Prion Disease: Information for Health Care and Public Health Professionals

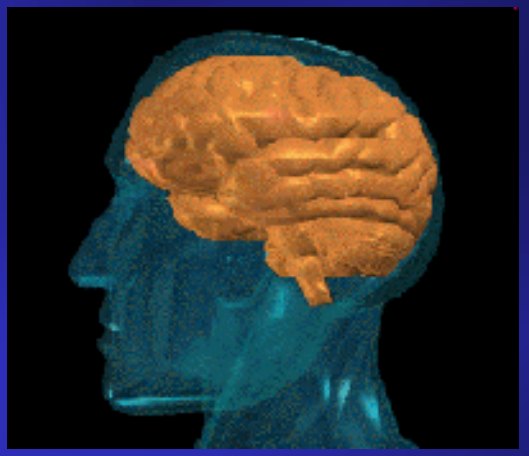
Michigan Department of Community Health

Updated 09/2012

Presentation Outline

- Prion Disease Etiology
- Animal Prion Diseases
- Human Prion Diseases
- Prion Disease Diagnostic Testing
- Prion Disease Infection Control
- Prion Disease Surveillance

Prion Diseases- What are they?



- Progressive, transmissible, and fatal diseases of the central nervous system
- Also known as Transmissible Spongiform Encephalopathies (TSE) due to the spongy appearance of brain tissue
- Long incubation periods, but usually rapidly progressive once clinical symptoms appear

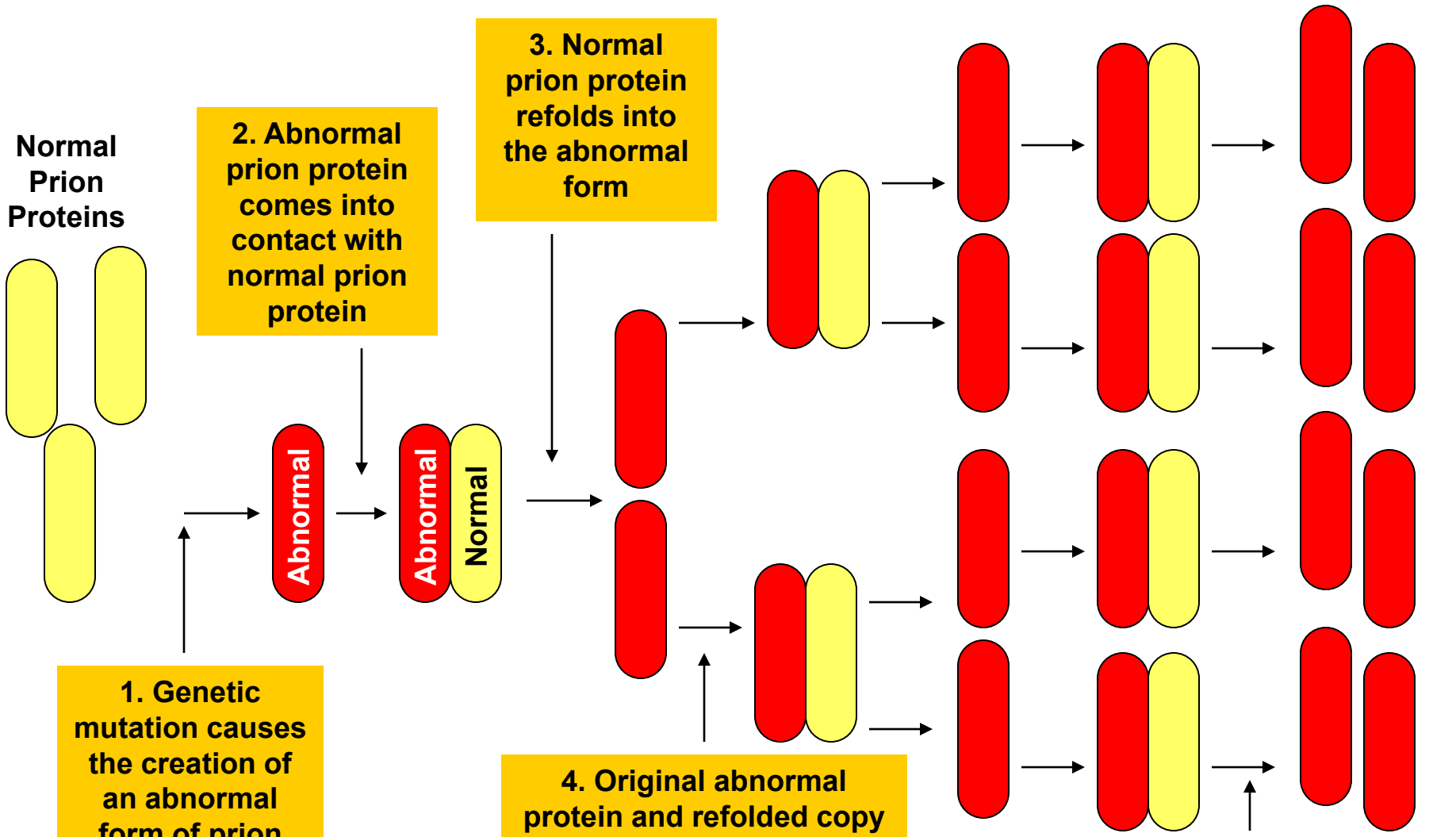
Prion Disease Etiology

- All humans have prion proteins as a normal part of their central nervous system
- Specific gene mutations may cause the production of an abnormal, misfolded prion protein
- The abnormal form of prion protein is more stable than the normal conformation
- When an abnormal prion protein encounters a normal prion protein, the normal protein refolds into the abnormal conformation

