RESEARCH ARTICLE

The Prevalence of Primary Dystonia: A Systematic Review and Meta-analysis

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ABSTRACT: Dystonia is a hyperkinetic movement disorder characterized by sustained muscle contractions that produce repetitive movements and abnormal postures. Specific information on the prevalence of dystonia has been difficult to establish because the existing epidemiological studies of the condition have adopted different methodologies for case ascertainment, resulting in widely differing reported prevalence. Medline and Embase databases were searched using terms specific to dystonia for studies of incidence, prevalence, and epidemiology. All population-based studies reporting an incidence and/or prevalence of primary dystonia were included. Sixteen original studies were included in our systematic review. Fifteen studies reported the prevalence of dystonia, including 12 service-based and three population-based studies. We performed a meta-analysis on the results of the service-based studies, and were able to combine data on the prevalence of several dystonia subtypes. From these studies, we calculated an overall prevalence of primary dystonia of 16.43 per 100,000 (95% confidence interval [CI]: 12.09–22.32). The prevalence of dystonia reported in the three population-based studies appears higher than that reported in the service-based studies. Only 1 of the 16 studies reported an incidence of cervical dystonia. This corresponded to a corrected incidence estimate of 1.07 per 100,000 person-years (95% CI: 0.86–1.32). Despite numerous studies on the epidemiology of dystonia, attempting to determine an accurate prevalence of the condition for health services planning remains a significant challenge. Given the methodological limitations of the existing studies, our own prevalence estimate of primary dystonia likely underestimates the true prevalence of the condition.

Key Words: prevalence studies; incidence studies; dystonia; blepharospasm

Dystonia is a condition characterized by sustained muscle contractions producing repetitive movements and abnormal postures. The primary generalized dystonias are progressive, disabling disorders that typically begin in youth. They are associated with mutations in a number of genetic loci, the majority of which follow an autosomal dominant pattern of inheritance with a reduced penetrance.[1] The primary focal dystonias nearly always occur in adults and typically involve the neck, face, or arms. Although many patients with focal dystonia report having relatives with the condition[2–4] and evidence exists for susceptibility genes that may act with environmental triggers,[5] specific genes for the condition have not been identified.

Dystonia often has an adverse effect on quality of life and can be markedly disabling[6]; however, specific information on the prevalence of dystonia that would be useful for health services planning has been difficult to establish. To date, the epidemiological studies published have adopted different methodologies for case ascertainment and have demonstrated widely different prevalences that have varied between regions. Whether this variance reflects differences in biological substrates or the methodological approaches of each study has been difficult to determine.

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

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The limitations of published studies reflect the difficulty of applying traditional methods of determining prevalence to dystonia. For example, case identification through computerized records of investigations or medications cannot be reliably applied to the study of dystonia, because only a proportion of patients receive treatment, and through varying specialties. Routine mortality and hospital admission data cannot be used as a proxy for prevalence because the condition has low morbidity and is generally nonfatal. Potentially, the greatest barrier to accurate ascertainment is that the diagnosis of dystonia is a clinical one, and uncertainty over diagnosis is relatively common. Family studies have highlighted this problem: One study of focal dystonia identified an additional undiagnosed affected relative in 25% of index cases.[7]

Ideally, door-to-door surveys would be used to identify all cases of dystonia regardless of medical seeking tendencies, previous diagnosis, or clinical severity. Although this would likely yield the most accurate data, dystonia is still a relatively rare condition, and this approach would therefore be limited by cost and practical considerations. Instead, the majority of published estimates of prevalence have been derived from less-resource-intensive service-based studies, which are prone to the limitations described above.

Given the variability in the reported prevalence of dystonia, we performed a systematic review of studies evaluating the epidemiology of dystonia. Where possible, meta-analysis was performed to provide estimates of prevalence of all types of dystonia, as well the effects of age and sex.

Patients and Methods

Medline and Embase databases were searched (December 2010) for studies of incidence, prevalence, and epidemiology using terms specific to dystonia (see Appendix for Medline search strategy). All studies reporting the incidence and/or prevalence of primary dystonia within a defined population after 1985 were included. We excluded studies that did not report on original research or were published in languages other than English or French. Two reviewers screened abstracts and identified full texts to determine study eligibility. The references of the included studies as well as review articles were screened to ensure that additional relevant studies were not missed.

