However, many are refractory to conventional treatments or less readily because the functional impact of WT is less than generalized action tremor. We excluded patients with writing dystonia, even if they had some tremor. This may be an artificial segregation, and account for our lack of dystonia association in this study. However, the tremor seen with writers cramp is usually very small amplitude, irregular and jerky, and position dependent. The WT in our group is a large amplitude very regular tremor phenomenologically dissimilar to writers cramp.

References