

# CREUTZFELDT-JAKOB DISEASE

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Creutzfeldt-Jakob disease (CJD) is a rare, degenerative neurological disease that affects humans and is always fatal. First identified in the 1920s, CJD belongs to a class of diseases known as transmissible spongiform encephalopathies or prion diseases. Prion diseases are widespread among animals and include scrapie in goats and sheep, and bovine spongiform encephalopathy (BSE) in cattle. The latter, known as “mad cow disease,” was identified in England in 1986, where it was attributed to the practice of feeding cattle meat-and-bone meal supplements made from scrapie-infected sheep or from rendered bovines already infected with BSE.<sup>1</sup> The consumption of BSE-infected beef has been associated with CJD.

## Variant CJD

There are four forms of CJD.<sup>2</sup> The form associated with the consumption of BSE-infected beef, the highly publicized *variant* Creutzfeldt-Jakob disease (vCJD), is extremely rare. Worldwide, most vCJD deaths have occurred in the United Kingdom, the focal point of the “mad cow disease” outbreak. To date, 139 people in the UK have died from vCJD. In addition, 7 people with probable cases are still alive.<sup>3</sup> The average age of onset of vCJD is 29, and the time between the appearance of symptoms and death is up to 14 months. There has been one vCJD death in Canada—a man who had been in the United Kingdom during the outbreak there.<sup>4</sup>

The risk of exposure to BSE-infected cattle has been far lower in Canada than in the United Kingdom. Three cases of BSE have been found in Canadian cattle; the first was a cow imported from Britain in 1987, the second, reported on May 20, 2003, was a cow raised in Alberta.<sup>5</sup> The most recent case, identified on December 23, 2003, was a cow in Washington State that had been born in Alberta.<sup>6</sup>

These isolated cases are in stark contrast to the United Kingdom, where 36,680 confirmed BSE

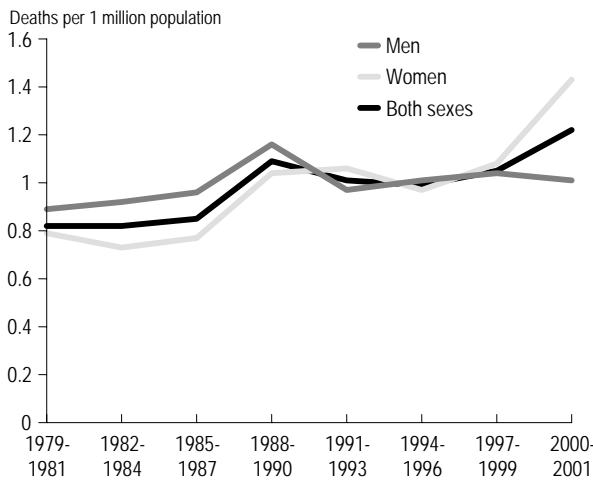
cases were reported in 1992 at the peak of the outbreak.<sup>7</sup> Although UK beef has not been imported into Canada for over 30 years, measures have been implemented to reduce the possibility of unrecognized BSE in Canadian herds:<sup>5</sup>

- limiting imports of live ruminants, meat and meat products to countries considered free of BSE;
- establishing BSE as a reportable disease in 1990;
- implementing a surveillance program in 1992 that has resulted in testing approximately 10,000 cattle brains for disease;
- banning the practice of feeding ruminant protein to other ruminants (cattle, sheep, goats, bison, elk, deer) since 1997; and
- introducing the Canadian Cattle Identification Program to ensure that the movements of all cattle and bison can be traced from birth to slaughter.

## Classical CJD

The three remaining forms of CJD—sporadic, familial and iatrogenic—are collectively known as *classical* Creutzfeldt-Jakob disease (cCJD). Sporadic CJD, for which the cause is unknown, accounts for 85% to 90% of cases. The familial, or hereditary, form comprises 10% to 15% of cases, while fewer than 1% are iatrogenic; that is, they result from medical examination or treatment. The iatrogenic form has been associated with corneal transplants, contaminated neurosurgical instruments, dura mater grafts, and a history of pituitary-derived (non-synthetic) human growth hormone use.<sup>2</sup> The age of onset for classical CJD is much older than that for variant CJD, averaging between 60 and 65. The time between exposure to the infection and the development of symptoms can extend from 1 to more than 30 years. Once symptoms appear, death rapidly follows, and most patients die in less than 6 months.

### Age-standardized mortality rates for Creutzfeldt-Jakob disease, by sex, Canada, 1979 to 2001



Data source: Vital Statistics Death Database

Note: Age-standardized to 1996 Canadian population; three-year averages for 1979 to 1999, two-year average for 2000 to 2001

## Deaths

Between 1979 and 2001, 599 deaths in Canada were attributed to Creutzfeldt-Jakob disease, only one of which was BSE-related. This was an annual average of 26 deaths, with the number ranging from a low

of 14 in 1979 to a high of 44 in 2001. More women than men died of CJD: 329 versus 270.

