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Short communication

Sensory trick in upper limb dystonia

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ABSTRACT

Introduction: Sensory trick is a specific maneuver that temporarily improves dystonia that is usually observed in 44%–89% of patients with cranial-cervical dystonia and in 20% of patients with upper limb dystonia. This study aimed to assess the prevalence of sensory trick in a cohort of 37 patients with idiopathic adult-onset upper limb dystonia and to determine whether sensory trick can be a useful tool to distinguish dystonic and non-dystonic tremor.

Methods: Thirty-seven right-handed patients with idiopathic upper limb dystonia and disturbed handwriting and 19 patients with non-dystonic action tremor in the upper limb causing writing disturbances participated into the study. Patients were asked to write a standard sentence twice, before and after applying a standardized sensory trick (gently grabbing right wrist with his left hand). Readability of the two sentences was assessed by three observers blinded to diagnosis.

Results: Five/37 patients (13%) self-discovered ST over disease history, while performing the standardized trick maneuver improved handwriting in 14/37 patients (38%). Interobserver agreement on the effectiveness of sensory trick among the three observers yielded a kappa value of 0.86 ($p < 0.0001$). The standardized trick was effective in 8/19 patients with dystonic tremor (42%) and in 0/19 patients with non-dystonic tremor ($p = 0.003$).

Conclusion: The results of applying a standardized non-spontaneous trick demonstrated that, in upper limb dystonia, ST may be more frequent than usually observed. Effective sensory trick, when present, may be a hallmark of idiopathic dystonia. The lack of effective sensory trick may help to identify non dystonic upper limb tremor.

1. Introduction

Sensory trick (ST), also called “geste antagoniste”, is a specific maneuver that temporarily improves dystonia and emphasizes the role of sensory-motor integration in the pathophysiology of dystonia [1]. Typically, ST may vary from person-to-person and with dystonia localization. Some individuals may also manifest multiple tricks [1].

The prevalence of ST seems to be greater in cranial-cervical dystonia than in upper limb dystonia (ULD) (44%–82% vs. 20%) [2]. This variability may reflect pathophysiological differences among focal dystonias or, alternatively, patients with cranial-cervical dystonia may merely have a greater ease to self-discover ST than patients with ULD [3,4]. In patients with cervical dystonia (CD) and head tremor, cessation or decrease of dystonic tremor induced by ST may help to distinguish dystonic and essential head tremor [5]. Information on the

helpfulness of ST in differentiating dystonic and non-dystonic tremor in the upper limb is to date lacking.

In this study, we assessed a group of patients with ULD and disturbed handwriting for spontaneous ST. Thereafter, patients were instructed to apply a standardized trick during handwriting, and the frequency of its effectiveness was evaluated. Finally, we investigated whether performing the standardized trick aided in distinguishing ULD patients with dystonic tremor from non dystonic patients with upper limb action tremor.

2. Patients and methods

Patients and controls were recruited between July 2017 and February 2018 among consecutive outpatients attending the movement disorder clinics of the Department of Medical Sciences and Public

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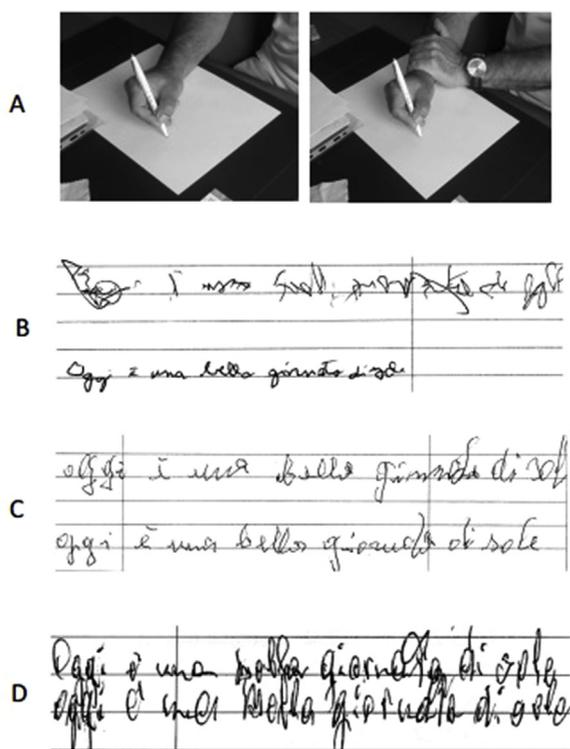


Fig. 1. Standardized sensory trick performed during handwriting (A); standard sentence, before (upper line) and after (lower line) applying the trick (B–D). Effective (B) and ineffective (C) trick in dystonic patients; ineffective trick in a patient with essential tremor (D).

