

SHORT REPORT

Sex prevalence of focal dystonias

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Abstract

The sex prevalence of idiopathic focal dystonia is reported from a data base review of all patients seen at the National Hospital of Neurology, Queen Square and King's College, London up to 1993. There was a higher prevalence of females to males in all categories of focal dystonia involving the craniocervical region. The female to male ratio for cranial dystonia was 1.92:1 (P < 0.01) and 1.6:1 (P < 0.001) for spasmodic torticollis. On the other hand, twice as many men than women had writer's cramp (M:F = 2.0:1, P < 0.01). At present, there is no clear explanation to account for this differences in the sex prevalence of different types of focal dystonia.

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There is some uncertainty about the female to male prevalence of patients with focal dystonia. A recent survey published in *The National Spasmodic Torticollis Association Newsletter*

(USA)¹ concerning the sex of persons with spasmodic torticollis showed that of 1812 respondents, 74% were female and 26% were male, giving a female to male ratio of 2.85:1. The female prevalence in this survey was well in excess of that described in the medical literature (table 1).²⁻⁹ We therefore undertook a study to look at our population of patients with isolated spasmodic torticollis as well as those with other forms of focal dystonia to determine the female to male ratios.

Patients and methods

We reviewed the data base of our patients with a diagnosis of idiopathic focal dystonia up to 1993. Patients with generalised or multifocal dystonia were eliminated, as well as those with neurological illness that might have caused their dystonia or those with exposure to drugs known to cause tardive dystonia. We divided the patients into different categories according to the type of focal dystonia: isolated spasmodic torticollis, isolated blepharospasm, isolated oromandibular dystonia, cranial dystonia, or Meige's syndrome (blepharospasm with oromandibular dystonia), isolated spasmodic dysphonia, and isolated writer's cramp. In each category, the number of females and males were counted and the female to male ratio was calculated (table 2); the χ^2 test was used to test statistical significance.

Results

Table 2 shows the results of our study—namely, the total number of patients, the number of females and males, and the female to male ratio for each category of focal dystonia. The female to male ratio varied from 1.6:1 to 3.3:1 for categories of dystonia involving the craniocervical region. For writer's cramp the female to male ratio was 1:2. For spasmodic torticollis, cranial dystonia, and writer's cramp this difference was statistically significant.

Discussion

In our study females were more prevalent than males in all categories of focal dystonia involving the craniocervical region. The female to male ratio for cranial dystonia was 1.92:1 and for spasmodic torticollis it was 1.60:1, lower than the figure of 2.85:1 obtained by The National Spasmodic Torticollis Association.¹

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Table 1 Large studies of patients with spasmodic torticollis in the medical literature

Study	Type of dystonia	No of patients*	F/M	Ratio
Friedman and Fahn ² 1986	cervical	116 (116)	72/44	1.6:1
Duane ³ 1988	cervical	347	—	1.6:1
Jahanshahi ⁴ et al 1990	cervical‡	72	37/35	1.0:1
Jankovic ⁵ et al 1990†	cervical‡	232 (209)	138/94	1.5:1
Chan ⁶ et al 1991	cervical‡	266 (214)	173/93	1.9:1
Rondot ⁷ et al 1991	cervical‡	347	123/76	1.6:1
Jankovic ⁸ et al 1991	cervical‡§	300	—	1.5:1
Deuschl ⁹ et al 1992†	cervical‡	100	54/46	1.2:1

*Total number of patients shown with, in parentheses, the number with pure spasmodic torticollis when known.

†Botulinum toxin study.

‡Included cases of segmental, multifocal, or generalised dystonia.

§Included cases of tardive dystonia.

Table 2 Patients with focal dystonia evaluated at the National Hospital and King's College up to 1993

Type of dystonia	No of patients	F/M	Ratio	P value (χ^2)
Isolated ST	424	261/163	1.60:1	< 0.001
Isolated B	102	68/34	2.00:1	0.02
Isolated OM	30	23/7	3.28:1	0.06
Cranial dystonia (CD)	178	117/61	1.92:1	0.01
Isolated SD	36	26/10	2.60:1	0.09
Writer's cramp	186	63/124	1.00:2	0.01

ST = spasmodic torticollis; B = blepharospasm; OM = oromandibular dystonia; CD = combination of blepharospasm and oromandibular dystonia; SD = spasmodic dysphonia.