Two reviewers independently extracted data and then confirmed their accuracy through discussion. Demographics and diagnostic data collected, the sources for these data, and the definitions and diagnostic criteria were recorded. Prevalence and incidence of the conditions within each study were recorded, as was stratification by age and gender, where given.

To ensure internal consistency and to permit accurate comparisons, studies examining similar populations and similar diagnoses, using similar methods, were grouped together. To assess for significant between-study heterogeneity, the Cochrane Q statistic was calculated and I^2 was used to quantify between-study heterogeneity. When significant heterogeneity was absent, the pooled prevalence per 100,000 people and 95% confidence intervals (CIs) were calculated using a fixed-effect model. When significant heterogeneity was present, a random-effects model was used. With a fixed-effect model, the studies are weighted using the inverse of the variance (larger studies receive more weight), and with a random-effects model, the inverse variance is corrected by a measure of between-study variation (tau-squared), thus reducing the effects of sample size. Because prevalence is a proportion, study estimates were combined using a log transformation to help normalize the data. Metaregression was used to determine whether a significant difference in the estimates of dystonia was present between men and women or between geographic locations. We investigated publication bias visually using funnel plots and statistically using both Begg and Mazumdar's and Egger et al.'s tests.[8,9]

Statistical Analyses

For all tests, $P < 0.05$ was deemed significant. All statistical analyses were carried out in *R* version 2.14.[10] The *meta* package was used to produce the pooled estimates, forest plots, and publication bias assessment.[11] The *metafor* package was used to conduct the metaregression using restricted maximum likelihood estimation.[12]

Results

Supporting Figure 1 outlines the selection process for the included studies. Sixteen studies met all inclusion criteria, with 15 reporting on prevalence (see Tables 1 and 2). Twelve of the studies were service based, reporting on patients accessing medical care for the diagnosis or treatment of dystonia, and three of the studies were population based. There was no evidence of publication bias across any of the meta-analyses. As a result of the small number of studies included in the analysis, we were likely underpowered to detect significant bias. However, we would not *a priori* expect a tendency for underpublication of findings outside an accepted range, because there is little agreement on definitive rates for incidence and prevalence of the various forms of dystonia.

Marras et al.'s[13] study was the only study reporting on an incidence of dystonia. They estimated the minimum incidence of primary cervical dystonia using the electronic medical records of Kaiser Permanente of

TABLE 1. Prevalence studies of dystonia

Study, Year	Country	Age	Population Size	Diagnostic Criteria	Data Source	Diagnosis Established by
Asgeirsson et al., *2006*[22]	Iceland	All	288,201	DMFR	Hospital/clinic chart review; administrative database; prescription drug database	Clinical assessment by a health professional; medical chart review
Butler et al., 2004[23]	England	All	2,605,100	DMFR	Mailed survey; hospital/clinic chart review	Clinical assessment by a health professional; medical chart review
Castelon Konkiewitz et al., 2002[24]	Germany	All	1,322,883	Fahn	Hospital/clinic chart review; administrative databases	Clinical assessment by a health professional; medical chart review
Cossu et al., 2006[25]	Italy	All	1,652,332	Unspecified	Hospital/clinic chart review; prescription drug database	Clinical assessment by a health professional; medical chart review
Das et al., 2007[15]	India	All	52,377	Unspecified	Stratified random sample door-to-door survey	Clinical assessment by a health professional
Defazio et al., 2001[26]	Italy	All	67,606	Fahn	Hospital/clinic chart review	Clinical assessment by a health professional; medical chart review
ESDE Collaborative Group, 2000[21]	France, Austria, England, Spain, Finland, Germany, Portugal, Italy	>20 years	5,792,937	Fahn	Hospital/clinic chart review; administrative database; prescription drug database	Medical chart review
Fukuda et al., 2006[18]	Japan	All	247,973	Fahn	Mailed survey; hospital/clinic chart review	Clinical assessment by a health professional; medical chart review
Kandil et al., 1994[14]	Egypt	All	42,000	Unspecified	Door-to-door survey	Unspecified
Le et al., 2003[20]	Norway	All	508,726	DMRF	Hospital/clinic chart review; administrative databases	Medical chart review
Matsumoto et al., 2003[17]	Japan	All	1,459,130	Unspecified	Hospital/clinic chart review; administrative databases	Clinical assessment by a health professional; medical chart review
Müller et al., 2002[16]	Italy	≥50	707	Fahn	Random population sample	Clinical assessment by a health professional
Papantonio et al., 2009[27]	Italy	≥17	541, 653	Unspecified	Hospital/clinic chart review; administrative databases; prescription drug database	Medical chart review; administrative data codes
Pekmezovic et al., 2003[28]	Serbia	≥20	1,602,226	Unspecified	Hospital/clinic chart review	Medical chart review
Sugawara et al., 2006[29]	Japan	All	1,166,967	Dystonia working group guideline 2004	Mailed survey; hospital/clinic chart review	Clinical assessment by a health professional; medical chart review

Abbreviations: DMRF, Dystonia Medical Research Foundation; Fahn, Fahn classification of dystonia; ESDE, The Epidemiological Study of Dystonia in Europe.

