

## Brief Reports

## Dystonic Head Tremor and the Coexistence of Headache

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### Abstract

**Background:** Head tremor may be observed in the presence of cervical dystonia and sometimes coexists with headache. We wished to investigate the presence of headache in dystonic head tremor.

**Methods:** We studied the files of 19 patients from our outpatient clinic (1997–2017) with dystonic head tremor and assessed the co-occurrence of headache. We also performed a literature search of the topic.

**Results:** Cervicogenic headache was present in nearly 37% of patients with dystonic head tremor. More than 85% of our patients presented with a “no-no” head tremor.

**Discussion:** Headache is common in dystonic head tremor. Cervicogenic headache seems to be more frequent in patients with dystonic head tremor than in the general population. Future studies should compare the presence of cervicogenic headache in essential head tremor patients with that in patients suffering from dystonic head tremor.

**Keywords:** Cervicogenic headache, dystonic head tremor, cervical dystonia

**Citation:** Hulzenga MA, Beumer D, Koehler PJ. Dystonic head tremor and the co-existence of headache. *Tremor Other Hyperkinet Mov.* 2017; 7. doi: 10.7916/D8BR94Q6

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**Editor:** Elan D. Louis, Yale University, USA

**Received:** June 1, 2017 **Accepted:** October 18, 2017 **Published:** November 2, 2017

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**Funding:** None.

**Financial Disclosures:** None.

**Conflicts of Interest:** The authors report no conflict of interest.

**Ethics Statement:** This study was reviewed by the authors’ institutional ethics committee and was considered exempted from further review.

### Introduction

Head tremor is commonly seen in clinical practice. It is known that mild cervical dystonia (CD) often coexists in patients with a head tremor, but this is frequently overlooked.<sup>1</sup> CD is a form of adult-onset primary dystonia. Two types of tremor have been described in patients with CD: dystonic tremor and tremor associated with dystonia.<sup>1</sup>

Chapter 11.2 of the International Classification of Headache Disorders (ICHD-3 beta) distinguishes between cervicogenic headache (chapter 11.2.1) and headache attributed to craniocervical dystonia (CCD, chapter 11.2.3).<sup>2</sup> Cervicogenic headache has a prevalence of 0.17% in the general population and a female preponderance.<sup>3</sup> Headache attributed to CCD is defined as headache caused by dystonia involving the neck muscles with abnormal movements or defective posturing of the neck or head as a result of muscular hyperactivity (Table 1).<sup>2</sup> These patients suffer from neck and posterior

head pain that has developed or worsened in temporal relation to CCD onset, and headache location needs to correspond with the location of the dystonic muscles.<sup>2</sup> In a study of 80 patients suffering from CCD, only one fulfilled the diagnostic criteria for headache attributed to CCD (1.3%). However, headache was present in 56.2% of the patients and seemed to be as prevalent as in healthy controls (without CCD).<sup>4</sup>

To our knowledge, the co-occurrence of dystonic head tremor and headache has not been studied. Following the presentation of a patient with cervicogenic headache and a head tremor, of which she did not seem to be aware, we wondered whether this was common, especially given a report that patients are often unaware of their own head shaking.<sup>5</sup> In this study, we examined patients with dystonic head tremor for the presence of (cervicogenic) headache to further elucidate the issue.

**Table 1. Diagnostic Criteria of Headache Attributed to Craniocervical Dystonia (ICHD-3 beta)**

<b>A</b>	Neck and posterior head pain fulfilling criterion C
<b>B</b>	Craniocervical dystonia is demonstrated by abnormal movements or defective posturing of the neck or head as a result of muscular hyperactivity
<b>C</b>	Evidence of causation demonstrated by at least two of the following: 1) headache has developed in temporal relation to the onset of craniocervical dystonia 2) headache has significantly worsened in parallel with progression of the craniocervical dystonia 3) headache has significantly improved or resolved in parallel with improvement in or resolution of the craniocervical dystonia 4) headache location corresponds to the location of the dystonic muscle(s)
<b>D</b>	Not better accounted for by another ICHD-3 diagnosis

### Methods

We searched for patients with dystonic head tremor treated at our outpatient clinic between 1997 and 2017 and assessed the presence of headache. Head tremor was classified as “yes-yes” tremor, “no-no” tremor, or “other” tremor. Diagnoses were made by general neurologists and movement disorder specialists. All patients were seen at least once by the same general neurologist with special interest in headache and long-term experience in movement disorder patients (P.K.). We retrospectively assessed patient files and described age, age at diagnosis, sex, type of head tremor (“yes-yes,” “no-no,” “other”), presence of voice tremor, and presence of headache.

In addition, we examined the medical literature by searching PUBMED and MEDLINE databases using the terms “tremor,” “cervical dystonia and headache,” and “head tremor and headache.”

### Results

#### Index case

As an illustrative case, we describe a 65-year-old female who first presented in 2000 with a cervicogenic headache that she had been suffering with for several years. In subsequent years, she was treated by her general practitioner (GP). In 2011, she was again referred to our outpatient clinic because of headache. Her GP mentioned a head tremor, of which she did not seem to be aware or at least did not complain. At examination, a dystonic “no-no” head tremor was noted with a slight tendency to turn her head to the right. A subtle tremor of her right arm was also observed, faster and of lower amplitude than is seen in essential tremor (ET). The head tremor increased during ophthalmoscopy, and the arm tremor was particularly visible during the Barré test (arms stretched in front of the patient). She was diagnosed with CD with dystonic tremor and secondary cervicogenic headache. As she did not complain of her head tremor, she only received low-dose amitriptyline for her cervicogenic headache, which provided sufficient relief. She has not consulted the outpatient clinic since.