Etiology Cont'd

- A cascade of normal prion proteins being converted into the abnormal form occurs
- The abnormal proteins cannot be broken down by the body and accumulate in the brain
- Holes in brain matter occur where the abnormal proteins accumulate
  - The term "spongiform" is derived from the spongy appearance of the brain
- Clinical disease results from the damage caused by the accumulation of these abnormal prion proteins

Progression of Prion Disease



Adapted from Robbins Basic Pathology 7th Ed., 2003

Animal Prion Disease

Animal Prion Disease

- Bovine Spongiform Encephalopathy (BSE)- “mad cow disease”, found in cattle
- Chronic Wasting Disease (CWD)- found in cervids (deer and elk)
- Scrapie- found in sheep
- Other less common prion diseases have been found in felines, mink, etc.

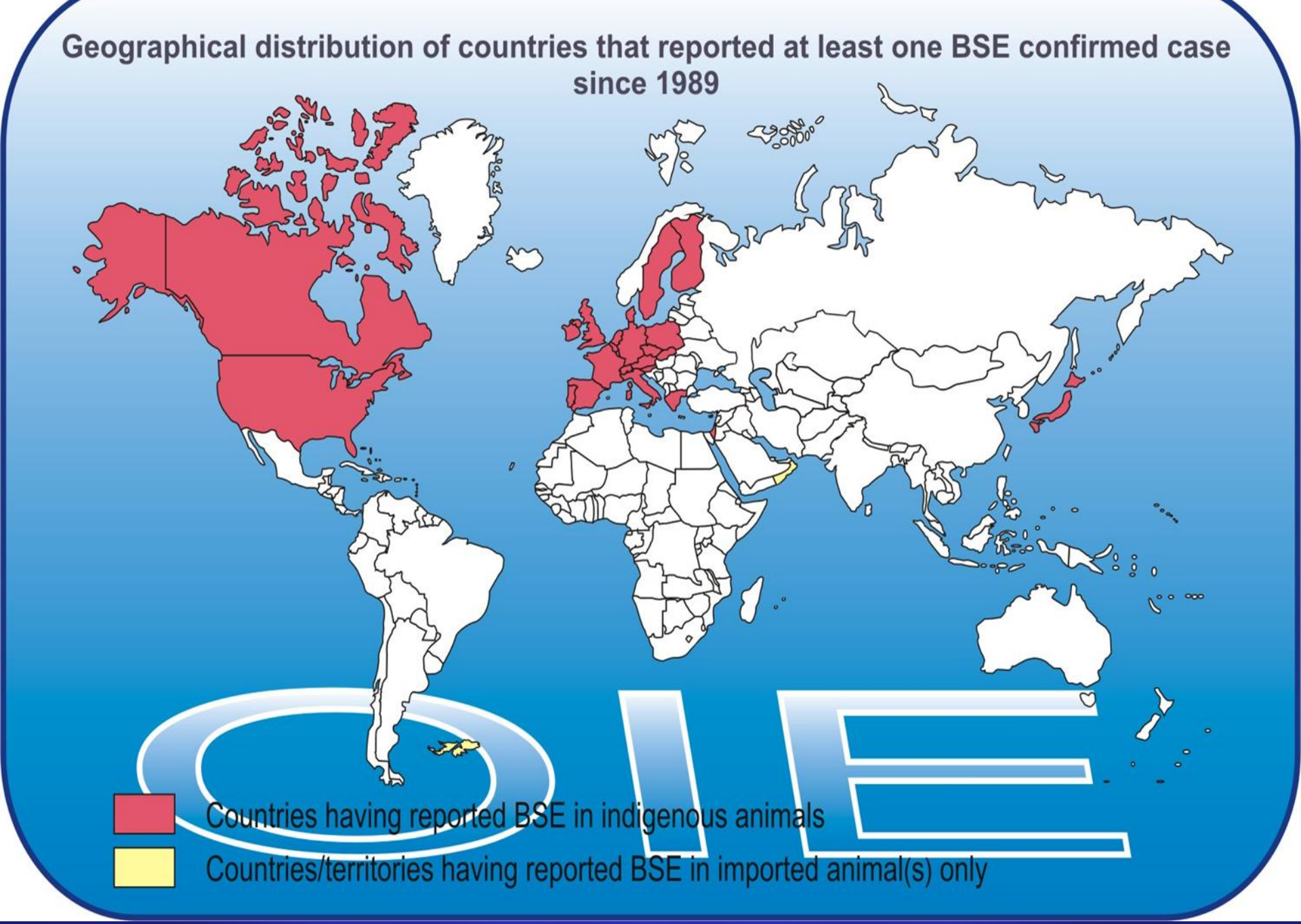
Bovine Spongiform Encephalopathy

- Commonly known as “Mad Cow Disease”
- BSE first recognized in 1986 in cattle in the United Kingdom (UK)
- Clinical signs in cattle:
  - Temperament changes (nervousness, aggression)
  - Heightened sensory perception
  - Abnormal posture
  - Loss of coordination
  - Weight loss
  - Excessive licking or itching


