

# Questionable Dystonia in Essential Tremor Plus: A Video-Based Assessment of 19 Patients

Sanjay Pandey, MD, DM\* and Sonali Bhattad, MD

In 2018, the consensus statement on the classification of tremor has included a new terminology “Essential Tremor Plus,” defined as tremor with the characteristics of essential tremor (ET) with additional soft neurological signs, including questionable dystonic posturing.<sup>1</sup> The consensus definition further states that the ET plus syndrome does not include other defined syndromes such as dystonic tremor and task-specific tremor. However, in a clinical setting interpretation of questionable dystonic posturing is subjective with a high rate of discordance and one of the major challenges is to differentiate ET plus from a dystonic tremor syndrome.<sup>2,3</sup>

We are reporting a video-based assessment of ET plus patients with a perspective of describing the spectrum of questionable dystonia. A retrospective chart review was done of 121 ET patients diagnosed using the 1998 criteria who attended our movement disorder clinic during the past 4 years. As per the new consensus criteria of ET, 42 patients were excluded (38 had a duration of <3 years and 4 had isolated head tremor). Of the remaining 79 patients, 45 (56.96%) were ET and 34 (43.03%) were ET plus (22 had questionable dystonia, 8 had rest tremor and 4 had gait difficulty). Dystonia was labelled as questionable if there was discordance between the 2 examiners (S.P., S.B.) regarding its presence. The video recordings of 22 patients with questionable dystonia were further reviewed, and 3 patients were removed from the final analysis because their video showed no clear tremor. Of the remaining 19 patients, questionable dystonia in different body segments was noted (see Video S1 and Table 1).

The mean age  $\pm$  standard deviation (SD) of the ET plus patients with questionable dystonia was significantly higher than ET patients ( $58.52 \pm 17.50$  vs.  $49.93 \pm 14.90$ , respectively; *t* test, *P* = 0.0499). The mean ( $\pm$  SD) duration of disease (in years) of the ET plus patients with questionable dystonia was also significantly higher than ET patients ( $11.84 \pm 9.52$  vs.  $7.51 \pm 3.25$ , respectively; *t* test, *P* = 0.0084). ET plus with questionable dystonia and ET were both more common in males (16/19, 84.21% vs. 34/45, 75.55%, respectively;  $\chi^2$  test, *P* = 0.4441). The thumb (left = 18 and right = 16) was

most frequently involved followed by the second digit (left = 10 and right = 13), wrist (left = 9 and right = 9), fifth digit (left = 7 and right = 7), third digit (left = 6 and right = 5), neck (retrocollis = 3, and anterocollis = 1), arm (n = 1), and forearm (n = 1).

Our study has provided some important observations. First, the thumb was more likely to be involved in questionable dystonia followed by the second digit and wrist. Second, ET plus patients with questionable dystonia were significantly older and had a longer duration of disease than ET patients, indicating that dystonic features appear with time. It will be interesting to follow-up these patients to know how many of them would develop obvious dystonia in the future. In our cohort, the tremor was asymmetric in 4 patients (cases 5, 11, 13, and 18) and it was jerky in 1 patient (case 8). Asymmetric jerky tremor is a feature of dystonic tremor, but none of our patients had other features of dystonia (sensory trick or null point).<sup>4</sup> Some of our patients also had spooning of hand, but they had no other features of dystonia. Kim and Louis<sup>5</sup> recently demonstrated spooning of hand as subtle dystonic posturing in 3 cases with hand tremors, including 2 cases of ET, but they also cautioned that overextending the arms may sometimes produce a picture that resembles spooning. Various neurophysiological tests can serve as an aid to resolve this diagnostic dilemma.<sup>4</sup> However, in a clinical setting, using electrophysiology may be difficult to do, and there is an urgent need for minimal criteria for the definition of dystonia.

One of the major limitations of our article is the retrospective evaluation of video recordings where tremor has not been documented uniformly. Also, the interrater agreement of the blinded review of these patients would be expected to be poor.

To conclude, our case series highlights the uncertainties that arise in the deep phenotyping of patients previously diagnosed as ET. Further studies of controls and long-term follow-up are needed to determine the significance of soft or questionable signs in ET plus. Such studies may lead to reconsidering the need for an ET plus category.

Department of Neurology, Govind Ballabh Pant Postgraduate Institute of Medical Education and Research, New Delhi, India

\*Correspondence to: Dr. Sanjay Pandey, Department of Neurology, Academic Block, Room No. 503, Department of Neurology, Govind Ballabh Pant Postgraduate Institute of Medical Education and Research, New Delhi, India 110002; E-mail: sanjaysgpgi2002@yahoo.co.in

**Keywords:** dystonia, essential tremor.

Relevant disclosures and conflicts of interest are listed at the end of this article.

