disease prevention news

August 26, 2002 www.tdh.state.tx.us/phpep/ Vol. 62 No. 18

Creutzfeldt-Jakob Disease Mortality in Texas, 1995-2001

The first case of suspect new variant CJD in the United States was identified in Florida in April 2002. This person is a 22-year-old United Kingdom (UK) citizen residing in the United States. Investigators concluded that the disease was contracted in the UK. In Texas multiple information sources, including death certificate reviews, revealed that all cases identified for years 1984-2001 have been classic, sporadic CJD.

The neurological disorder, Creutzfeldt-Jakob disease (CJD), is a transmissible spongiform encephalopathy (TSE) thought to be caused by prions, proteinaceous particles devoid of nucleic acid.[1,2] Classic CJD, the most common form, occurs sporadically worldwide in people over age 60. It is characterized by rapidly progressive dementia accompanied by severe muscle spasms and incoordination.[3] The incidence of classic CJD is 1 case per million population per year. [3] Death usually occurs within 12 months after onset with an average of 8 months.[3] From 5% to 10% of patients have a less rapid course and survive more than 2 years. [4] Classic CJD has no known route of acquisition and no known treatment. Case definitions of the other three CJD types: iatrogenic, familial, and new variant (nv) CJD are listed on the following page.

The Centers for Disease Control and Prevention (CDC) has been studying CJD deaths in the United States since the identification of a new form of human CJD (nvCJD) first described in the United Kingdom in 1996. Variant CJD has been linked to bovine spongiform encephalopathy (BSE) also known as "mad cow disease."[5,6] Variant CJD differs from classic CJD in a number of aspects: it affects younger individuals, has a shorter clinical course, has an abnormal but not typical EEG, and has brain histopathology that is different but recognizable as CJD. The first likely case of new variant CJD was identified in the United States in Florida in April 2002. [7] This person is a 22-year-old United Kingdom (UK) citizen residing in the United States. Investigators concluded that the disease was contracted in the UK. [7] According to the April 18, 2002, CDC press release, 125 persons with nvCJD have been identified worldwide. Most of these patients had multiple-year exposures in the UK from 1980 through 1996 during the occurrence of the BSE outbreak in cows. [7]

Texas Surveillance: Medical records are requested and reviewed for any Texas death reported or identified as CJD through death certificate reviews . The review allows classification by subtype and degree of confidence in the diagnosis. The types are sporadic, familial, iatrogenic, and variant CJD; the degree of confidence is expressed as definite, probable, possible (suspect), or not a case. [8]

Table 1. Age Distribution of Confirmed Cases of CJD in Texas, 1995-2001

Age Group	1995	1996	1997	1998	1999	2000	2001	Total
<55	2	1	2	1	2	3	0	**11**
≥55	10	15	11	6	7	5	7	**61**
Total	**12**	**16**	**13**	**7**	**9**	**8**	**7**	**72**

Both definite and probable cases are included in reports to CDC and to estimate incidence rates for Texas.

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In Texas an earlier review of surveillance reports and death certificates from 1984 through 1994 revealed that all patients who had CJD had classic, sporadic disease.[9] Likewise, there have been no reports of nvCJD, familial, or iatrogenic disease from January 1, 1995, through December 31, 2001. Reports confirmed that 72 Texas residents died from CJD during this 7-year period: 38 cases met definite CJD case criteria, and 34 met probable CJD case criteria. The average crude death rate was 0.5 deaths per million population per year.[10] The age distribution and distribution by public health region are shown in Tables 1 and 2.

Case Definitions

Definite CJD: Neuropathologically confirmed; and/or immunocytochemically confirmed, PrP positive (Western blot) and/or scrapie associated fibrils; the findings of spongiform encephalopathy in cerebral and/or cerebellar, and/or subcortical grey matter, and/or encephalopathy with prion protein (PrP) immunoreactivity.

Probable CJD: Progressive dementia, typical periodic high-voltage complexes on electroencephalogram (EEG), and at least 2 of the following clinical features: myoclonus, visual disturbance, cerebellar disturbance, pyramidal dysfunction, extrapyramidal dysfunction, and/or akinetic mutism. The duration of illness (onset of illness to time of death) was less than 2 years.

Possible CJD: Same as probable CJD but without an EEG or with an EEG finding not typical for CJD and a duration of the illness less than 2 years.

Iatrogenic CJD: Progressive cerebellar syndrome in a pituitary hormone recipient; CJD with a recognizable exposure risk, eg, dura mater transplant.