APHIS

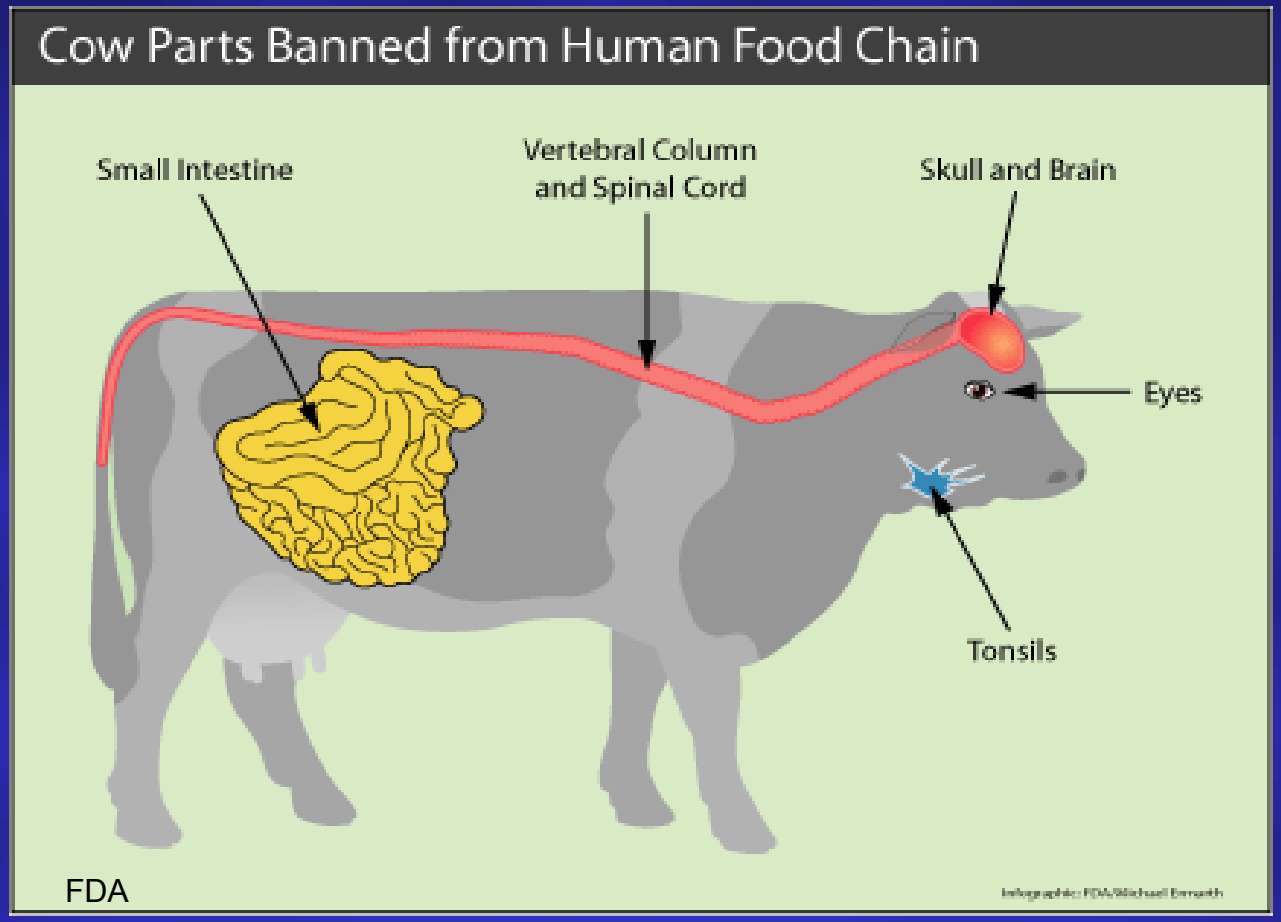
- Linked to the human illness variant Creutzfeldt-Jakob Disease (vCJD)

Geographical distribution of countries that reported at least one BSE confirmed case since 1989