Northern California to identify cases first diagnosed between 1997 and 1999. They identified 66 cases of primary cervical dystonia during a total of 8,215,110 person-years. This corresponded to an incidence of 0.8 per 100,000 person-years (95% CI: 0.63, 1.03) and a corrected estimate of 1.07 per 100,000 person-years (95% CI: 0.86–1.32).

From the service-based studies, we were able to combine data on the prevalence of primary dystonia, focal dystonia, focal and segmental dystonia, generalized dystonia, blepharospasm, cervical dystonia, laryngeal dystonia, oromandibular dystonia, limb dystonia, and writer's cramp. We calculated sex-specific prevalence for focal/segmental dystonia, cervical dystonia, and blepharospasm. We were unable to perform a meta-analysis to derive age-specific estimates as a result of variable grouping of ages across studies. Data from studies that provided age-specific estimates are summarized in Table 2.

The population-based studies consisted of two studies that relied on door-to-door surveys: Kandil et al. evaluating a population in Upper Egypt[14] and Das et al. evaluating a population in Calcutta, India.[15] A third study evaluated a random selection of patients over the age of 50 in Bruneck, South Tyrol, Italy.[16] These three studies generated markedly different results.

Kandil et al. studied 7,000 families (42,000 subjects) from urban, suburban, and rural communities in Upper Egypt from March 1988 to June 1990.[14] Subjects were interviewed at home, and those identified by screening interview and examination as having abnormal movements were reinterviewed and examined to confirm and assess the etiology of their

TABLE 2. Prevalence of dystonia by age group

Study	Dystonia Subtype	Age Group	Prevalence/10^5
Das et al., 2007[15]	Primary dystonia	<29	7.6
		30–49	31.3
		50–69	177.9
		>70	130.8
Defazio et al., 2001[26]	Blepharospasm	<29	0
		30–49	0
		50–69	26.6
		60–69	31.9
		>69	74.0
Le et al., 2003[20]	Focal/segmental dystonia	<29	4.1
		30–49	23.8
		50–69	51.4
		>70	58.7
Papantonio et al., 2009[27]	Focal/segmental dystonia	18–34	3.9
		35–54	9.9
		55–74	27.4
		>75	16.3
Pekmezovic et al., 2003[28]	Focal dystonia	20–49	7.2
		50–59	19.9
		60–69	10.9
		>70	31.4
ESDE, 2000[21]	Focal dystonia	20–49	6.4
		50–59	17.9
		60–69	20.6
		70–95	17.8

disorder. The researchers did not specify the qualifications of those performing the screening and subsequent assessments, but stated that further clinical assessments and investigations required for diagnosis were carried out at Assiut University Hospital (Assiut, Egypt). The researchers did not specify for how many subjects this detailed evaluation was undertaken.

The researchers reported on a prevalence of focal dystonia of 10 per 100,000. All cases were cervical dystonias and all were observed in 30 to 40 year olds, producing an age-specific prevalence of 203 per 100,000. No cases of generalized dystonia were recorded, and no differences were observed between the regions studied.

Das et al. sampled the population of Calcutta with random, two-stage, stratified selection of subjects.[15] A nonmedical team performed the door-to-door surveys, administering to a senior household member a screening questionnaire directed at the entire household. A study neurologist then examined any screened positive cases and recorded clinical data. Two senior neurologists then confirmed any screened positive cases by review of the paper history and, in some instances, with in-person examinations.

Of 52,377 subjects screened, 23 were diagnosed with primary dystonia. This generated a crude prevalence of 43.91 per 100,000 (95% CI: 28.41–64.81). All cases were focal dystonias, predominantly manifesting as writer's cramp and primary writing tremor, followed by facial/cervical dystonia. Age of onset was earlier for women (mean, 43.5 years) than men (mean, 46.6 years). Limb dystonia was more common among men and facial dystonia more common among women, but cervical dystonia was equally common.

