Published in final edited form as:

Mov Disord. 2011 September ; 26(11): 2107–2109. doi:10.1002/mds.23827.

Extreme Task-Specificity in Writer's Cramp

Ejaz A. Shamim, MD, MS^{1,2}, Jason Chu, BS, MS^{1,3}, Linda H. Scheider, BS^{1,4}, Joseph Savitt, MD, PhD⁵, H. A. Jinnah, MD, PhD⁶, and Mark Hallett, MD¹

¹ Human Motor Control Section, National Institute of Neurological Disorders and Stroke (NINDS), National Institutes of Health (NIH), Bethesda, MD, 20892

² Kaiser Permanente Mid-Atlantic Permanente Medical Group, Department of Neurology, 5100 Auth Way, Suitland, MD 20746

³ Neurosurgery Resident, Emory University School of Medicine, Atlanta, GA

⁴ Virginia Commonwealth University School of Medicine, Richmond, VA

⁵ Department of Neurology, Johns Hopkins University School of Medicine, Baltimore, MD

⁶ Departments of Neurology, Human Genetics & Pediatrics, Emory University, Atlanta GA

Abstract

Correspondence: Ejaz A. Shamim MD, MS, Human Motor Control Section, Medical Neurology Branch, National Institute of Neurological Disorders and Stroke, National Institutes of Health (MSC 1428), 10 Center Drive Room 7D37, Bethesda, MD 20892 USA, shamime@mail.nih.gov.

Author's Roles

Ejaz A. Shamim (1A, 1B, 1C and 3A and 3B)

Jason Chu (1B, 1C and 3B)

Linda Scheider (1B, 1C, and 3B)

Mark Hallett (1A, 1B, and 3B)

Joseph Savitt (1A, 1B, and 3B)

H A Jinnah (1A, 1B, and 3B)

Disclosures

Dr. Shamim completed speakers' training for Allergan, Inc. and has participated as the site PI for an Allergan-sponsored clinical trial, CD-PROBE. He has received research support from the NIH/NINDS Intramural program and the Kinetics Foundation.

Dr. Savitt receives research grant support from Solvay Pharmaceuticals and Molecular Biometrics

Dr. Jinnah has served as a consultant for Savient Pharmaceuticals, and Psyadon Pharmaceuticals. He has previously received grants from the NIH, the Lesch-Nyhan Syndrome Children's Research Foundation, the Tomorrow Foundation, and the Bachmann-Strauss Dystonia & Parkinson's Foundation. He currently is funded by grants from the Dystonia Medical Research Foundation and the NIH (NS067501, HD53312, NS061349 and NS03592).

Dr. Hallett serves as Chair of the Medical Advisory Board for and receives honoraria and funding for travel from the Neurotoxin Institute. He may accrue revenue on US Patent #6,780,413 B2 (Issued: August 24, 2004): Immunotoxin (MAB-Ricin) for the treatment of focal movement disorders, and US Patent #7,407,478 (Issued: August 5, 2008): Coil for Magnetic Stimulation and methods for using the same (H-coil); in relation to the latter, he has received license fee payments from the NIH (from Brainsway) for licensing of this patent. Dr. Hallett's research at the NIH is largely supported by the NIH Intramural Program. Supplemental research funds came from the US Army via the Henry Jackson Foundation, Ariston Pharmaceutical Company via a Cooperative Research and Development Agreement (CRADA) with NIH, and the Kinetics Foundation via a Clinical Trials Agreement (CTA) with NIH. Dr. Hallett serves as Chair of the Medical Advisory Board of the Benign Essential Blepharospasm Foundation and Chair of the Medical Advisory Board of the International Essential Tremor Foundation. He serves on editorial advisory boards for Clinical Neurophysiology, Brain, Acta Neurologica Scandinavica, Journal of Clinical Neurophysiology, Italian Journal of Neurological Sciences, Medical Problems of Performing Artists, Annals of Neurology, Neurology and Clinical Neurophysiology, The Cerebellum, NeuroRx, Current Trends in Neurology, Faculty of 1000 Medicine, Brain Stimulation, Journal of Movement Disorders (Korea), and is Editor-in-Chief of World Neurology. He receives publishing royalties from Blackwell Publisher, Cambridge University Press, Springer Verlag, Taylor & Francis Group, Oxford University Press, John Wiley & Sons, Massachusetts Medical Society, Wolters Kluwer, and Elsevier. He has received honoraria for lecturing from Columbia University, the Parkinson and Aging Research Foundation, University of Maryland, University of Wisconsin, State of New York, and University of Navarra. He has received a fee from PGA Tour Inc.

Background—Focal hand dystonia may be task-specific as is the case with writer's cramp (WC). In early stages, the task-specificity can be so specific that it may be mistaken for a psychogenic movement disorder.

Methods—We describe four patients who showed extreme task specificity in WC. They initially only had problems writing either a single letter or number. Although they were largely thought to be psychogenic, they progressed to typical WC.

Conclusions—Early recognition of this condition may provide an opportunity for early initiation of treatment.