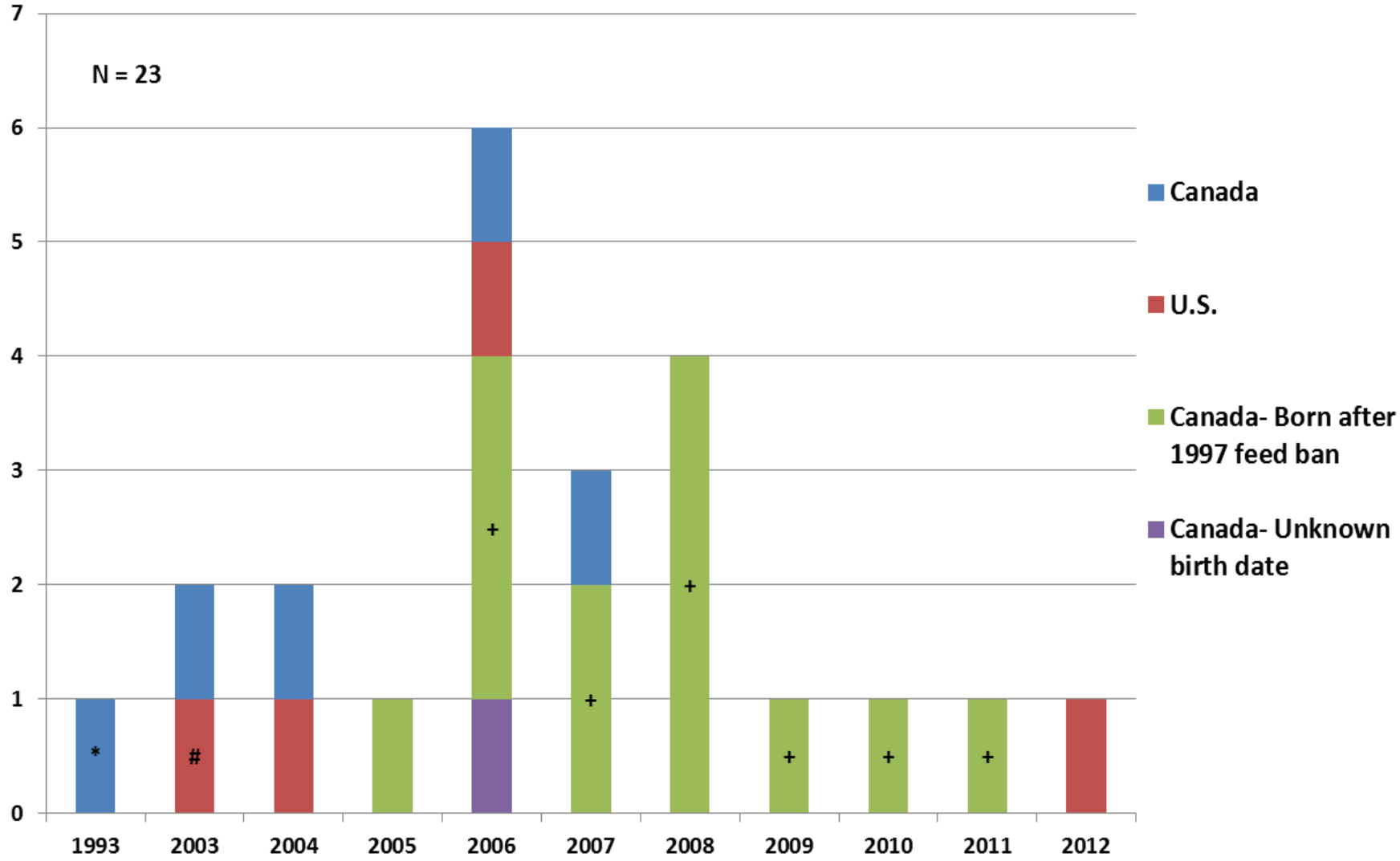
Source: http://www.oie.int/en/animal-health-in-the-world/bse-specific-data/map/

Bovine Spongiform Encephalopathy



Tissues known as "specified risk materials" are banned from the human food chain and for use in animal feed from cattle over 30 months of age. This feed ban went into effect in 1997.

BSE Cases in North America, by Year and Country of Death 1993, 2003-2012



* Imported from UK into Cananda

\# Imported from Cananda into U.S.

