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Cervical Dystonia Incidence and Diagnostic Delay in a Multiethnic Population

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Correction added December 6, 2019, after first online publication: The author byline and affiliation for Raymond Lo has been updated to include middle initial and name of university.

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Abstract

Background: Current cervical dystonia (CD) incidence estimates are based on small numbers in relatively ethnically homogenous populations. The frequency and consequences of delayed CD diagnosis is poorly characterized.

Objectives: To determine CD incidence and characterize CD diagnostic delay within a large, multiethnic integrated health maintenance organization.

Methods: We identified incident CD cases using electronic medical records and multistage screening of more than 3 million Kaiser Permanente Northern California members from January 1, 2003, to December 31, 2007. A final diagnosis was made by movement disorders specialist consensus. Diagnostic delay was measured by questionnaire and health utilization data. Incidence rates were estimated assuming a Poisson distribution of cases and directly standardized to the 2000 U.S. census. Multivariate logistic regression models were employed to assess diagnoses and behaviors preceding CD compared with matched controls, adjusting for age, sex, and membership duration.

Results: CD incidence was 1.18/100,000 person-years (95% confidence interval [CI], 0.35–2.0; women, 1.81; men, 0.52) based on 200 cases over 15.4 million person-years. Incidence increased with age. Half of the CD patients interviewed reported diagnostic delay. Diagnoses more common in CD patients before the index date included essential tremor (odds ratio [OR] 68.1; 95% CI, 28.2–164.5), cervical disc disease (OR 3.83; 95% CI, 2.8–5.2), neck sprain/strain (OR 2.77; 95% CI, 1.99–3.62), anxiety (OR 2.24; 95% CI, 1.63–3.11) and depression (OR 1.94; 95% CI, 1.4–2.68).

Conclusions: CD incidence is greater in women and increases with age. Diagnostic delay is common and associated with adverse effects.

Keywords

cervical dystonia; diagnostic delay; incidence

Cervical dystonia (CD) is among the most common forms of focal dystonia in the neurology clinic.^{1–9} However, the true frequency of CD is not established.^{10,11} Most estimates of CD prevalence are based on referral clinic populations.^{2–5,7,12–14} There are many advantages to studying population incidence, including the minimization of bias. Previous studies estimating CD incidence are based on small numbers of cases in homogenous populations or rely on decades-old data that may now poorly represent increased awareness of dystonia among medical providers.^{15–17} The aims of this study were to determine the incidence of CD in a large, ethnically diverse population, identify the characteristics associated with a delayed diagnosis of CD, and delineate possible risk factors for developing CD.

Methods

Study Population and Data Extraction

Kaiser Permanente Northern California (KPNC) is an integrated healthcare system with more than 3 million members that serves a diverse population across 14 Northern Californian counties. Based on data from the 2003 California Health Interview Survey, Kaiser Permanente member demographics are similar to the non-Kaiser Permanente population.¹⁸

Cases and controls were ascertained through a review of the comprehensive clinical and administrative data-bases of KPNC. The multistage case identification process used by this study is detailed in Figure 1 and is based on our preliminary work.¹⁵

Case Selection

Incident cases of CD were identified through a 3-stage case identification procedure. First, inpatient and outpatient visits and pharmacy records for all KPNC and non-KPNC services were screened for specific diagnostic codes and write-in diagnoses for primary dystonia. Second, for all individuals with a CD diagnosis, a complete chronological list of all health service utilizations from the beginning of KPNC membership was generated for each individual. Each individual's list was reviewed by 1 or more movement disorders specialists to classify individuals as likely or unlikely to have CD. Third, individuals judged likely to have CD underwent a more extensive review by a movement disorders expert, including when possible a neurological examination in person or via review of a standardized videotape data collection by a trained research assistant during a home visit. Research diagnostic criteria appropriate to epidemiologic studies were developed by an expert panel convened by the Dystonia Medical Research Foundation (including C.M.T., S.B., and C.C.) and operationalized for use in KPNC. A final diagnosis of CD required (1) a diagnosis by a neurologist, (2) no consistent change in neurologist's diagnosis after index date, (3) no antecedent neuroleptic use, and (4) no identifiable associated medical or neurologic disorder known to include dystonia. Individuals were excluded if their dystonia was diagnosed prior to January 1, 2003 (i.e., prevalent cases) or after December 31, 2007, or if they were not members of the KPNC system at the time of CD diagnosis.