Familial CJD: Definite or probable CJD plus definite or probable CJD in a first-degree relative; neuropsychiatric disorder plus disease-specific precursor protein mutation.

Definite New Varient CJD: Neuropathologically confirmed and/or immunocytochemically confirmed; numerous widespread kuru-type amyloid plaques surrounded by vacuoles in both cerebellum and cerebrum; spongiform change most evident in the basal ganglia and thalamus with sparse distribution throughout the cerebral cortex. Included is neuronal loss and focal astrocytosis and high density prion protein accumulation, particularly in the cerebellum and cerebrum.

Suspect New Variant CJD: Illness onset or death by age 55; illness duration of at least 6 months; a psychiatric presentation with anxiety, depression, social withdrawal and other behavioral changes with progression to neurological abnormalities such as progressive cerebellar syndrome, chorea, pyramidal and extrapyramidal signs; early onset of dysesthesias in limbs and face; forgetfulness and other memory impairment, dementia, and myoclonus in the late stages; an abnormal but not diagnostic EEG.

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Table 2. Distribution of Confirmed Sporadic CJD Cases by Public Health Region, 1995-2001

PHR	1995	1996	1997	1998	1999	2000	2001	Total
1	0	1	1	2	0	0	2	**6**
2	0	1	1	0	0	1	0	**3**
3	2	3	1	1	1	2	2	**12**
4	0	2	5	0	2	0	0	**9**
5	0	2	1	0	0	0	0	**3**
6	4	6	4	1	1	4	1	**21**
7	1	1	0	1	0	1	1	**5**
8	2	0	0	0	3	0	0	**5**
9	1	0	0	1	1	0	0	**3**
10	1	0	0	1	0	0	0	**2**
11	1	0	0	0	1	0	1	**3**
Total	**12**	**16**	**13**	**7**	**9**	**8**	**7**	**72**

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Additional information is available at the TDH website, www.tdh.state.tx.us/ideas/factsht/cjd.htm. *For information about national surveillance data or about how to obtain diagnostic services, visit the CJD website at* www.cjdsurveillance.com.

Erratum

In the July 29, 2002, printed issue of *DPN*, (Vol. 62 No. 16), the YTD totals were missing. These totals should be as follows: Measles, 0; Mumps, 9; Pertussis, 377; Rubella, 1, and Tetanus, 1. The Internet issue was corrected on July 31.

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The electronic version of *Disease Prevention News,* the subscription form, and a searchable index of issues from 1995 are available at the followingwebsite:
www.tdh.state.tx.us/phpep/

TDH Publication #E59-10940

West Nile Encephalitis : Human Cases in Texas

The date of onset of the first confirmed human case of West Nile encephalitis (WNE) in Texas was July 3, 2002. As of Monday, August 19 , 2002, 25 human cases of WNE have been reported from the following counties: Harris (13), Orange (5), Dallas (3), Montgomery (2), Jefferson (1), and Trinity (1 [acquired elsewhere]). West Nile virus (WNV) has been detected in humans, birds, mosquitos, and horses in 31 Texas counties, for a total of 318 cases. One human death in Harris County was confirmed Monday afternoon. The Texas Department of Health (TDH) conducts routine surveillance for mosquito-borne diseases and has been preparing for the appearance of WNV in Texas since the first human cases were reported in the northeastern United States.

Long before the virus was detected in Texas, TDH began to develop a WNV response plan designed to minimize human illness through vector identification, early disease diagnosis and vector control, geographic tracking, and public education. Current efforts include streamlining protocols for surveillance and intervention. Testing priority is currently focused on Texas counties where WNV has not yet been identified. All human cases with WNE symptoms and also laboratory results that confirm infection with WNV must be reported to TDH.

The April 22, 2002, *DPN* features an indepth report on WNV, which includes a history of the disease as well as symptoms of various arboviral encephalitides, differential diagnosis, diagnositic laboratory tests, instructions for laboratory submission of samples for testing, and primary prevention measures. Access this issue online at www.tdh.state.tx.us/phpep/dpn/issues/dpn62n09.pdf.

The TDH Zoonosis Control Division updates information on the current West Nile fever outbreak at this website: www.tdh.state.tx.us/zoonosis/diseases/Arboviral/westNile/westnile.asp. Also included on this website is the West Nile Virus Response Plan for Public Health Departments & Mosquito Control Districts.

For further information contact the Zoonosis Control division at 800/252-8239 (press 3) or 512/458-7255.