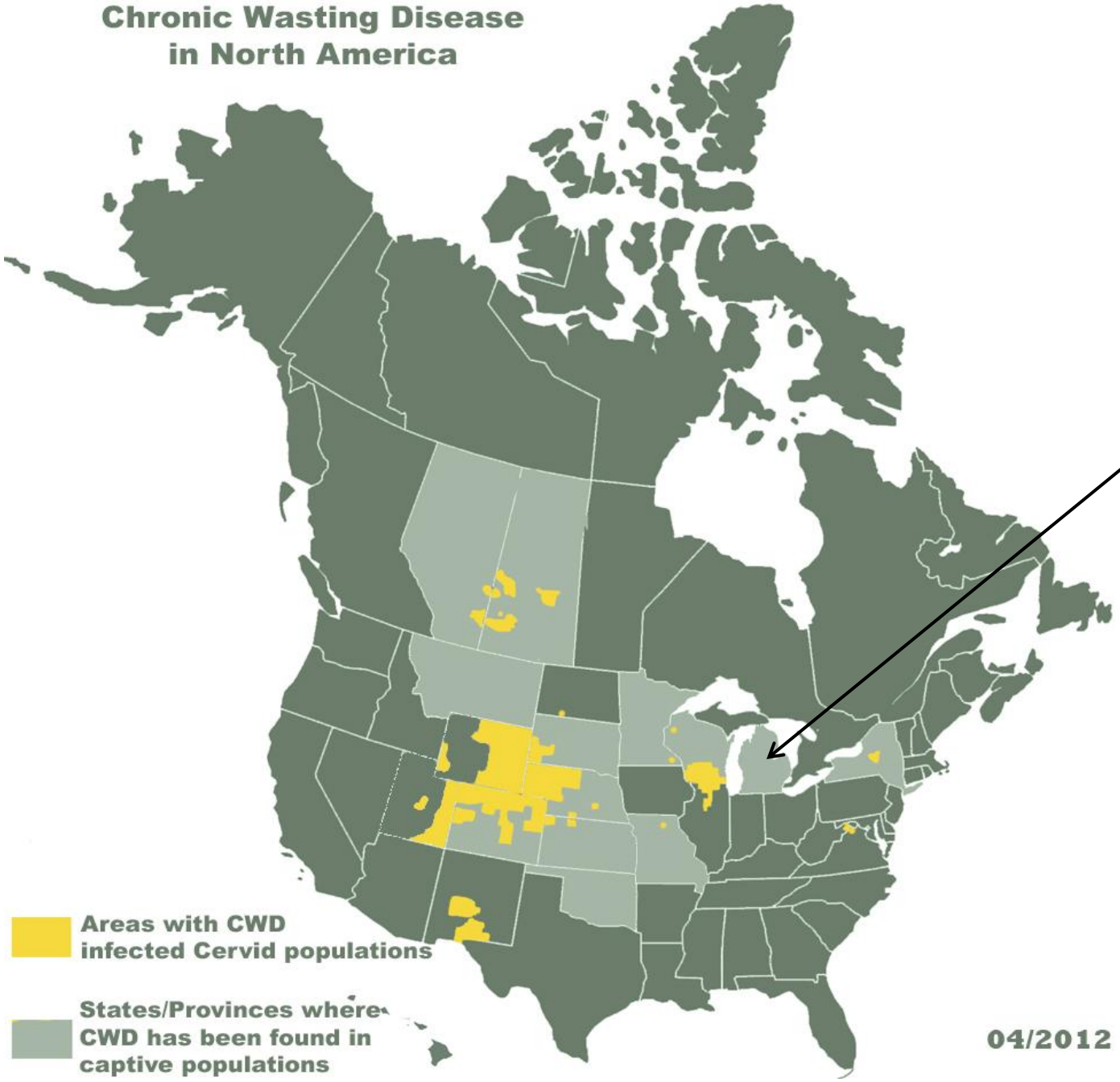
\+ Born after March 1st, 1999

Source: Adapted from CDC- http://www.cdc.gov/ncidod/dvrd/bse/

Chronic Wasting Disease

- Prion disease affecting cervids (deer and elk)
- Transmission- animal to animal, and possibly from the environment
- Clinical signs:
  - Weight loss
  - Excessive salivation and grinding of the teeth
  - Behavioral changes

- No evidence to date that CWD can cause human disease



To date, one case of CWD in a captive (non-wild) deer has been found in Michigan

Source: Chronic Wasting Disease Alliance- www.cwd-info.org

Human Prion Disease

Forms of Human Prion Disease

- Human prion diseases occur in 3 distinct forms
  - Sporadic (Classical)
    - Disease results from a spontaneous genetic mutation that creates the abnormal form of prion protein
    - No known inherited or environmental cause

  - Familial (Inherited)
    - Individuals carry a gene that makes it more likely the abnormal form prion protein will be created by the body
  - Acquired
    - Disease results from the introduction of the abnormal prion protein from an outside source
    - Includes iatrogenic cases caused by medical procedures and variant CJD cases linked to the consumption of BSE contaminated products

Types of Human Prion Disease

- **Sporadic (Classical)**
  - Sporadic Creutzfeldt-Jakob Disease (CJD)
    - NOT associated with BSE ("Mad Cow")
  - Sporadic Fatal Familial Insomnia
- **Familial (Inherited)**
  - Familial CJD
  - Fatal Familial Insomnia
  - Gerstmann-Straüssler-Scheinker Syndrome
- **Acquired**
  - Variant CJD (vCJD)
    - Human form of BSE ("Mad Cow")
  - Iatrogenic CJD
  - Kuru

Sporadic Human Prion Disease

- **Sporadic Creutzfeldt-Jakob Disease (CJD)**
  - The most common type of human prion disease accounting for ~85-90% of CJD cases
  - Caused by a spontaneous mutation
    - Not associated with BSE ("Mad Cow"), not inherited
  - Symptom onset generally around age 60 and include progressive dementia, behavior changes and incoordination
  - Average duration of disease from symptom onset to death is 4-6 months
  - Amyloid plaques are NOT present in the brain

Familial Human Prion Disease

- **Familial CJD**
  - Hereditary form of disease with dominant inheritance
  - Accounts for 5-10% of CJD cases
  - People with this inherited mutation are predisposed to producing the abnormal form of the prion protein
  - Carrying the gene mutation does not mean the person will definitely develop disease
  - Symptom onset generally in early 50's
  - Course of disease is longer than the sporadic type

Familial Human Prion Disease

- **Fatal Familial Insomnia (FFI)**
  - Rare, inherited prion disease that primarily affects the thalamus region of the brain
    - As the disease progresses, individuals lose the ability to sleep
  - Onset generally between the ages of 40-60
  - Death usually occurs 7-36 months after onset of symptoms
  - Mild spongiosis, amyloid plaques absent
  - A sporadic version of FFI that is not genetically linked also exists

