

Video Abstracts

Dementia in Essential Tremor: A Visual Record

Maria Anna Zdrodowska^{1†}, Tess E. K. Cersonsky^{1†}, Arash Salardini¹, Stephanie Cosentino^{2,3} & Elan D. Louis^{1,4,5*}

¹ Department of Neurology, Yale School of Medicine, Yale University, New Haven, CT, US, ² Taub Institute for Research on Alzheimer's Disease and the Aging Brain, College of Physicians and Surgeons, Columbia University, New York, NY, US, ³ Department of Neurology, College of Physicians and Surgeons, Columbia University, New York, NY, US, ⁴ Department of Chronic Disease Epidemiology, Yale School of Public Health, Yale University, New Haven, CT, US, ⁵ Center for Neuroepidemiology and Clinical Neurological Research, Yale School of Medicine, Yale University, New Haven, CT, US

Abstract

Background: Research studies have shown an association between essential tremor (ET) and dementia, although dementia in ET is not often regarded as a clinically important issue.

Phenomenology Shown: We present three tangible visual records of patients with ET who have developed concurrent, comorbid dementia.

Educational Value: ET is a risk factor for dementia. This non-motor feature of the disease has substantial effects on the lives of patients and their families.

Keywords: Essential tremor, dementia, Clinical Dementia Rating, cognitive decline

Citation: Zdrodowska MA, Cersonsky TEK, Salardini A, Cosentino S, Louis ED. Dementia in essential tremor: A visual record. Tremor Other Hyperkinet Mov. 2019; 9. doi: 10.7916/d8-eh2t-5m82

*To whom correspondence should be addressed. E-mail: elan.louis@yale.edu

†These authors contributed equally to critical revision of the manuscript and video.

Editor: Elan D. Louis, Yale University, USA

Received: January 7, 2019 **Accepted:** February 1, 2019 **Published:** March 6, 2019

Copyright: © 2019 Zdrodowska et al. This is an open-access article distributed under the terms of the Creative Commons Attribution–Noncommercial–No Derivatives License, which permits the user to copy, distribute, and transmit the work provided that the original authors and source are credited; that no commercial use is made of the work; and that the work is not altered or transformed.

Funding: NINDS R01NS086736.

Financial Disclosures: None.

Conflicts of Interest: The authors report no conflicts of interest.

Ethics Statement: This study was performed in accordance with the ethical standards detailed in the Declaration of Helsinki. The authors' institutional ethics committee has approved this study and all patients have provided written informed consent. All patients that appear on video have provided written informed consent; authorization for the videotaping and for publication of the videotape was provided.

An increasing number of independent studies from around the world (Italy, first study in 2001; USA; Spain; Germany; Turkey; Korea) are now showing that, on average, essential tremor (ET) cases have poorer cognitive performance than age-matched controls. These cognitive changes are not strictly subclinical and they have been associated with greater functional difficulty. There is also an emerging understanding that the cognitive problems in ET patients are not necessarily static, can become severe, and that the rate of progression seems to be above and beyond that expected in age-matched people without ET. A cross-sectional epidemiological study reported an association between ET and mild cognitive impairment (MCI) (odds ratio 1.57), and several prospective, population-based, epidemiological studies, one in Madrid and the other in New York, demonstrated an association between ET and both prevalent and incident dementia. In these

studies, 11.4–25.0% of ET cases had prevalent dementia vs. 6.0–9.2% of controls. Furthermore, in both studies, the risk of incident dementia at follow-up was higher among individuals with baseline ET vs. those without baseline ET (relative risk 1.64–1.89).¹ A third study also found that a subset of ET cases had a similar increased risk of dementia (relative risk 2.1). ET patients with cognitive problems are *not simply* older adults with ET that happen to also have dementia (i.e., a chance co-occurrence).² Studies reporting the presence of dementia in ET compare ET cases with age-matched controls, and show a higher prevalence of dementia in ET, thereby indicating that dementia is *disease-linked* and not merely *age-linked*. By analogy, the cognitive problems in Parkinson's disease (PD) are not just “old people” with MCI or dementia who happen to also have PD; there is both clinically and pathologically a separable PD cognition entity. The same is likely