Finally, Müller et al. evaluated the prevalence of primary dystonia in a random population sample of individuals over age 50 participating in the Bruneck study.[16] As part of this study, a baseline examination was performed in 1990, and follow-up evaluations were performed at 5-year intervals. During the second follow-up, subjects remaining in the study (92%) were screened for dystonia by a movement disorders specialist. A standardized history and neurologic examination was obtained from all patients. If dystonia was suspected, videotape recordings were made and independently reviewed by three other movement disorders specialists. Patients were classified as having definite dystonia when all three examiners made a diagnosis of the condition. For positive cases, a medical work-up for secondary causes was performed.

A total of 15 potential cases were identified at the time of the on-site examination. Nine were excluded with alternative diagnoses. Therefore, primary dystonia was present in 6 of 707, resulting in a prevalence of 732 per 100,000 (95% CI: 319–1,564) in the general population over age 50. Only two of the cases had been previously diagnosed.

Twelve studies assessed the prevalence of dystonia in clinics and hospitals serving defined areas (Table 1). With the exception of three studies from Japan, the locations for these studies were primarily in Europe.

Because these serviced-based studies used similar methodologies, we were able to combine their data for analysis. We calculated an overall prevalence of primary dystonia of 16.43 per 100,000 (95% CI: 12.09–22.32; $I^2 = 96.4\%$; $Q = 110.8$; $df = 4$; $P < 0.0001$; see Fig. 1). The overall prevalence of focal and segmental dystonias was 15.36 per 100,000 (95% CI: 12.06–19.55; $I^2 = 95.6\%$; $Q = 157.8$; $df = 7$; $P < 0.0001$; see Fig. 2). Prevalence was higher in women 22.35 (95% CI: 12.05–41.46) than in men 14.58 (95% CI: 9.43–22.52); however, this difference was not statistically significant by metaregression.

We calculated an overall prevalence of cervical dystonia of 4.98 per 100,000 (95% CI: 3.58–6.94; $I^2 = 93.1\%$; $Q = 115.2$; $df = 8$; $P < 0.0001$; see Fig. 3). Metaregression analysis revealed no significant differences between men and women for cervical dystonia, with a prevalence in women of 6.48 per 100,000 (95% CI: 3.49–12.04) and in men of 4.98 per 100,000 (95% CI: 4.12–6.01). We calculated an overall prevalence of blepharospasm of 4.24 per 100,000 (95% CI: 2.92–6.18; $I^2 = 94.3\%$; $Q = 157.5$; $df = 9$; $P < 0.0001$; see Fig. 4); again, metaregression analysis revealed no significant differences between sexes, with 4.78 cases of blepharospasm per 100,000 (95% CI: 4.11–5.57) in women and 3.08 per 100,000 (95% CI: 1.63–5.82) in men.

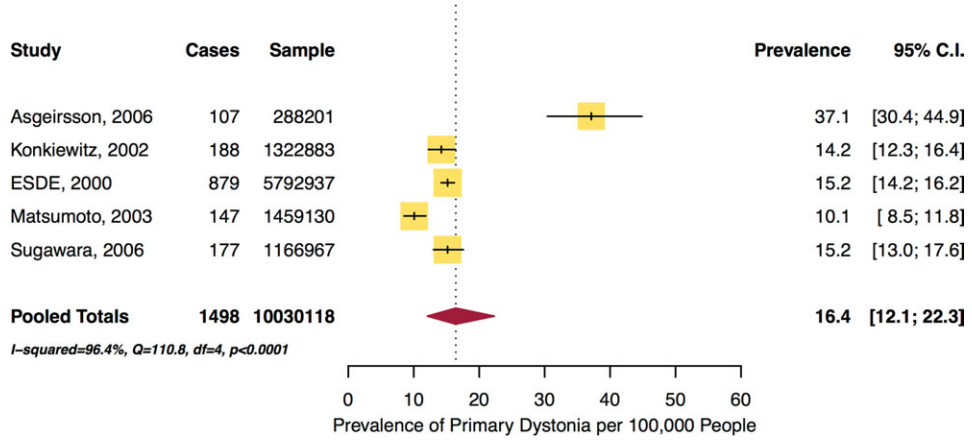

FIG. 1. Prevalence of primary dystonia (per 100,000).