The CJD mortality rate rose slightly over the 1979-to-2001 period. The three-year average age-standardized mortality rates for men increased from 0.89 to 1.01 deaths per million, and from 0.79 to 1.43 per million for women. For both sexes together, the rate rose from 0.82 to 1.22 deaths per million population, which is consistent with rates in other countries.

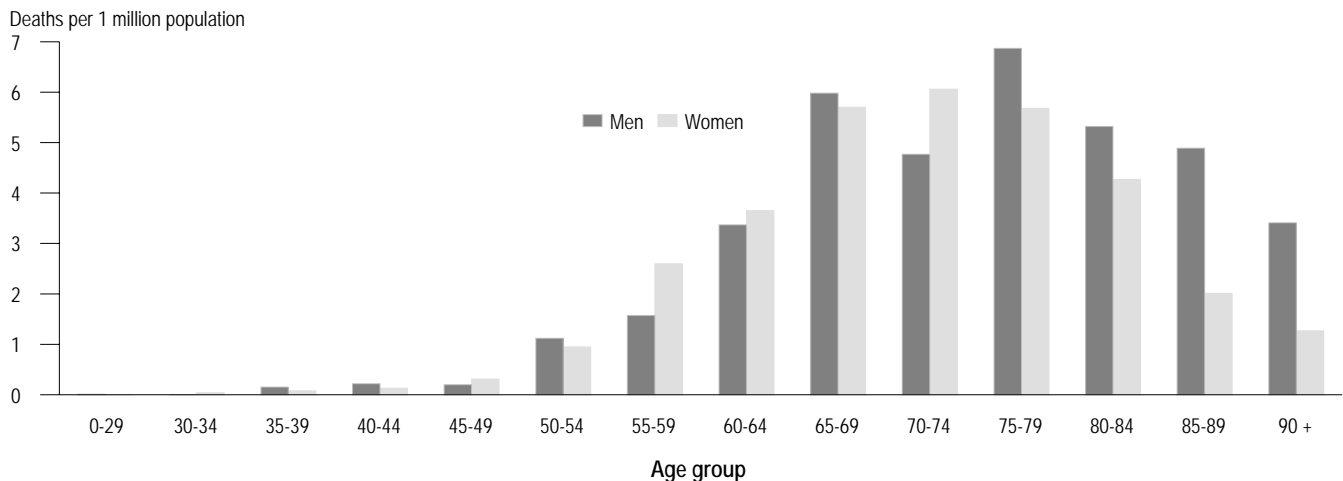
CJD mortality rates rise sharply with age, especially after 50. Rates were highest at ages 75 to 79 for men, and at ages 70 to 74 for women.

Between 1979 and 2001, CJD mortality among the provinces ranged from a low of 0.5 deaths per million population in Newfoundland to a high of 1.3 in Nova Scotia.

## Diagnosis and autopsies

Confirming a diagnosis of CJD can be difficult because the clinical symptoms are similar to those of other neurological disorders such as Alzheimer's disease. Brain scans and tonsil biopsy are used to establish a probable diagnosis, but a confirmed diagnosis can only be made with a microscopic examination of the brain tissue after the patient has died. While it might be anticipated that autopsies

### Mortality rates for Creutzfeldt-Jakob disease, by age and sex, Canada, 1979 to 2001



Data source: Vital Statistics Death Database

would follow the majority of suspected CJD deaths in Canada, between 1979 and 2001, autopsies were performed in 45% of cases. Nonetheless, this is much higher than the 16% of autopsies that were performed for all deaths.

## Risk difficult to estimate

The fear and media attention that surround Creutzfeldt-Jakob disease and its bovine counterpart, “mad cow disease,” intensified throughout 2003 with the discovery of two BSE-infected cows in North America. Much of the concern focuses on the economic impact. In 2001, the World Health Organization Director General, Dr. Gro Harlem Brundtland, referred to the issue of BSE and its link to CJD as a “global emergency” likely to cost “several tens of billions of dollars.”<sup>8</sup>

## Data source

Information about Creutzfeldt-Jakob Disease deaths was taken from the **Vital Statistics Death Database**, which is based on death certificates submitted by the provinces and territories and maintained by Statistics Canada. The *International Classification of Diseases* (ICD) categorizes Creutzfeldt-Jakob Disease under code 046.1 for deaths from 1979 to 1999 (ICD-9), and code A81.0 for deaths in 2000 and 2001 (ICD-10). No sub-classification distinguishes between the classical and variant forms. However, Health Canada’s surveillance system monitors all referrals with suspected CJD. These cases are followed until an autopsy report, or other evidence, confirms the diagnosis and distinguishes variant from the classical forms.

From the point of view of simply counting occurrences, concern about the public health implications may be unwarranted: between 1979 and 2001, Canadians were more likely to die from extreme cold or from falling off a ladder or scaffold than from CJD. However, the menace of CJD lies in the unknown size of the infected population. The long period between infection and the onset of symptoms makes it difficult to

estimate the numbers of people who may be at risk of developing vCJD, particularly as no tests can identify it prior to the onset of symptoms.

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