#### Included patients

We found data from 19 patients with dystonic head tremor. The mean age was 64 years (SD 13.1), and 13 (68.4%) were female.

The mean age at the time of diagnosis of dystonic head tremor was 58 years (SD 13.1), and 17 (89.5%) patients presented with a dystonic “no-no” tremor. Headache was present in 10 (52.6%). Cervicogenic headache was present in seven of 10 patients, while the other three had migraine, undefined, and tension-type headache. Three cases of CD were treated by botulinum toxin. Two of these patients were also suffering from headache, and the botulinum toxin did not provide a sufficient response for the headache in either case. A voice tremor was present in four (21.1%) patients. Limb tremor was not described in any of the other patients. The results are presented in Table 2.

#### Literature search

Entering the terms “tremor,” “cervical dystonia and headache,” and “head tremor and headache” provided three and 18 results in the PUBMED online database, respectively. None of these articles addressed the specific subject we were interested in, the co-occurrence of cervicogenic headache and head tremor in CD. Using the same terms in the MEDLINE online database also did not yield any relevant articles.

### Discussion

Among our patients with dystonic head tremor, we found headache in 52.6% of them, which matches the prevalence in earlier studies of headache in CCD (56.2% and 58.6%, respectively).<sup>4</sup> Headache prevalence in the general population is 47%.<sup>6</sup>

Cervicogenic headache occurred in nearly 37% in our patient population, which is more frequent than the prevalence in the general population (0.17%).<sup>3</sup> CD was subtle in most of the patients and only three were being treated with botulinum toxin, which in general was not effective for headache (of which two of the three patients were suffering). Barbanti had more positive results with botulinum toxin treatment in patients with primary CCD; among 45 patients, the headache improved or disappeared after treatment in 12 (26.7%).<sup>4</sup> Although “Headache attributed to CCD” is a well-known clinical syndrome, dystonia in head tremor may be subtle and go unrecognized.<sup>1,7</sup> ET is sometimes mistakenly diagnosed in such cases, even if there is no accompanying limb tremor.<sup>8</sup>

The ICHD-3 beta diagnosis “Headache attributed to CCD” (11.2.3)<sup>2</sup> may be considered if the head tremor is accompanied by

Table 2. Characteristics of Patients with Dystonic Head Tremor

Patient	Age (Years)	Age of Diagnosis (Years)	Sex	Tremor Capitis	Voice Tremor	Headache	Dystonia Treatment
1	73	62	Female	Other (complex with jerky movements)	Yes	Mentioned left-sided cervicogenic headache	+
2	65	60	Female	“No-no”	No	Yes, cervicogenic headache	-
3	48	44	Female	“Yes-yes”	No	No	+
4	66	61	Male	“No-no”	No	No	-
5	55	40	Female	“No-no”	No	Yes, migraine	-
6	78	71	Female	“No-no”	Yes	No	-
7	72	53	Male	“No-no”	Yes	No	-
8	75	72	Female	“No-no”	Yes	Yes, but mentioned before onset of cervical dystonia	-
9	55	52	Male	“No-no”	No	Yes, cervicogenic headache and neck pain	+
10	60	58	Male	“No-no”	No	No	-
11	72	68	Female	“No-no”	No	No	-
12	56	49	Female	“No-no”	No	No	-
13	61	57	Female	“No-no”	No	Yes, cervicogenic headache	-
14	74	64	Female	“No-no”	No	No	-
15	63	63	Male	“No-no”	No	Yes, cervicogenic headache	-
16	71	71	Female	“No-no”	No	Yes, right-sided headache	-
17	85	85	Female	“Yes-yes/no-no”	No	Yes, cervicogenic headache	-
18	57	57	Female	“No-no”	No	No	-
19	24	24	Male	“No-no”	No	Tension-type headache	-

evident dystonia, but this seems to be rare.<sup>2,4,9</sup> We could not attribute the symptoms that our index patient presented with to this kind of headache with certainty since there was no temporal relationship between the onset of headache and that of her head tremor. In general, criteria 1 to 3 of C (Table 1) are often not met as the temporal relation to the onset of the CCD is not rarely unknown, especially if the

tremor is the leading symptom. The same is true for item 2 (worsening in parallel with CCD progression). Finally, item 3 is also infrequently met as the dystonia may be subtle and the tremor not sufficiently severe to be eligible for treatment.

CD movements may include a rhythmic, tremor-like component with a superimposed, larger amplitude, irregular movement.<sup>10</sup>

When the head is turned to the direction of the desired posture, the tremor may decrease (“null point” often induced by the “geste antagoniste”). When the head is turned to the opposite direction, the tremor may increase. However, as mentioned above, these features may be very subtle in head tremor.

A striking 16 out of 19 (84.2%) patients presented with a pure “no-no” head tremor. Previously published literature provides contradictory information about the relationship between head tremor and the underlying diagnosis.<sup>6-8</sup> Conclusions regarding the direction of the head tremor therefore cannot be drawn.

### Limitations

Since our study included only a small group of patients, confirmation of our findings in a larger study group would be desirable. Considering the retrospective nature of the present series, that should preferably be done in a prospective study.

### Future considerations

In our small clinical sample, we found cervicogenic headache to be more prevalent in patients with dystonic head tremor than in the general population. Other, preferably prospective, studies should evaluate this finding. Patients with both head tremor in ET and CD should be included to find out whether there are differences between the two types of head tremor with respect to the prevalence rates and headache types.

In conclusion cervicogenic headache in dystonic head tremor seems to be much more frequent than in the general population. The diagnostic criteria for headache attributed to CCD (ICHD-3beta 11.2.3) are rarely met.

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