Neuroepidemiology 2013;40:99–100
DOI: [10.1159/000342312](https://doi.org/10.1159/000342312)

‘When Did My Tremor Start?’: Can Essential Tremor Patients Pinpoint Their Disease Onset?

Julián Benito-León

Department of Neurology, University Hospital 12 de Octubre, Department of Medicine, Faculty of Medicine, Complutense University, and Centro de Investigación Biomédica en Red sobre Enfermedades Neurodegenerativas, Madrid, Spain

Essential tremor (ET) is one of the most common neurological disorders among adults, and it is the most common tremor disorder [1–8]. As in other neurodegenerative diseases, age of onset in ET is chiefly assessed by the patients’ (or occasionally their caregivers’) self-report, which may be of little documentary value. Surprisingly, there have been few data that have examined the accuracy of patients’ reports of age of onset of ET.

In ET, dating the onset is not a trivial issue. Indeed, age of onset is a variable of considerable importance. First, late-onset ET could have different biochemical and neuropathological underpinnings from earlier-onset ET [9, 10]. Second, as in other neurodegenerative disorders, such as Parkinson’s disease [11, 12], an association between older age of onset and mild cognitive impairment and dementia has been reported in ET (i.e. ET patients with older age of onset are more likely to have mild cognitive impairment and to be demented than are ET patients with younger age of onset, and later onset of ET is a risk factor for incident dementia) [13–15]. Third, age of onset is crucial when determining the rate of progression in cross-sectional studies [9].

In this issue of *Neuroepidemiology*, Louis [16] analyzed the agreement in data on age of onset in 86 patients with ET from a prospective, epidemiological study in New York. Patients with ET were assessed at two time points (baseline and follow-up), and data on age of onset were collected each time by self-report. Despite the observation that agreement between the baseline and follow-up reports was high ($p = 0.85$, $p < 0.001$), and greater agreement was associated with several clinical factors including age, medication use, embarrassment, depressive symptoms, cognitive test score and disease duration, the difference in reported age of onset at the two time points (age of onset_{baseline} – age of onset_{follow-up}) did range widely (from –47 to 32 years), and in 1/5 of cases was >10 years.

The results of this study are of special interest. Aside from the fact that only a prior pilot study, conducted by the same group, had addressed this reliability issue in ET [17], there were no other published data to aid clinicians and researchers.

The study had several strengths, including the large sample size, the exhaustive study of the patients and the well-thought-out approach to the statistical analyses. The study was not without limitations. Specifically, only 86 of 376 ET patients initially enrolled were followed up. However, these 86 enrollees were similar to the larger group of 376 from which they were drawn in terms of baseline tremor severity, baseline tremor duration and baseline age [16].

The study again demonstrates that age of onset, overall, is reliably reported. Yet investigators should approach these self-reports with some degree of caution. Differences in reported age of onset in ET can vary widely, and in up to 1/5 of patients they may be substantial. Louis [16] has proposed several approaches to supplement self-reported data in future studies. One approach would be to examine hand-writing samples (e.g. signatures, letters) retrospectively, as an objective record of the possible onset and development of tremor. Another one would be to use collateral history from family members. Despite this, the main question is: ‘When does ET really start?’ The answer still remains uncertain. Given the heterogeneous nature of ET [18], this may differ depending on the cause/s of the syndrome. When and where the neuropathological process develops in ET still remains uncertain.

Acknowledgment

Dr. Benito-León is supported by R01 NS039422 from the National Institutes of Health, Bethesda, Md., USA.

References

- 1 Benito-León J, Louis ED: Essential tremor: emerging views of a common disorder. *Nat Clin Pract Neurol* 2006;2:666–678, quiz 691.
- 2 Benito-León J, Louis ED: Clinical update: diagnosis and treatment of essential tremor. *Lancet* 2007;369:1152–1154.
- 3 Benito-León J, Louis ED: Update on essential tremor. *Minerva Med* 2011;102:417–439.
- 4 Benito-León J: How common is essential tremor? *Neuroepidemiology* 2009;32:215–216.
- 5 Benito-León J: Essential tremor: one of the most common neurodegenerative diseases? *Neuroepidemiology* 2011;36:77–78.

- 6 Benito-Leon J, Bermejo-Pareja F, Morales JM, Vega S, Molina JA: Prevalence of essential tremor in three elderly populations of central Spain. *Mov Disord* 2003;18:389–394.
- 7 Benito-Leon J, Bermejo-Pareja F, Louis ED; Neurological Disorders in Central Spain Study Group: Incidence of essential tremor in three elderly populations of central Spain. *Neurology* 2005;64:1721–1725.
- 8 Louis ED, Ferreira JJ: How common is the most common adult movement disorder? Update on the worldwide prevalence of essential tremor. *Mov Disord* 2010;25:534–541.
- 9 Louis ED, Faust PL, Vonsattel JP, et al: Older onset essential tremor: more rapid progression and more degenerative pathology. *Mov Disord* 2009;24:1606–1612.
- 10 Shill HA, Adler CH, Beach TG, et al: Brain biochemistry in autopsied patients with essential tremor. *Mov Disord* 2012;27:113–117.
- 11 Hughes TA, Ross HF, Musa S, et al: A 10-year study of the incidence of and factors predicting dementia in Parkinson's disease. *Neurology* 2000;54:1596–1602.
- 12 Hobson P, Meara J: Risk and incidence of dementia in a cohort of older subjects with Parkinson's disease in the United Kingdom. *Mov Disord* 2004;19:1043–1049.
- 13 Benito-León J, Louis ED, Mitchell AJ, Bermejo-Pareja F: Elderly-onset essential tremor and mild cognitive impairment: a population-based study (NEDICES). *J Alzheimers Dis* 2011;23:727–735.
- 14 Benito-León J, Louis ED, Bermejo-Pareja F; Neurological Disorders in Central Spain Study Group: Elderly-onset essential tremor is associated with dementia. *Neurology* 2006;66:1500–1505.
- 15 Bermejo-Pareja F, Louis ED, Benito-León J; Neurological Disorders in Central Spain (NEDICES) Study Group: Risk of incident dementia in essential tremor: a population-based study. *Mov Disord* 2007;22:1573–1580.
- 16 Louis ED: Age of onset: can we rely on essential tremor patients to report this? Data from a prospective, longitudinal study. *Neuroepidemiology* 2013;40:93–98.
- 17 Louis ED, Schonberger RB, Parides M, Ford B, Barnes LF: Test-retest reliability of patient information on age of onset in essential tremor. *Mov Disord* 2000;15:738–741.
- 18 Benito-Leon J: Essential tremor: from a monosymptomatic disorder to a more complex entity. *Neuroepidemiology* 2008;31:191–192.