Medical records review, videotaped exam review, and clinical examinations conducted by movement disorders experts in subsets included characterization of the abnormal posture, the presence or absence of superimposed movements, presence or absence of geste antagoniste, presence or absence of associated features, type of therapy, and response to therapy as well as absence of another cause for the movement abnormality.

In these validation subsets, agreement between utilization-defined CD and medical record review was 88%, and between utilization defined CD and examination was 96%.

Control Selection

Two control populations were assembled. For analyses based on the electronic medical record, control participants were identified to determine the relative risk of clinical

diagnoses preceding a diagnosis of CD. A total of 10 controls were selected at random from the entire KPNC member base and matched to each case on birth year, sex, membership length, facility, and zip code (as a proxy for socioeconomic status). Electronic medical record (EMR) information was then pulled from the KPNC databases. For analyses based on interview data, control participants were selected at random from the KPNC member base and matched 1-to-1 to enrolled cases based on birth year, sex, and membership length. If a control did not complete the entire interview, another matched control was selected for the participating case.

Data Acquisition

Data were acquired from the review of the EMR, health services utilization reports, and a mailed questionnaire. Information regarding incidence and antecedent clinical diagnoses was obtained from the EMR. The patient questionnaire provided detailed information on the frequency, character, and impact of diagnostic error in persons with CD as well as data regarding family history and demographics.

Standard Protocol Approvals, Registrations, and Patient Consents

This study was conducted with approval from the KPNC institutional review board. Informed consent was obtained from each participant.

Statistical Analysis

All statistical analyses were conducted using the SAS/STAT statistical software package, version 9.1 (SAS Institute Inc., Cary, NC). Average annual incidence was calculated overall and for the sex-specific, race/ethnicity-specific, and age-specific groups.¹⁹ Incidence rates and confidence intervals were estimated assuming a Poisson distribution of cases within the Kaiser Permanente population. Direct standardization of rates for age, race/ethnicity, and sex used the 2000 U.S. census population. Incidence rates were presented per 100,000 person-years.

Behaviors and antecedent diagnoses were compared between CD cases and their interviewed controls (1:1) or EMR control (1:10). Adjusted odds ratios (ORs) and 95% confidence intervals (CIs) were calculated adjusting for birth year, sex, membership length, facility, and zip code (as a proxy for socioeconomic status).

Results

CD Incidence and Participant Demographics

A total of 200 cases of incident primary CD were identified during 15,489,433 person-years of observation. Incidence standardized to the U.S. 2000 census population was 1.18 per 100,000 person-years (95% CI, 0.35–2.0). The age-specific, sex-specific, and race/ethnicity-specific rates are summarized in Table 1. CD incidence was significantly greater in women and whites. The difference between mean age at incident diagnosis between men and women approached significance (men = 47 years [95% CI, 42.29–51.85], women = 54 years [95% CI, 51.61–56.43]). Incidence appeared to increase with age through the seventh decade Figure 2.

CD Diagnostic Delay

Of the 200 CD cases, we had permission to contact and approach 114 cases. A total of 80 cases agreed to respond to questionnaires concerning risk factors, diagnosis, and treatment (70%); of those, a total of 58 cases (51%) responded to the survey addressing details of diagnosis. Diagnosis of CD was delayed a median of 730 days after the onset of self-reported symptoms; a median of 3 physician visits occurred before the diagnosis was made. Characteristics of diagnostic delay derived from the patient questionnaire are outlined in Table 2. Of the patients, 50% reported unpleasant or harmful effects as a result of this delay. Of those surveyed, 66% reported receiving treatment for an incorrect diagnosis prior to being diagnosed with CD. The most common treatments included muscle relaxants, physical therapy, anti-inflammatories, acupuncture, antidepressants, and trigger point injections. A quarter of those who received treatment prior to CD diagnosis reported unpleasant or harmful side effects from those treatments. Following CD diagnosis, 83% of patients received botulinum toxin injections; of these, 89% reported that botulinum toxin injections were helpful. The features associated with diagnostic delay are summarized in Table 3.