We conducted stratified analyses and metaregression to determine whether the prevalence of cervical dystonia or blepharospasm differed by location. Three studies conducted in Japan were compared to six studies conducted in Europe. This analysis suggests that the prevalence of cervical dystonia is significantly lower in Japan than in Europe, with a reported prevalence of 2.52 (95% CI: 2.00, 3.18) in Japan and 6.71 (95% CI: 4.84, 9.30) in Europe ($P = 0.0015$). The prevalence of blepharospasm was not significantly different between the Japanese and European studies ($P = 0.3697$).

For several forms of dystonia, data did not permit calculation of sex-specific prevalence. Therefore, we calculated overall prevalences for limb dystonia (1.24 per 100,000; 95% CI: 0.35–4.36; $I^2 = 96.4\%$; $Q = 110.9$; $df = 4$; $P < 0.0001$), writer's cramp (1.65 per 100,000; 95% CI: 0.89–3.04; $I^2 = 83\%$; $Q = 17.7$; $df = 3$; $P = 0.0005$), oral mandibular dystonia (0.52 per 100,000; 95% CI: 0.15, 1.77; $I^2 = 89.7\%$; $Q = 39$; $df = 4$; $P < 0.0001$), laryngeal dystonia (1.54 per 100,000; 95% CI: 0.65–3.61; $I^2 = 94.1$; $Q = 67.5$; $df = 4$; $P < 0.0001$), and generalized dystonia (0.44 per 100,000; 95% CI: 0.26–0.74; $I^2 = 46.9\%$; $Q = 5.6$; $df = 3$; $P = 0.13$).

Discussion

The prevalence of dystonia reported on in the three population-based studies[14–16] in our analysis is much higher than in the service-based studies. The latter numbers are likely underestimates, because the numbers of patients seeking treatment is almost certainly lower than the prevalence of primary dystonia in the general population. Müller et al.'s Bruneck[16] study provides some evidence for this: Of the 6 patients identified, only 2 had been previously diagnosed with dystonia. Additionally, patients who have previously been diagnosed with dystonia, but who have stopped treatment or were last examined before the capture dates, may be missing in service-based estimates.

The population-based studies[14–16] are not without limitations. One of the principal limitations of the two

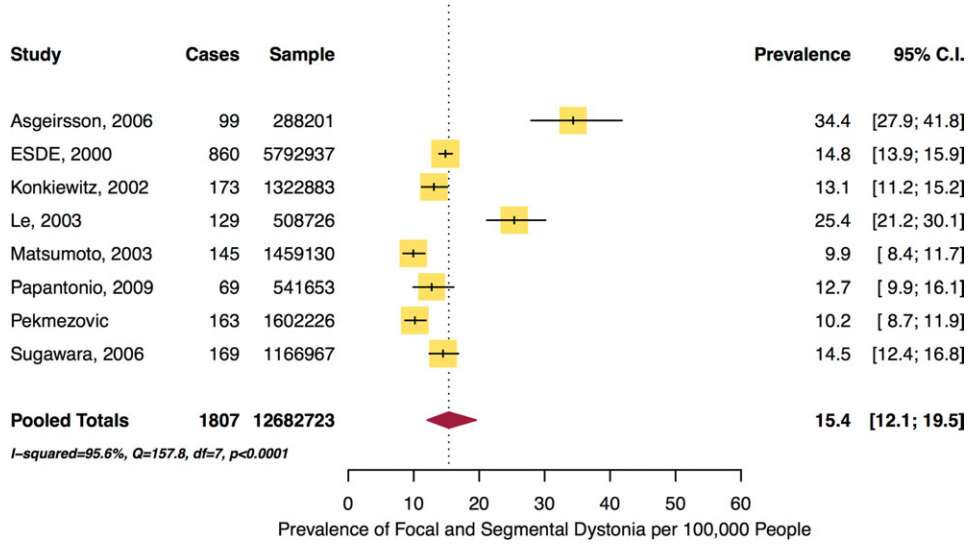

FIG. 2. Prevalence of focal and segmental dystonia (per 100,000).