Received 29 April 2019; revised 24 July 2019; accepted 20 August 2019.

Published online 23 October 2019 in Wiley Online Library (wileyonlinelibrary.com). DOI: 10.1002/mdc3.12838

TABLE 1 Questionable dystonia in different body segments

| Case no. | Age,<br>y | Sex    | Duration,<br>y | Questionable Dystonia |    |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
|----------|-----------|--------|----------------|-----------------------|----|---|---|---|-------|---------|-----|---|--------|---|---|---|-------|---------|-----|-------------------|-------------------|------------------|------------------|--|--|--|--|--|
|          |           |        |                | Right                 |    |   |   |   |       |         |     |   | Left   |   |   |   |       |         |     | Neck              |                   |                  |                  |  |  |  |  |  |
|          |           |        |                | Digits                |    |   |   |   |       |         |     |   | Digits |   |   |   |       |         |     | Antero-<br>collis | Latero-<br>collis | Retro-<br>collis | Torti-<br>collis |  |  |  |  |  |
|          |           |        |                | 1                     | 2  | 3 | 4 | 5 | Wrist | Forearm | Arm | 1 | 2      | 3 | 4 | 5 | Wrist | Forearm | Arm |                   |                   |                  |                  |  |  |  |  |  |
| 1        | 87        | Male   | 5              | +                     | +  | + |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 2        | 83        | Male   | 40             | +                     |    |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 3        | 77        | Male   | 15             | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 4        | 72        | Male   | 24             | +                     |    |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 5        | 70        | Male   | 5              | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 6        | 68        | Male   | 10             | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 7        | 68        | Male   | 10             | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 8        | 65        | Male   | 3              | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 9        | 61        | Male   | 3              | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 10       | 60        | Male   | 3              | +                     |    |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 11       | 60        | Female | 4              |                       |    |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 12       | 60        | Male   | 25             | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 13       | 58        | Male   | 4              |                       |    |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 14       | 50        | Female | 20             | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 15       | 47        | Male   | 12             | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 16       | 42        | Female | 15             | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 17       | 40        | Male   | 5              | +                     |    |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 18       | 26        | Male   | 14             | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| 19       | 18        | Male   | 8              | +                     | +  |   |   |   |       |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |
| Total    |           |        |                | 16                    | 13 | 5 | 2 | 7 | 9     |         |     |   |        |   |   |   |       |         |     |                   |                   |                  |                  |  |  |  |  |  |

## Author Roles

(1) Research project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript: A. Writing of the first draft, B. Review and Critique.

S.P.: 1A, 1B, 1C, 2A, 2B, 2C, 3A, 3B

S.B.: 1A, 1B, 1C, 2A, 2B, 2C, 3A, 3B

## Disclosures

**Ethical compliance statement:** This retrospective video review was part of our ongoing study titled “Clinical and Electrophysiological Profile of Essential Tremor Plus Patients” approved by the institutional ethics committee, Maulana Azad Medical College, New Delhi. A written informed consent was obtained from patients. We have also received consent from patients for the publication of the video. The patients gave consent to be videoed for publication both in print and online. 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.

**Funding Sources and Conflicts of Interest:** No specific funding was received for this work. The authors declare that there are no conflicts of interest relevant to this work.

## Financial Disclosures from the Previous 12 Months:

The authors declare that there are no additional disclosures to report.

## References

- Bhatia KP, Bain P, Bajaj N, et al. Consensus Statement on the classification of tremors. from the task force on tremor of the International Parkinson and Movement Disorder Society. *Mov Disord* 2018;33(1):75–87.
- Rajan R, Pandey S, Anandapadmanabhan R, Srivastava AK. Interrater and intrarater agreement on the 2018 consensus statement on the classification of tremors. *Mov Disord* 2018;33:1966–1967.
- Fasano A, Lang AE, Espay AJ. What is “essential” about essential tremor? A diagnostic placeholder. *Mov Disord* 2017;33(1):58–61.
- Pandey S, Sarma N. Tremor in dystonia. *Parkinsonism Relat Disord* 2016;29:3–9.
- Kim CY, Louis ED. “Spooning”: a subtle sign of limb dystonia. *Tremor Other Hyperkinet Mov* 2018;8:607. <https://doi.org/10.7916/D8B00NRV>

## Supporting Information

Supporting information may be found in the online version of this article.

**Video S1.** Questionable dystonic posturing in different body segments of essential tremor patients.