Medical Conditions Preceding CD Diagnosis

Patients ultimately diagnosed with CD were more likely to be given certain diagnoses when compared with controls. These diagnoses included essential tremor (OR 68.1; 95% CI, 28.2–164.5), cervical disc disease (OR 3.8; 95% CI, 2.83–5.17), neck sprain/strain (OR 2.68; 95% CI, 1.99–3.62), anxiety (OR 2.24; 95% CI, 1.63–3.11), and depression (OR 1.94; 95% CI, 1.4–2.68). Patients with CD were significantly more likely to have a diagnosis of neck trauma or pathology 3 and 5 years preceding the index date when compared with controls ($P = 0.000$ and $P = 0.002$, respectively). Three years prior to diagnosis, patients with CD were significantly more likely than controls to be diagnosed with tremor, essential tremor, depression, and anxiety ($P < 0.001$ for each), but not migraine ($P = 0.577$).

Discussion

In this study of CD in patients within the KPNC system, we found an estimated CD incidence of 1.18/100,000 person-years based on 200 cases over 15.4 million person-years. This study incorporates the largest, most ethnically diverse cohort of incident cases of CD to date. Our estimated incidence of 1.18 per 100,000 person-years is similar to the preliminary incidence estimate we previously published for the years 1997 to 1999 (1.07 per 100,000 person-years).¹⁵ This current report is based on 3 times as many incident cases of CD, more than twice as many person-years of observation, compared with our prior estimate. Because of the larger sample size, we are now able to evaluate differences among subgroups with more confidence and precision.

We found that the incidence of CD differs by sex. The incidence of CD was 3.5 times greater for women than for men. This strengthens our previous finding that the minimum estimated incidence of CD was 2.5 times greater for women.¹⁵ The majority of prior studies estimated CD prevalence, not incidence; these studies found sex prevalence ratios (female:male) ranging from 1.1–3.6:1.^{4,7,16,20} However, unlike our study, these data were mostly derived

from homogenous populations or from a small number of centers.²⁰ Although the incidence of CD is higher for women, the age of CD onset may be later for women than for men. The sex difference observed in our study may have several explanations. One possibility is rooted in genetics. Although most cases of CD have no known cause, a family history of dystonia remains an important risk factor.²¹ There are genetic links for many forms of dystonia, but these are not usually inherited on sex chromosomes.²² However, our observation that the incidence of CD differs by race also raises the possibility of genetic contributions. Currently identified CD-related genetic mutations and variants are not sufficient to explain the observed sex difference, although variable penetrance, epigenetic modification of gene expression, or gene–environment interactions may play a role. Differences in sex hormones are another possible explanation for this sex discrepancy. Rodent studies suggest that estrogen may protect the nigrostriatal dopaminergic system from damage through enhancing dopamine synthesis, increasing dopamine receptor sensitivity, and stimulating plasticity in this circuit.^{23–26} The decrease in estrogen during the perimenopausal and postmenopausal periods may be a driver of the observed increase in CD incidence for women later in life when compared with men.^{4,7,13} Sex differences may also result from differences in health-seeking behaviors or medical provider diagnostic bias. These could affect both the overall sex-specific incidence rates as well as apparent differences in age distribution. Lastly, these observations may also be the result of exogenous risk factors, such as occupation. Additional study of genetic, molecular, and environmental factors is necessary to better understand the relationship of sex to the development of CD.