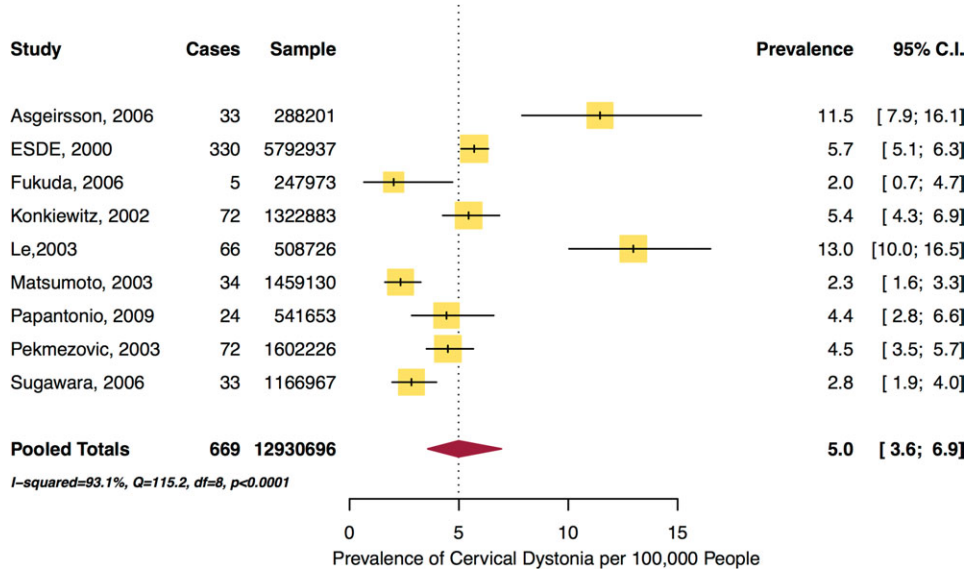

FIG. 3. Prevalence of cervical dystonia (per 100,000).

door-to-door studies[14,15] was the relatively low sensitivity of the initial screening. The prevalence of dystonia could have been higher if neurologists had examined all subjects, thus capturing milder cases.

Such limitations were not a feature of the population-based study performed in Bruneck,[16] which used a very different methodology and which may explain why this study reported the highest prevalence.[16] The study was based on a relatively small total population of 13,704 participants and a limited number of cases that generated wide CIs. However, even under the impossible assumption of zero prevalence in the population below age 50, the calculations based on the observed cases for the age range of 0 to 90 years still produce a prevalence of 220 per 100,000 (95% CI: 98–481). Even the lower limit of this estimate clearly exceeds the prevalence reported on in all service-based studies. The investigators defended their estimates by contrasting their rigorous approach to that of the service-based studies, as well as to the published population-based studies that have relied on initial screening conducted by non-neurologists without stringent diagnostic criteria. Given that a definite diagnosis was only accepted when three movement disorders specialists independently diagnosed dystonia upon video review, there was a low likelihood of false positives.

Our meta-analysis of the service-based studies generated a relatively narrow CI for the prevalence of primary dystonia: 16.43 per 100,000 (95% CI: 12.09–22.32). However, a notable feature on review of these

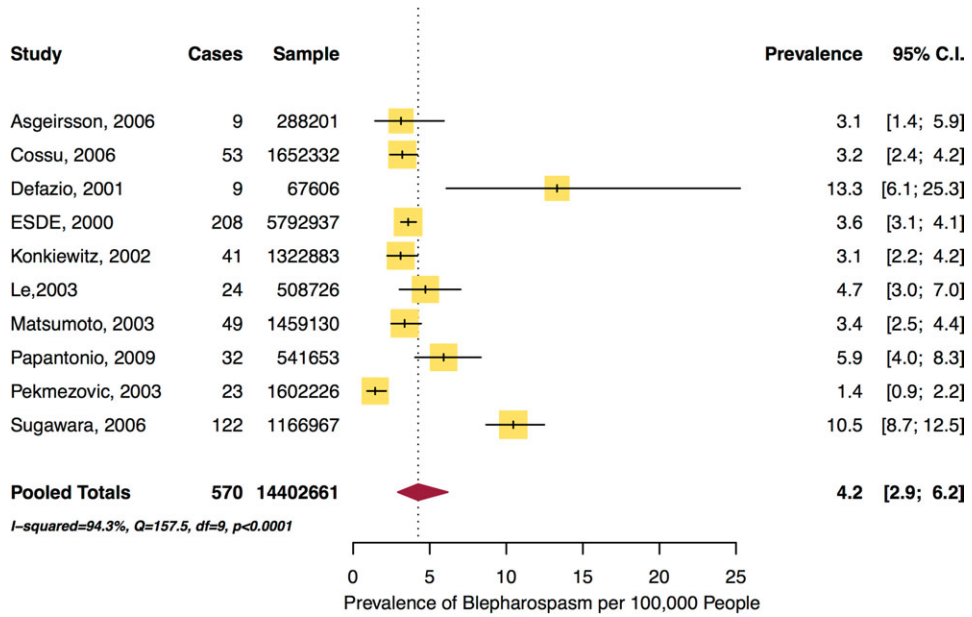

FIG. 4. Prevalence of blepharospasm (per 100,000).

