E-METHODS

The methods we used for this study rely heavily on the approach our group developed for previous studies of neurodegenerative disease. The Indian Health Service (IHS) provides and collects data on healthcare for members of federally recognized American Indian and Alaska Native tribes. Of eight Navajo healthcare Service Units, five are operated by the IHS and three by the Navajo government. The study data, obtained from the IHS National Patient Information Reporting System, include all hospital discharge and outpatient visit records from IHS- and tribally-operated medical facilities in the Navajo Nation, as well as facilities and providers that contract to provide healthcare services. This study followed a pilot epidemiological study of Parkinson disease in a single Service Unit.

We endeavored to determine the average annual incidence and point prevalence rates of Huntington disease among Navajo people receiving IHS or tribal health care during 2001–2011. The unit of analysis for this study was the patient. Data for electronic inpatient and outpatient visit records with the International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM) code for Huntington disease (333.4) listed as one of up to 15 diagnoses were selected for eligible people. Patients with Huntington disease were eligible to have their records included in the analysis if they were enrolled, resident members of the Navajo Nation, and had a visit to a tribal or IHS clinical facility in the Navajo Nation. To be a member of the Navajo Nation a person must have ¼ degree blood quantum or the equivalent of one Navajo grandparent,
For retrospective incidence estimates, cases were required to have at least two Huntington disease-associated inpatient or outpatient visits during 2002–2011 with no such visits during 2001. Point prevalence estimates were calculated as of the midpoint in the study period, July 1, 2006, and included all patients with at least two inpatient or outpatient visits that listed the code for Huntington disease during 2001–2011, with at least one visit before July 1, 2006, and who were determined to be alive on July 1, 2006 by having any visit in the IHS healthcare system between July 1, 2006 and December 31, 2011.1,2 This approach was taken because death certificate and IHS data are not linked. Two visits were used in the case definition to reduce the impact of single coding errors.7 The population used for the prevalence rate was estimated by calculating the total number of Navajo people with an inpatient or outpatient visit record within the IHS/tribal health care system on or before July 1, 2006 and at least once after July 1, 2006.

The results of the primary analysis were unexpected so we conducted three post hoc analyses using the IHS data. First, we determined the number of cases and estimated the prevalence rate in AI/AN people living on or near reservations across the United States during the same time period (2001-2011). Second, we examined Navajo Nation inpatient records to determine whether any Huntington disease-associated visits occurred during from 1980–2000. Patient identifiers were unavailable for this earlier time period so only individual visits could be examined. Finally, we estimated the probability of finding no cases in the Navajo Nation on prevalence day, July 1, 2006, under different rate assumptions. The probability was calculated using the binomial distribution as the chance of 0/217,158 people having HD (or equivalently the chance of 217,158 of 217,158 people not having HD) under the assumption that the underlying
prevalence rate for Navajo people is the same as for the reference population and the probabilities of each patient having Huntington disease (HD) are independent of one another (e.g. for a reference prevalence rate of 5.7/100,000 the calculation of probability was: \(1 - \frac{5.7}{100,000}\)).

The analyses include all Navajo people who received direct or Purchased/Referred Care from the IHS/tribal health care system during the study period. Because of the isolation of the Navajo Nation and the uncommon stability of the population, the approach captured 98% of eligible Navajo. The Navajo Nation Human Research Review Board granted study approval.

To enhance ascertainment, the lead author on this paper (PHG), who is also a movement disorder specialist, and, any given time, the only neurologist in the Navajo Nation, made additional inquires: He himself consults on many difficult cases, by telephone or in person, from across the reservation. In seven years of practice including consultation, travel and speaking engagements at every hospital and community government seat reservation-wide, he has not encountered any Navajo people with Huntington disease. For this study, he also contacted regional Huntington’s clinics at the University of New Mexico, which he helped found in 2001, and at the University of Arizona, learning that no Navajo people were enrolled.

Supplement References