Diagnostic delay is an important barrier to appropriate treatment for patients with dystonia, but few studies have characterized the magnitude of this obstacle. Our study suggests a greater percentage of misdiagnosis than prior studies, with 50% of patients endorsing harmful effects as a result of the delay.^{3,27,28} The consequences of diagnostic delay can be severe and include unnecessary healthcare costs, such as inappropriate testing and treatment. There are many reasons for CD delayed diagnosis, including mild or slowly progressive symptoms, fluctuation of symptoms not seen during a physician visit, focus by the physician on managing pain rather than the underlying cause of pain, and misdiagnosis as a psychogenic illness.^{28,29} Even in healthcare systems with equal access to care, such as KPNC, delayed diagnosis remains an important barrier. Given the significant improvement in health-related quality of life in patients who receive botulinum toxin therapy for dystonia, it is imperative that physicians maintain a high level of suspicion for CD when examining a patient with neck muscle spasm.^{30,31} Possible interventions to mitigate diagnostic delay include more robust education on diagnosing dystonia in medical school or through continued medical education courses.

Participants in this study were given multiple diagnoses preceding the diagnosis of CD. These alternate diagnoses may reflect comorbid conditions, diagnostic errors, or etiologic factors. For example, depression and anxiety could represent a misattribution of CD symptoms to a psychogenic cause or may reflect the prominent mood disorders that can accompany a disabling neurological disease.^{28,32} Potential triggers of CD in the literature include peripheral trauma, such as neck sprain or whiplash.^{21,33–35} We found that patients diagnosed with CD were significantly more likely to be diagnosed with cervical disc disease or neck trauma preceding CD diagnosis when compared with controls. A relationship

between peripheral trauma and development of CD has been previously observed,^{35–37} such as in one case-control study where 16 of 95 respondents reported a history of injury within 4 weeks of CD development.³⁸ The pathophysiology of a peripherally—rather than centrally—mediated association remains unclear, and there continues to be debate regarding its existence.³⁹ One possibility is a gene–environment interaction, where the peripheral trauma incites development of CD in someone with a genetic predisposition.^{40,41} Aberrant peripheral inputs—such as from trauma—may provoke reorganization of neuronal circuitry more centrally.^{41,42} There is the suggestion that patients with dystonia have aberrant synaptic plasticity mediated by acetylcholine resulting in altered basal ganglia networks.⁴³ It is also possible that pain secondary to CD resulted in neck imaging that revealed cervical disc disease, which is common in the general population, and so this radiographic finding may be present but not contributory. Overall, the relationship between peripheral trauma and dystonia remains a challenging topic to study because of recall bias and cause-effect bias, for example, unrecognized dystonia may result in injury rather than the converse. A larger prospective study is warranted to better elucidate this potential association.

This investigation includes several strengths. We chose to minimize survival bias and differences in diagnostic practices by studying incident cases of CD during a specified 5-year period rather than all prevalent CD cases. Our strict case definition and physician review of all suspected cases minimized the false–positive rate. In addition to its large size, there are many advantages to studying CD in the KPNC population. All Kaiser Permanente members receive essentially equal access to healthcare and membership is precisely enumerated. Members benefit from equal access to expert care, including movement disorders specialists. The multiethnic KPNC cohort is demographically similar to the non-Kaiser Permanente population,¹⁸ maximizing the generalizability of our findings. Centralized data collection within the Division of Research that spans across the Kaiser Permanente organization is another strength.

This study has several limitations. We did not conduct population-wide examinations, so individuals never diagnosed are not included in our study. This may result in an underestimation of CD incidence. The sample sizes for racial groups other than whites were small and the categories were broad—for example, the classification of Asian within KPNC includes persons from multiple regions including the Indian subcontinent and the Pacific Islands—limiting our conclusions on racial differences. Lastly, our questionnaire characterizing diagnostic delay, mailed to the 114 of the 200 CD cases we had permission to contact, was only answered by 58 cases. This is a potential source of bias as there is a possibility of overestimating or underestimating the burden of delay.