Table 3 Large studies of patients with blepharospasm in the literature

Study	Type of dystonia	No of patients*	F/M	Ratio
Cohen <i>et al</i> ¹⁰ 1986†	Blepharospasm‡	75 (54)	47/28	1.7:1
Carruthers and Stubbs ¹¹ 1987†	Blepharospasm‡	47 (22)	27/20	1.35:1
Grandas <i>et al</i> ¹² 1988	Blepharospasm‡§	264 (58)	170/94	1.8:1

*Total number of patients shown with, in parentheses, the number with pure blepharospasm when known.

†Botulinum toxin study.

‡Included cases of segmental, multifocal, or generalised dystonia.

§Included cases of tardive dystonia.

Table 4 Large studies of patients with spasmodic dysphonia in the medical literature

Study	Type of dystonia	No of patients*	F/M	Ratio
Jankovic and Ford ¹¹ 1983	Spasmodic dysphonia‡	21 (6)	20/7	2.85:1
Ludlow <i>et al</i> ¹⁴ 1988†	Spasmodic dysphonia	16 (16)	15/1	15:1
Rosenfield <i>et al</i> ¹⁵ 1988	Spasmodic dysphonia‡	41	32/9	3.5:1
Blitzer <i>et al</i> ¹⁶ 1988†	Spasmodic dysphonia‡	73 (26)	42/31	1.35:1

*Total number of patients with, in parentheses, the number with pure spasmodic dysphonia when known.

†Botulinum toxin study.

‡Included cases of segmental, multifocal, or generalised dystonia.

The much greater number of females in their survey is possibly due to a selection bias, with more females than males responding. We have also found that nearly two thirds of patients attending our outpatient botulinum toxin clinic to receive injections for spasmodic torticollis are female. This figure is higher than the increase in female to male ratio seen when we looked at the total population of our patients with spasmodic torticollis. Clearly referral bias can be a confounding factor in such studies and the female to male ratio seems to depend on the specific population studied.

There have been many large studies of focal dystonia (tables 1, 3 and 4)²⁻¹⁶ showing a preponderance of females to males for forms of focal dystonia involving the craniocervical region. However, many of these studies have included cases with craniocervical dystonia as part of multifocal or generalised dystonia, and some even included cases of tardive dystonia. Some studies were designed to evaluate the efficacy of botulinum toxin, with the inherent problem of selection bias in such series. Nevertheless, there seems to be a slight but clear preponderance of females with various types of focal dystonia involving the craniocervical region.

Why does craniocervical dystonia affect women more than men? Duane³ has raised the possibility that women are more at risk for these forms of dystonia because of specific oestrogen receptors within the CNS which could influence involuntary motor function. Oestrogens can affect the nigrostriatal dopaminergic system.¹⁷ Thyroid disease and other autoimmune conditions, which are more common in women than men, have also been implicated in the pathophysiology of cervical dystonia by some authors³ but refuted by others.^{6,7}

On the other hand, if the same gene is responsible for focal and generalised idiopathic torsion dystonia,¹⁸ it is possible that females are less susceptible to the more extensive forms of the disease and may exhibit a minor expression of the gene. In two studies of patients with idiopathic generalised dystonia reported by

Fletcher *et al*¹⁹ and Bressman *et al*²⁰ there was no clear evidence that generalised dystonia was less prevalent among females. Among the 107 index cases with generalised dystonia studied by Fletcher *et al*¹⁹ 45 were male and 62 were female, whereas in the 43 patients reviewed by Bressman *et al*²⁰ 23 were male and 20 were female. In their families, those with focal dystonias also showed no clear sex predominance.^{20,21}

Interestingly, in some other forms of focal dystonia—for example, writer's cramp—males are preponderant as seen in our study (table 2) and in the medical literature.²² Among 91 patients with writer's cramp, Sheehy *et al*²² found 36 females and 55 males giving a female to male ratio of 1:1.5. Typist's cramp, on the other hand, occurs mainly in women²² and golfer's cramp mainly in men.²³

Our study confirms a clear but mild preponderance of females with various types of craniocervical dystonia and of males with writer's cramp. Why this is so remains to be discovered.

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