Familial Human Prion Disease

- **Gerstmann-Straüssler-Scheinker Syndrome (GSS)**
  - Rare, inherited prion disease
  - Presents with difficulty speaking and unsteadiness, dementia occurs later in the course of disease
  - Onset is generally during ages 40-60
  - Disease course may last from 1-10 years
  - Amyloid plaques present in the brain

Acquired Human Prion Disease

- **Iatrogenic CJD**
  - Disease caused by the introduction of abnormal prion protein during a medical procedure
  - Documented transmission from:
    - Contaminated neurosurgical instruments
    - Tissue transplants: dura mater, cornea
    - Pooled human growth hormone
    - Variant CJD from blood transfusions
      - Sporadic CJD is not transmitted by blood transfusions

Acquired Human Prion Disease

- **Iatrogenic CJD Cont'd**
  - The incubation time for the disease is dependent on the route of inoculation
    - Incubation time is shorter when the abnormal prion protein is introduced directly into the brain or central nervous system
  - The risk for iatrogenic CJD has been nearly eliminated by the use of more stringent decontamination methods for neurosurgical instruments and the discontinuation of the use of cadaver derived dura mater and human growth hormone
    - No cases from contaminated instruments have been identified since 1976

Acquired Human Prion Disease

- **Kuru**
  - Prion disease discovered in the 1950's
  - Found in Papua New Guinea among members of the Fore tribe
  - Thought to be associated with the handling and possible consumption of brain during funeral rituals
  - The disease is basically non-existent at this time

Acquired Human Prion Disease

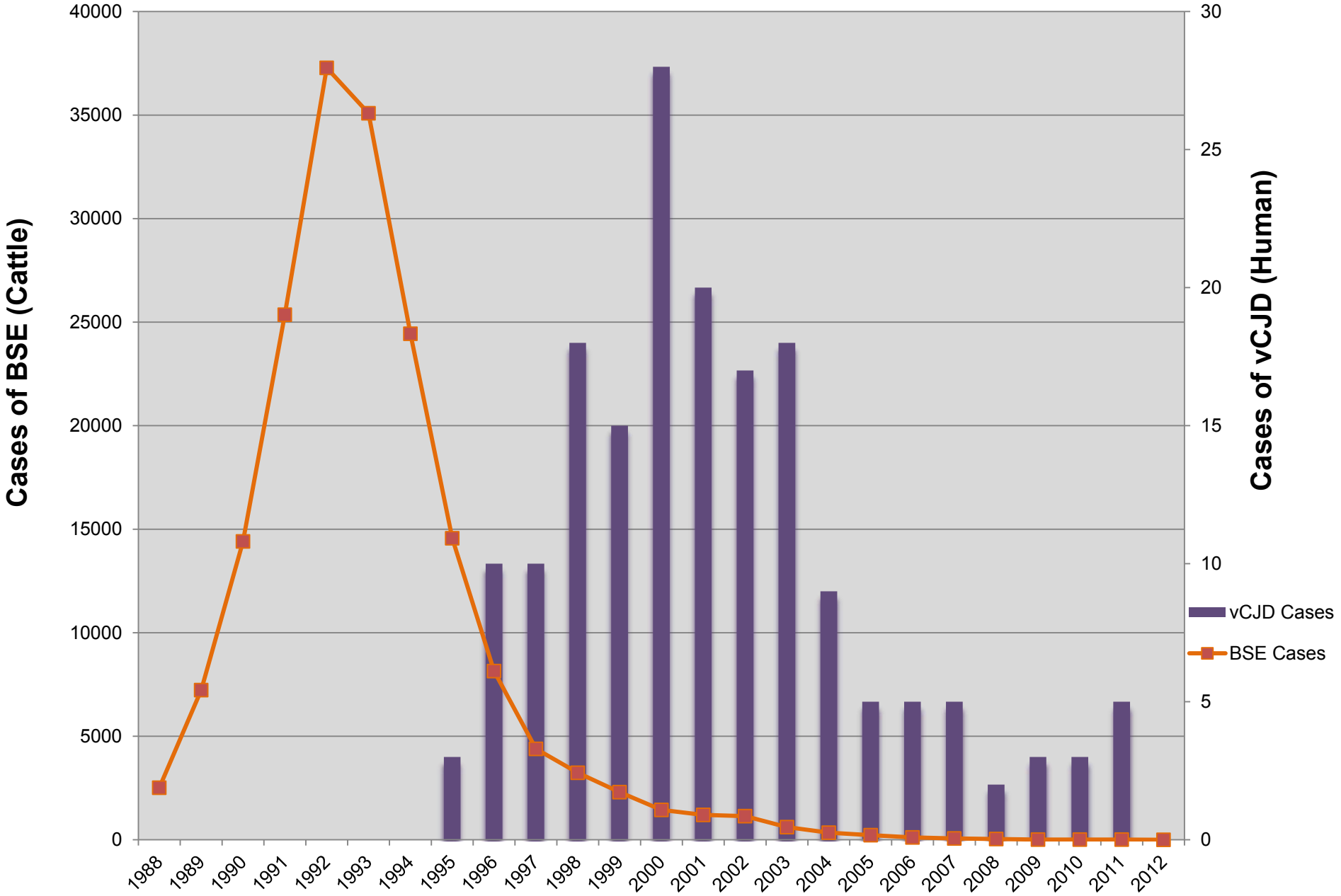
- **Variant CJD (vCJD)**
  - Human form of BSE ("Mad Cow")
  - First case reported in the United Kingdom in 1995
  - Average age of onset is the late 20's
  - The duration of vCJD is longer than sporadic CJD and can last from 8 to 38 months
  - Disease presentation for variant CJD is quite different from sporadic CJD
  - Numerous amyloid plaques found in both the cerebellum and cerebrum