Keywords

Dystonia; Movement Disorders; Clinical Neurology

Introduction

Dystonias are characterized by excessive involuntary contractions of muscles leading to abnormal posturing. Dystonias that affect discrete body parts, such as focal hand dystonia (FHD) may be task-specific. Typically FHD occurs in individuals who repeatedly perform very precise tasks for prolonged periods of time usually under stressful conditions. As a result, musicians, typists, dart throwers, billiard players and others can be affected with life-altering dysfunctions. Animal models have shown the importance of repetitive activities in the development of WC (1). Hereditary factors are also important (2).

Task specificity in FHD is poorly understood. All other aspects of hand function are usually unaffected and the neurological examination is normal. Since the initial recorded description of task specificity in FHD by Sir Charles Bell (3) and the description of WC by Gowers (4) in the 1800s, this specificity has puzzled clinicians. The unusual task-specificity led to it being classified as a psychogenic movement disorder until the 1980s when it was recognized together with other dystonias as an organic entity (5). Here, we describe four patients who had "extreme task specificity" as an early manifestation of WC. Three of the four initially were thought to have a psychogenic movement disorder.

Patient #1

A 55-year-old right-handed Caucasian male presented with a one-year history of difficulty signing his name. He signed his name 200 to 1000 times per day for the past several years under stressful conditions where deadlines had to be met and employees and bills had to be paid. His initial symptom was difficulty with initiating his signature, which starts with the letter "J" (see video 1). Only in the context of signing his name was this difficult. Initially, when he printed or wrote this letter in other contexts, there were no problems. Later, he began to have trouble with the letter "J" even in other contexts, and a diagnosis of WC was more obvious.

On examination, he used excessive pressure while writing and had mirror movements with his right hand when he wrote with the left; his neurological examination was otherwise normal. Using a stamp to sign his name has been very helpful.

Patient #2

A 49-year-old left-handed Caucasian male presented with a three year history of progressive difficulty writing the number "7." He only had trouble making the vertical line down. This progressed to involve the number "9," and the letter "C," in the same manner. He felt a cramping sensation in the forearm while making these vertical lines. Writing the number "1"

was not a problem. He later had difficulty with all aspects of writing (see video 2). He was a mechanic for over 20 years. Three years ago, he started carving birds for 2 hours daily. His free time was spent carving, which required him to make very precise short vertical movements with his hands using the right index finger and thumb to stabilize the carving tool. He had to be gentle yet forceful when making these repetitive movements, of the arm, hand, wrist, and finger. Ultimately, he had difficulty carving.

His neurological examination showed mirror movements in his left hand while writing with his right hand. He held the pen in an awkward position with fingers and wrists flexed (see video 2). He stretched his hands frequently while writing.

He tried medicines without benefit including primidone, gabapentin, propranolol or carbidopa/levodopa. His was told by physicians, neurologists and psychiatrists that the ailment was psychological. About 2 years after the onset of symptoms he was diagnosed with WC. Currently, using a thick pen helps alleviate the cramping sensation and carving remains a problem.

Patient #3

A 52-year-old right-handed Caucasian woman presented with a 10 year history of trouble writing the letters “m” and “n”. She was an accounting executive and her job required a great deal of writing with lots of stress and frequent deadlines. She worked 70 to 80 hours weekly. The writing later affected all letters and numbers and she had difficulty writing even short thank-you notes and frequently broke pens because of the amount of pressure she exerted. She never had difficulty writing on a black board. Her ability to play the piano was unaffected. She had normal electrodiagnostic studies and MRI of the brain and cervical spine. She saw many physicians and her symptoms were considered a manifestation of underlying emotional stress, so she stopped working.

Her neurological examination was normal. With writing, her thumb, fingers and wrist flexed and she felt a cramping sensation in her forearm (see video 3). She used excessive pressure when writing. With continued writing, the pen fell out of her hand. She had mirror movements with her right hand as she wrote with her left.

Patient #4

A 52-year-old right-handed Caucasian man presented with an eight year history of trouble writing. He was an accountant and cartographer for the National Guard. He participated in daily drills where he had to make a dot on a map and circle the dot and then write a couple of words where bombing practice was to occur. Although these were just practice drills, they were very tense situations. He started having difficulty making the dot. He would try to make a dot but could not place the pen on the map. His superiors told him the problem was stress-related. He soon developed difficulty writing words. He then sought the help of physicians, psychiatrists, and orthopedic surgeons without any answers. He was also a banjo player and subsequently noticed that his fingers would curl while playing. He was diagnosed with FHD about 11 years after onset.

The patient’s neurological examination was notable for awkward posturing with hyper-extension at the wrist joint and fingers causing frequent change in his grip while writing (see video 4). With playing the banjo, his fingers curled and he was unable to extend them (see video 4). He had mirror movements with the right hand while he wrote with the left hand.

For several years, BTX helped but this later became ineffective. He has stopped playing the banjo and began typing.