In conclusion, CD is a disease with a female and white preponderance, with an incidence that increases with age. Despite increased awareness of this common movement disorder, there remain significant delays with diagnosis and treatment for patients with CD. Quality of life can greatly improve with simple therapies. Education of primary care physicians and general neurologists on the recognition of CD and screening tools with increased sensitivity and specificity are needed to assist with making an accurate and timely diagnosis in this vulnerable patient population.

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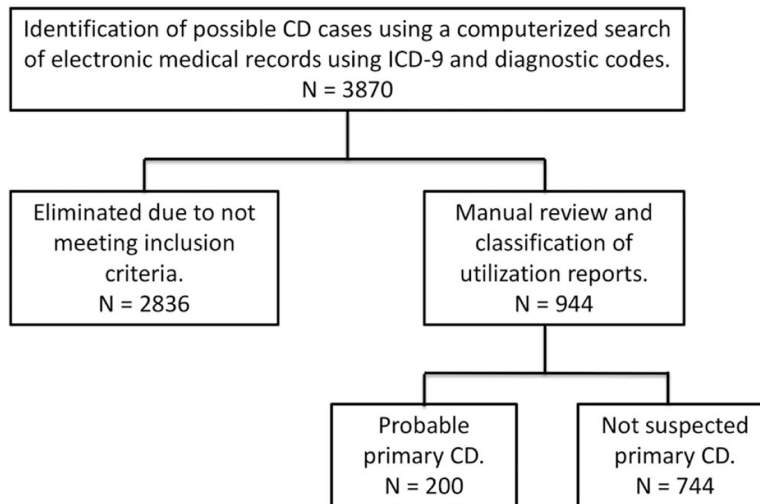


FIG. 1. Case identification process for patients with cervical dystonia (CD) obtained from a multiethnic membership of a health maintenance organization in Northern California.