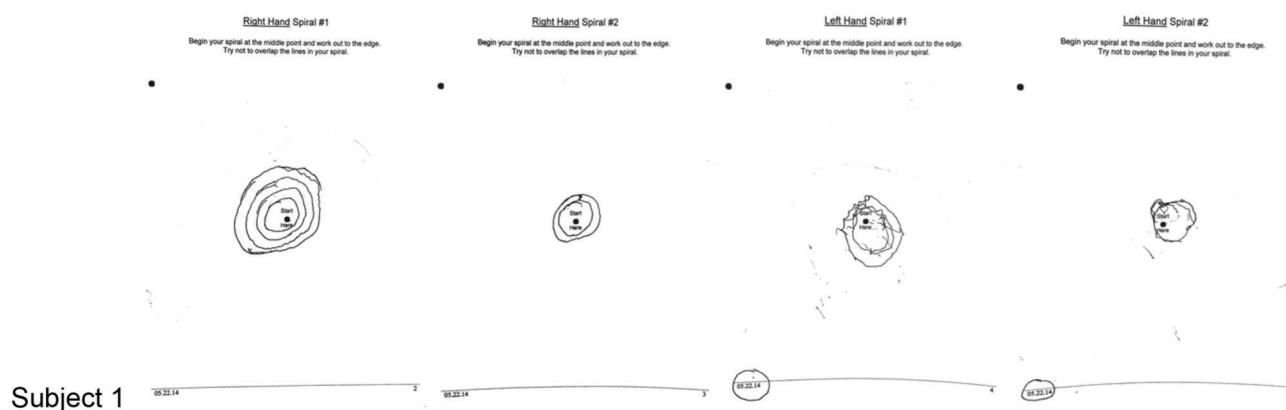


Figure 1. Abnormal Spiral Drawn by an ET Patient with Dementia. Patient 1 was unable to correctly complete the spiral drawing task because she did not understand the task. Rather than drawing spirals of increasing diameter, she attempted to draw one or two concentric circles and then got confused and stopped.



Video 1. Three ET Patients with Co-morbid Dementia. Patient 1. This 85-year-old female has had essential tremor for 10 years. Her most recent total tremor score was 20 out of 36, indicative of moderate tremor. She has also experienced cognitive decline during the past 3 years, progressing during that period from Clinical Dementia Rating (CDR) = 1 (mild dementia) to CDR = 3 (severe dementia). Her most recent Mini-Mental State Examination score was 1 out of 30. In this video, the patient has difficulty performing the specified motor commands. With her right hand, she initially touches the pen and her own nose to command, but then goes on to have difficulty touching the pen as the task proceeds. Additionally, the hand gesture she uses for touching the pen is abnormal, with an open hand tilted to one side. She is then briefly confused when asked to use her left hand and struggles to complete the task correctly. This patient's difficulties may be the result of several different impairments such as those related to verbal comprehension, working memory, response inhibition, and/or ideomotor apraxia (i.e., impaired ability to perform a skilled gesture with a limb upon verbal command). **Patient 2.** This 84-year-old female has had ET for 12 years. Her most recent total tremor score was 16 out of 36. During the previous three years, cognitive decline was observed from CDR = 0 (no dementia) to CDR = 2 (moderate dementia). Her most recent Mini-Mental State Examination score was 13 out of 30. Here she demonstrates problems with alternating between touching her nose and a pen. When using her right hand, she seems to have difficulty locating the pen and reaches for the examiner's hand instead. This may reflect a spatial disturbance such as optic ataxia, which arises from an inability to locate objects in visual space. However, it may also reflect a language disturbance, particularly comprehension of the word 'pen'. **Patient 3.** This 99-year-old female has had ET for 24 years. Her most recent total tremor score = 27 out of 36. During the past three years, cognitive problems have developed, with a CDR = 1 (mild dementia) progressing to a CDR = 2 (moderate dementia). Her most recent Mini-Mental State Examination score was 17 out of 30. She has some difficulty initially following directions. When asked to repeat the sequence with her left hand, she comments, "I can't find where I oughta be". Her difficulty with correctly placing the left hand may be due to visuospatial dysfunction, dysfunctional left hemibody schema, or loss of mental set for the task instructions.

true for ET. That is, ET itself appears to be a risk factor for developing a gradient of cognitive impairments. Here, we present three tangible visual records of patients with ET who have developed dementia (Video 1, Figure 1). We do so to underscore the point that dementia is an issue for a not insignificant number of the ET patients whom we encounter in clinical settings, and not merely an academic problem on paper. Its presence has profound prognostic import for ET patients; indeed, increasing the Clinical Dementia Rating score is associated with a 3.80-fold increased risk of mortality among ET patients.³

References

1. Louis ED. Non-motor symptoms in essential tremor: a review of the current data and state of the field. *Parkinsonism Relat Disord* 2016;22:S115–118. doi: 10.1016/j.parkreldis.2015.08.034
2. Shill HA, Hentz JG, Jacobson SA, Belden C, Sabbagh MN, Beach TG, et al. Essential tremor in the elderly and risk for dementia. *J Neurodegener Dis* 2014;2014:328765. doi: 10.1155/2014/328765
3. Zubair A, Cersonsky TEK, Kellner S, Huey ED, Cosentino S, Louis ED. What predicts mortality in essential tremor? A prospective, longitudinal study of elders. *Front Neurol* 2018;9:1077. doi: 10.3389/fneur.2018.01077