Health, University of Cagliari, and the Department of Basic Medical Sciences, Neurosciences and Sense Organs, University of Bari, Italy. Inclusion criteria were the presence of idiopathic focal/segmental dystonia diagnosed according to published criteria [6]; dystonic involvement of the upper limb (with or without tremor) and handwriting disturbance; and age at dystonia onset > 18 years. Exclusion criteria were neuroleptic treatment in the six months before the onset of dystonia and other causes of secondary dystonia [7]. All patients with dystonia onset before 26 years were tested and proved negative for DYT1 mutation [1].

Dystonic patients were compared with control patients affected by non-dystonic action tremor in the upper limb causing dysgraphia. Action tremor was diagnosed according with Bhatia et al. [8] Patients and controls were asked to write an Italian standard sentence, “oggi è una bella giornata di sole” (today is a nice sunny day). Thereafter, subjects were asked to rewrite the sentence while performing a standardized trick, that is gently grabbing the right wrist with their left hand (Fig. 1A). The readability of the sentence was judged by three observers who were blinded to diagnosis. ST was considered to be effective only when all three observers agreed on the improvement in readability (Fig. 1B–D). Patients were under botulinum toxin treatment and handwriting assessment was performed three to four months after the last treatment.

Table 1

Demographic and clinical features of patients with upper limb dystonia and tremor vs. non-dystonic patients with upper limb action tremor.

	Dystonic patients with tremor (n. 19)	Non-dystonic patients with tremor (n. 19)	P
N. of men (%)	12 (63%)	12 (63%)	1
Mean age (years) ± SD	63.2 ± 9.4	71.9 ± 9.5	0.02
Mean age at disease onset (years) ± SD	42.1 ± 14	64.1 ± 9.7	p < 0.0001
N. of patients with focal dystonia (%)	10	NA	NA
N. of patients with effective sensory trick (%)	8 (42%)	0	0.003
Mean disease duration (years) ± SD	21.4 ± 9.8	7.5 ± 6.5	p < 0.0001

Data were expressed as mean and standard deviation unless otherwise indicated. Groups were compared by chi-squared test, Fisher's test, and Mann-Whitney *U* test as appropriate. Interobserver agreement on the effectiveness of ST was assessed by kappa statistics. Statistical significance was set at the 0.05 level. Sensitivity was the proportion of subjects who showed an effective standardized ST and had dystonic tremor; specificity was the proportion of subjects suffering from non-dystonic tremor who screened negative on performing standardized ST.

3. Results

During the study period, 37 right-handed ULD patients were eligible for and accepted to participate into the study. There were 18 men and 19 women aged 60.3 years (SD, 13.2). Mean age at dystonia onset was 43.7 years (SD, 12.9) and mean disease duration was 18.1 years (SD, 10.1). Focal ULD was diagnosed in 22 patients, ULD as part of a segmental/multifocal dystonia was diagnosed in 15 patients. Upper limb dystonic tremor was observed in 19/37 patients.

Five out of 37 patients (13%) self-discovered ST over their disease history: ST was still effective at study time in 4/5. Performing the standardized trick improved handwriting in 14/37 patients (38%). Interobserver agreement on the effectiveness of ST among the three observers yielded a kappa value of 0.86 ($p < 0.0001$). Patients who showed an effective trick and those who did not were similar for age (63.4 ± 11.9 vs. 58.2 ± 13.8 years, $p = 0.14$), sex (9 men and 5 women vs. 9 men and 14 women, $p = 0.14$), age at disease onset (42.9 ± 14.1 vs. 44.3 ± 12.3 years, $p = 0.38$), disease duration (20.6 ± 10 vs. 16.1 ± 10 years, $p = 0.12$), focal/segmental distribution of dystonia (8/6 vs. 14/9, $p = 0.8$), and presence of dystonic tremor (8/6 vs. 11/12, $p = 0.6$).

To see whether ST distinguished between dystonic and non-dystonic upper limb action tremor, we compared the 19 patients with dystonic tremor to 19 right-handed patients with non-dystonic action tremor in the upper limb (10 patients with essential tremor and 9 patients with parkinsonian action tremor -seven also had rest tremor-). Patients with dystonic and non-dystonic tremor did not significantly differ for sex but differed for age, age at disease onset, disease duration, and effectiveness of ST (Table 1). ST was effective in 42% of patients with dystonic tremor and ineffective in all patients with action tremor due to essential tremor or Parkinson's disease. Overall, ST differentiated dystonic and non-dystonic tremor with 42% sensitivity and 100% specificity.