Worldwide Cases of Variant CJD

- Since vCJD was first identified in 1995, a total of 222 cases from 12 countries have been identified
  - 173 from the United Kingdom (U.K.)
  - 25 from France (1 possibly exposed in U.K.)
  - 5 from Spain
  - 4 from Ireland (2 possibly exposed in U.K.)
  - 3 from the United States (2 exposed in U.K., 1 in Saudi Arabia)
  - 3 from the Netherlands
  - 2 from Portugal
  - 2 from Italy
  - 2 from Canada (1 possibly exposed in U.K.)
  - 1 each from Japan, Saudi Arabia, and Taiwan (Taiwan case possibly exposed in U.K.)

Source: NCJDRSU- http://www.cjd.ed.ac.uk/vcjdworld.htm

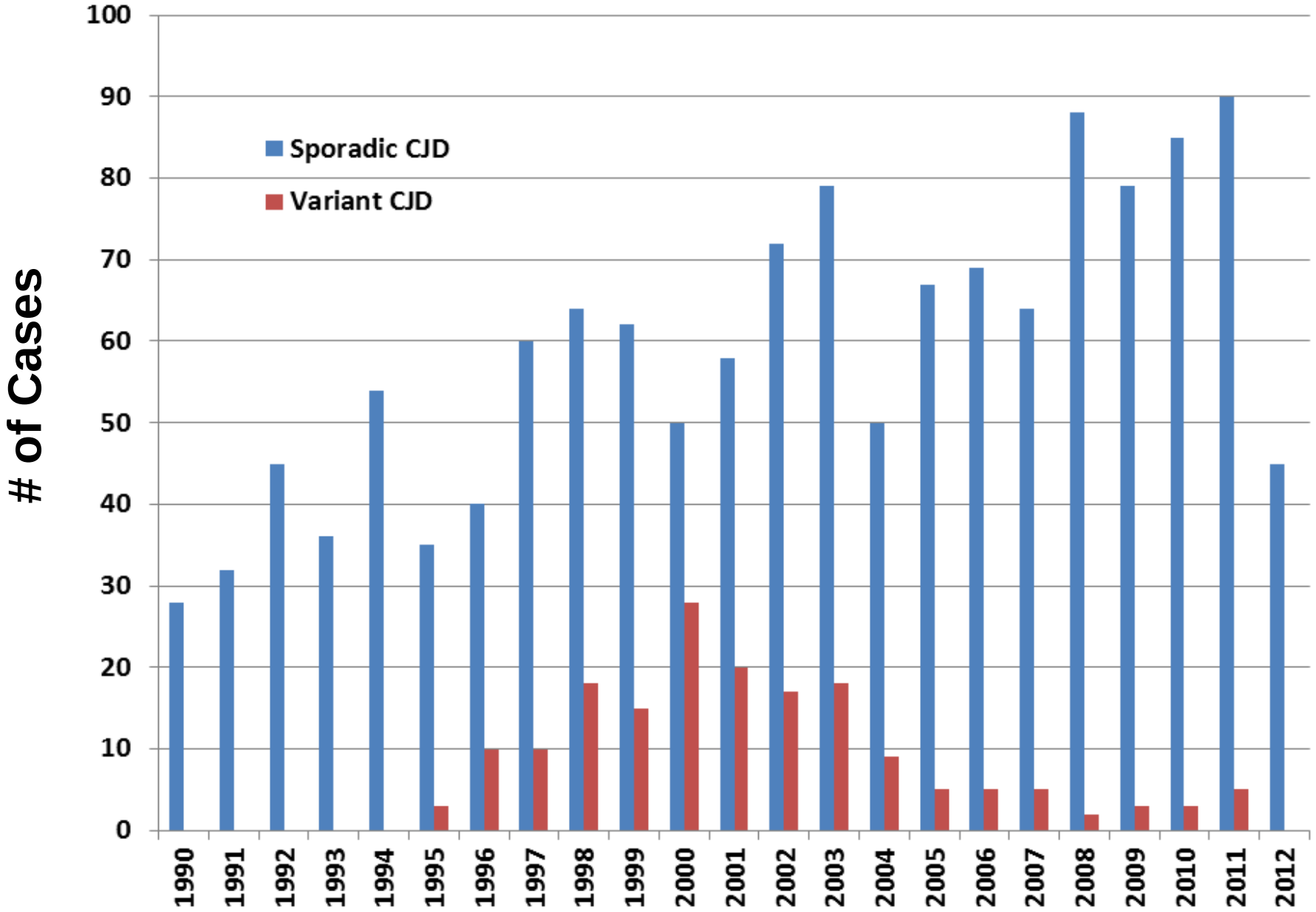
Cases of BSE and Variant CJD in the U.K. 1988-2012*



Source: NCJDSU - http://www.cjd.ed.ac.uk

* As of 09/2012

Sporadic and Variant CJD Cases in the U.K.*



Source: NCJDSU - http://www.cjd.ed.ac.uk/figures.htm

* As of 09/2012

Exposure to BSE in the U.K.