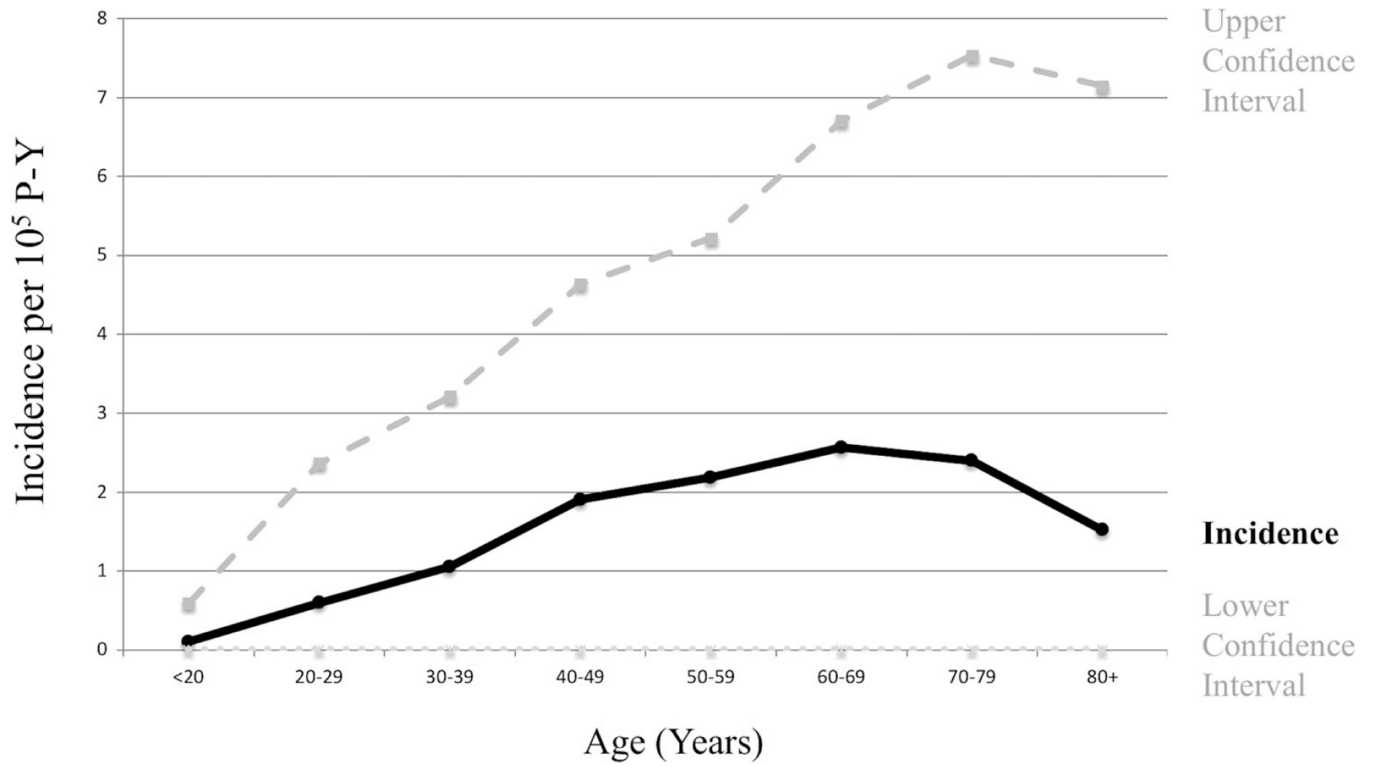


FIG. 2. Incidence of cervical dystonia by decade standardized to the U.S. population during the year 2000. P-Y, person-years.

TABLE 1.

Demographics of patients with CD derived from the Kaiser Permanente electronic medical record

Demographic	Total Patients	Incidence per 100,000 Person-Years	LCI	UCI
All patients	200	1.18	0.35	2.00
Sex				
Male	42	0.52	0.13	0.91
Female	158	1.81	1.08	2.53
Age, y				
<20	4	0.10	0	0.59
20–29	11	0.60	0	2.37
30–39	23	1.06	0	3.21
40–49	47	1.91	0	4.63
50–59	50	2.19	0	5.22
60–69	37	2.57	0	6.70
70–79	21	2.40	0	7.54
80+	7	1.52	0	7.15
Racial category				
African American	10	0.62	0	2.51
Asian	20	0.46	0	1.50
White	167	1.56	0.36	2.76
Native American	2	0.93	0	7.41
Other	1	0.13	0	1.45
Ethnic category				
Hispanic	28	0.97	0	2.82
Non-Hispanic	172	1.21	0	2.13
Sex	Total	Mean Age, y	LCI	UCI
Male	42	47.07	42.29	51.85
Female	158	54.02	51.61	56.43

CD, cervical dystonia; LCI, lower confidence interval; UCI, upper confidence interval.

TABLE 2.

Characteristics of diagnostic delay in patients ultimately diagnosed with CD derived from patient questionnaire

Characteristic	Mean \pm SD	Median
Days between symptom onset and CD diagnosis	1674 \pm 2543	730
Number physicians seen prior to CD diagnosis	4 \pm 5	3
Number of diagnoses prior to CD diagnosis	Frequency	Percent
0	20	35
1	21	36
2	7	12
3	5	9
4	3	5
> 4	2	3

CD, cervical dystonia; SD, standard deviation.

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TABLE 3.

Consequences of diagnostic error derived from patient questionnaire

Consequence of Diagnostic Error	Total Patients	Percent
Patients with unpleasant/harmful effects because of delay in CD diagnosis, N = 52		
Yes	26	50
No	14	27
No delay in diagnosis	12	23
Unpleasant/harmful effects from delay in CD diagnosis, N = 26		
Any unpleasant/harmful effect	26	100
Personal worry/anxiety	22	88
Family problems	12	50
Friendship problems	11	48
Job problems	15	65
Emotional well-being	23	89
Problems with recreational activities	17	74
Unpleasant/harmful effects from treatment received prior to CD diagnosis, N = 36		
Total patients	9	25
Impact of correct CD treatment, N = 58		
Work resumption	17	29

Total number of patients who answered each question differ as a result of patient participation.

CD, cervical dystonia.