studies was the great variation of reported estimates of prevalence across Europe. No good evidence exists for such geographic variation, and the reported prevalences differ to such an extent that uneven success in identifying patients seems the most plausible explanation. For example, the study of dystonia in Iceland yielded the highest prevalence of primary, focal/segmental, and cervical dystonia. Although genetic factors could be responsible for the higher prevalence of dystonia, the higher prevalence can also be readily attributed to a more-complete ascertainment of cases. Iceland's small population of 288,201 people is served by neurologists in only one neurological department in a hospital in the capital and a single neurologist in a regional hospital—and both hospitals participated in the study. The study also employed many other methods of identifying cases, including contacting other specialists who might treat patients with dystonia, review of hospital medical records, and the records of the hospital's botulinum toxin outpatient practice.

Differences in ascertainment may also explain the differences in reported frequencies of dystonia subtypes across countries, in particular, comparing Europe and Japan. Matsumoto et al.'s study of patients in Kyoto reported that blepharospasm was more prevalent than cervical dystonia, which is opposite to the pattern demonstrated in most European populations.[17] Although the reported prevalence of blepharospasm in Kyoto was similar to those reported in Italy, Matsumoto et al.[17] noted, at the time of their study in 2003, that ophthalmologists in Japan were only just beginning to provide treatment for blepharospasm, despite this being common practice elsewhere, and that this could have increased the prevalence of the condition noted in neurology clinics in Japan. Likewise, Fukuda et al., reporting in their 2006 study that the prevalence of facial dystonia was approximately four times higher than in the previous survey of Tottori prefecture in 1993 (6.5 versus 1.6 per 100,000),[18,19] stated that this finding probably related to the increased number of patients being evaluated, diagnosed, and treated since the introduction of botulinum toxin in 1997. They noted that, in contrast, the prevalence of torticollis did not differ from that observed in their 1993 survey; whereas that of writer's cramp increased by approximately three times.

In general, we can conclude that the studies appearing to show differences between populations in the prevalence of dystonia and dystonia subtypes do not allow a distinction to be made between the effects of genetics, heterogeneous diagnostic criteria, and healthcare-seeking patterns.

Overall, our study documented increasing prevalence of dystonia with age (Table 2), and some evidence suggests that the age of emergence of dystonia may also be partially ethnically related, because the study from Oslo documented later onset of disease in 719 non-Jewish Caucasians than in Ashkenazi Jews and later onset in ethnic Europeans than in first-generation Asian and African immigrants.[20] In general, the literature suggests that men develop dystonia at an earlier age and have a shorter time to diagnosis,[13,17,21] suggesting a sex-related biologic difference that may reflect greater severity of disease in males. Our own metaregression analysis did not reveal sex-related differences in prevalence.

Conclusion

In conclusion, despite the publication of studies on the epidemiology of dystonia, attempting to determine an accurate prevalence of the condition for health-service planning remains a challenge. Given the methodological limitations of the studies included in our own meta-analysis, it is likely that the combined prevalence of primary dystonia of 16.43 per 100,000 is an underestimate, with many cases remaining undiagnosed. More-accurate assessments of prevalence could be obtained with population-based studies employing rigorous assessments that would necessitate a substantial commitment of resources; however, the dystonias are a source of significant disability, lost productivity, and impaired quality of life. In view of the effective treatments that are now available for the condition, increased awareness of dystonia among both health professionals and the public is warranted to ensure appropriate healthcare planning and the provision of treatment.

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APPENDIX

Medline Search Strategy

1 exp Dystonia/
2 exp Torticollis/
3 1 or 2
4 exp Incidence/
5 exp Prevalence/
6 exp Epidemiology/
7 4 or 5 or 6
8 3 and 7
9 *Dystonia/ep [Epidemiology]
10 *Torticollis/ep [Epidemiology]
11 9 or 10
12 8 or 11
13 limit 12 to yr=”1985 –Current”
14 limit 13 to animals
15 13 not 14