Discussion

Although task specificity in focal dystonias is a well-known phenomenon, the nature of this specificity is not well understood. Because of the curious nature of task specificity, patients are sometimes thought to have a psychogenic problem, leading to significant frustration until a diagnosis is established. Only one of the four patients was diagnosed in a relatively short period of time. The other three went from one physician to the next until a diagnosis was established. In one case, it took more than 10 years. Early recognition can be life-altering (6), may decrease frustration in an already disheartened individual, and may allow the patient to function with appropriate treatment.

Some clinicians may argue that patient 1 may have writer's block which might be a psychological phenomenon, but WC seems more likely. Pressured writing and history of repetitive movement are seen in patients with WC. The development of dystonia in the right hand when he was asked to write with the left hand represents a phenomenon called "mirror dystonia" which is frequently seen in patients with WC. Jedynek et al reported that it was seen in 44% of the 65 patients they studied with WC (15). This patient represents a good example of how WC diagnosis can be confusing even for experts in the earliest stage of the disease.

In the etiology of FHD and WC, performing a very precise repetitive task for prolonged periods of time is a frequent trigger. Epidemiological studies in musicians who are required to perform very precise repetitive movements for prolonged periods of time under stressful conditions (7) have supported this notion. The importance of performing repetitive activity in patients with WC was recognized even in the earliest description of the disease in the late 1800s (4). All four of our patients performed repetitive activities for long periods of time. For patient 2, daily carving and mechanical activities may have triggered the FHD. None of our patients had affected family members.

Unusual task specificity can be seen in other focal dystonias and can be considered bizarre leading to a psychogenic diagnosis. Perhaps it was this bizarre exceptional specificity, which led to the descriptive term, "professional neuroses," which was later confused as a psychological phenomenon (8). Unusual task specificity can be seen in many focal dystonias (9, 10, 11, 12). With embouchure dystonia, trumpet players may begin with dystonia with certain range of notes, which later generalizes to all notes (13). The underlying mechanism leading to loss of specificity over time is not clear. Loss of surround inhibition in patients with FHD may lead to abnormal plasticity of other parts of the brain over time (14).

It is important for physicians and especially neurologists and psychiatrists to be wary of the fact that WC can start as a very task-specific problem involving only a single letter or number in patients performing repetitive writing or fine motor tasks during stressful situations. Early recognition can help allay frustration for patients and provide some explanation to an already disheartened individual.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

We thank D. Schoenberg for skillful editing. This endeavor was supported in part by the Intramural Program of the National Institutes of Health or NS067501.

References

1. Byl NN, Merzenich MM, Cheung S, Bedenbaugh P, Nagarajan SS, Jenkins WM. A primate model for studying focal dystonia and repetitive strain injury: effects on the primary somatosensory cortex. *Phys Ther.* 1997; 77:269–284. [PubMed: 9062569]
2. Tarsy D, Simon DK. Dystonia. *N Engl J Med.* 2006; 355:818–829. [PubMed: 16928997]
3. Bell, C. *The Nervous System of the Human Body.* Washington Duff Green; 1833.
4. Gowers, W. *A Manual of the Diseases of the Nervous System.* Philadelphia: P. Blakiston; 1888.
5. Sheehy MP, Marsden CD. Writer's cramp-a focal dystonia. *Brain.* 1982; 105 (Pt 3):461–480. [PubMed: 7104663]
6. Pullman S, Hristova A. Musician's dystonia. [Editorial]. *Neurology.* 2005; 64(2):186–187. [PubMed: 15668411]
7. Frucht SJ. Focal task-specific dystonia in musicians. *Adv Neurol.* 2004; 94:225–230. [PubMed: 14509677]
8. Marsden CD, Sheehy MP. Writer's cramp. *Trends Neurosci.* 1990 Apr; 13(4):148–539. [PubMed: 1692172] Sachdev P. Golfers' cramp: clinical characteristics and evidence against it being an anxiety disorder. *Mov Disord.* 1992; 7:326–332. [PubMed: 1484526]
9. Song IU, Kim JS, Kim HT, Lee KS. Task-specific focal hand dystonia with usage of a spoon. *Parkinsonism Relat Disord.* 2007
10. Scolding NJ, Smith SM, Sturman S, Brookes GB, Lees AJ. Auctioneer's jaw: a case of occupational oromandibular hemidystonia. *Mov Disord.* 1995; 10:508–509. [PubMed: 7565835]
11. Bonanni L, Thomas A, Scorrano V, Onofrij M. Task-specific lower lip dystonia due to mantra recitation. *Mov Disord.* 2007; 22:439–440. [PubMed: 17149705]
12. Ilic TV, Potter M, Holler I, Deuschl G, Volkmann J. Praying-induced oromandibular dystonia. *Mov Disord.* 2005; 20:385–386. [PubMed: 15580567]
13. Frucht SJ. Embouchure dystonia--Portrait of a task-specific cranial dystonia. *Mov Disord.* 2009; 15;24(12):1752–62.
14. Hallett M. Neurophysiology of dystonia: The role of inhibition. *Neurobiol of Dis.* 2011; 42 (2): 177–84.
15. Jedynak PC, Tranchant C, de Beyer DZ. Prospective clinical study of writer's cramp. *Mov Disord.* 2001 May; 16(3):494–9. [PubMed: 11391744]