4. Discussion

In this sample of patients with ULD and disturbed handwriting, 13% of patients self-discovered ST over their disease history. When patients were instructed to apply a standardized trick, the frequency of effective ST raised to 38%. In addition, we observed that ST distinguished dystonic and non-dystonic tremor with 42% sensitivity and 100% specificity. The lack of effect of the standardized ST in the non dystonic tremor group would support the standardized maneuver as a ST rather than a mere stabilization of tremor.

The rate of self-discovered ST observed in this sample of ULD patients was close to the 20% frequency of effective ST reported by a few prior publications [9,10]. The results of applying a standardized non-

spontaneous trick to the entire patient population demonstrated that, in ULD, ST may be potentially more frequent than usually observed. The 38% frequency of effective standardized trick found in our sample is close to the lower limit of the range of ST frequency reported in cranio-cervical dystonia [3,4]. Of note, we tested only one type of standardized trick whereas ST may have variable phenomenology. It is possible that other types of ST might have been effective in ameliorating ULD in the patients who did not respond to our standardized manoeuvre. Therefore, the true frequency of effective ST in this sample might have been underestimated. The aforementioned observations would suggest that the apparent variable frequency of spontaneous, self-discovered ST observed in patients with CD, BSP, and ULD does not reflect pathophysiological differences but probably the variable ease/difficulty of patients with different focal dystonias to self-discover ST.

The presence of an effective ST did not correlate with any demographic/clinical variable. This is at variance with some studies reporting an inverse correlation between rate of ST and dystonia duration [11,12], even though other studies did not find any association between ST and disease duration or severity [5]. It should also be noticed that previous studies assessing the relationships between ST and dystonia duration referred to spontaneous self-discovered tricks whereas we studied a standardized, non-spontaneous trick manoeuvre.

ST correctly identified only 42% of patients with dystonic tremor and 100% of patients with non dystonic tremor. Overall, our findings confirm that ST, when present, may be a hallmark of idiopathic dystonia; the 100% specificity also indicates that excluding an effective ST may help to identify non dystonic upper limb tremor.

This study may have strengths and limitations. Recruiting consecutive ULD patients in a multicentre setting gave a case series resembling the general population of cases. Although ULD patients were relatively old at study time, ULD and handwriting disturbance developed at a younger age, similar to the general population of patients with ULD. By contrast, patients with writing tremors due to essential tremor and Parkinson's disease were older than ULD, as it is commonly observed [6,8]. Assessors were unaware of the study hypotheses and blinded to patient's diagnosis. However, assessors were not blinded to ST and the use of a subjective measure of motor performance might have biased judgement about the outcome of the handwriting task. To minimize such a bias, however we considered sensory trick to be effective only when all three observers agreed on the substantial improvement in readability. Furthermore, our satisfactory reliability data indicated that there is validity in an approach that is usually used in the clinic. Control group was heterogeneous to reflect the heterogeneity of clinical conditions other than ULD possibly affecting handwriting. The difference in age and disease duration between dystonic and non-dystonic tremor patients may have affected the outcome, as there is a trend towards an increased number of STs in patients with lower dystonia duration. However, the greater disease duration in the dystonic tremor group does not support such a bias. Finally we were aware that in parkinsonian patients associated bradykinesia and rigidity rather than action tremor may affect hand motor performance. Nevertheless we included these patients in the control group because in clinical practice parkinsonian action tremor may contribute to the differential diagnosis of upper limb dystonic tremor.

In conclusion, the effectiveness of a standardized ST in a consistent proportion of ULD patients points towards recommending to ULD patients to systematically identify and elicit STs, a clinical clue that supports the dystonic nature of upper limb tremor as compared to essential tremor or other types of tremors. Despite of the transitory nature of benefit, ST may be especially important since botulinum toxin is not always effective in task specific ULD. In addition, identifying a trick may also lead to the fabrication of effective mimicry devices.

Ethical standards

The study was approved by institutional review boards (approval No. 14178, University of Bari). Informed consent was obtained from all subjects according to declaration of Helsinki. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

Conflicts of interest

Declaration of interest: none. All authors have approved the final article.

Appendix

Author Roles

1. Research project:
 - A. Concept: Dagostino S, Ercoli T, Defazio G
 - B. Organization: Gigante AF, Pellicciari R, Fadda L
 - C. Execution: Dagostino S, Ercoli T, Gigante AF, Pellicciari R
2. Statistical Analysis:
 - A. Design: Dagostino S, Ercoli T, Defazio G
 - B. Execution: Fadda L, Defazio G
 - C. Review and Critique: Dagostino S, Ercoli T, Gigante AF

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