- It is estimated that over 50 million infectious doses of BSE contaminated products were consumed by individuals in the U.K. during the primary years of the BSE outbreak
- The relatively low number of human cases of variant CJD suggests that the species barrier (inherent genetic and biologic differences between species) provided protection from the disease for most people
  - Surveillance continues to try and determine if certain individuals have longer incubation periods or can be "carriers" and never develop disease

Variant CJD in the U.S.

- **No domestically acquired cases of variant CJD have been discovered to date in the United States**
  - **3 total cases of variant CJD have been diagnosed in the U.S.**
    - **Cases are attributed to the country where the diagnosis is made, not where the infection was acquired**
    - **All 3 individuals were foreign born and likely exposed prior to moving to the U.S. (2 U.K., 1 Saudi Arabia)**
      - **Based on the long incubation period of prion disease**

Sporadic vs. Variant CJD

- The two diseases are distinguished based on clinical presentation and laboratory testing results
  - Clinical presentations
    - Sporadic CJD generally begins with neurologic symptoms while variant CJD often presents with psychological symptoms
  - Affect different age groups
    - Sporadic CJD is generally found in those 60+, variant in late 20's
  - Durations of illness
    - Sporadic CJD disease course from onset of symptoms usually lasts 4-5 months, variant 13-14 months
  - Present differently on CTs, EEGs, and MRIs
  - Are easily distinguished through the examination and testing of brain autopsy tissue

Clinical and Pathologic Characteristics of Sporadic CJD and Variant CJD

Characteristics	Sporadic CJD	Variant CJD
Median Age at Death	68 Years	28 Years
Median duration of illness	4-5 months	13-14 months
Clinical signs and symptoms	Dementia; early neurologic signs	Prominent psychiatric/behavioral symptoms; painful dysesthesiasis; delayed neurologic signs
Periodic sharp waves on electroencephalogram	Often present	Often absent
"Pulvinar sign" on MRI*	Not reported	Present in >75% of cases
Presence of "florid plaques" on neuropathology	Rare or absent	Present in large numbers
Immunohistochemical analysis of brain tissue	Variable accumulation	Marked accumulation of protease-resistance prion protein
Presence of agent in lymphoid tissue	Not readily detected	Readily detected
Increased glycoform ratio on immunoblot analysis of protease-resistance prion protein	Not reported	Marked accumulation of protease-resistance prion protein

*An abnormal signal in the posterior thalami on T2- and diffusion-weighted images and fluid-attenuated inversion recovery sequences on brain magnetic resonance imaging (MRI); in the appropriate clinical context, this signal is highly specific for vCJD

Table adapted by the CDC from Belay E., Schonberger L. Variant Creutzfeldt-Jakob Disease and Bovine Spongiform Encephalopathy. Clin Lab Med 2002;22:849-62.

Diagnostic Testing

Diagnostic Testing Resource

- National Prion Disease Pathology Surveillance Center (NPDPSC)
  - Located at Case Western Reserve University in Cleveland, OH
  - Funded by in part the CDC and NIH for prion disease testing and surveillance
  - Website: www.cjdsurveillance.com

NPDPSC Diagnostic Testing

Laboratory Tests Performed:

- **Fixed brain tissue** (from biopsy or autopsy):
  - Histopathology
  - Immunohistochemistry
- **Frozen brain tissue** (from biopsy or autopsy):
  - Western blot- determines presence and type of prion protein
  - DNA extraction and prion gene sequencing- assess the presence and type of mutations
- **CSF**:
  - Assessment of 14-3-3 and Tau protein levels

Source: NPDPSC

Testing Considerations

- **Tissue from a brain autopsy is currently the only method available to definitively determine the presence of prion disease or rule it out**
- **A brain biopsy is not a preferred method for diagnosis**
  - **Sample of brain tissue taken may not be the location where the prion accumulations are present**
  - **A positive biopsy is definitive for prion disease, but a negative biopsy can not rule-out disease**
  - **Should only be used when a treatable condition is suspected**

Source: NPDPSC

Testing Considerations

- Only testing on frozen brain tissue can confirm or exclude the diagnosis of prion disease and identify the type of prion disease present
- Immunohistochemical examination on fixed tissue provides a definitive diagnosis of prion disease only when positive, but may not be able to identify the type of prion disease
- CSF and blood examinations provide information that may be helpful to physicians in making a clinical diagnosis, but are not considered confirmatory tests

Source: NPDPSC

NPDPSC Services

- Autopsy services and testing can be arranged for through the National Prion Disease Pathology Surveillance Center (NPDPSC)
- The NPDPSC can:
  - Provide assistance in finding a facility to perform an autopsy in the event that the referring facility is not equipped to handle one
  - Provide a pathologist at the referring facility location to perform the autopsy
  - Cover the cost of transportation if the autopsy is to be performed outside the referring facility
  - Perform laboratory testing on CSF, blood, and brain tissue

Source: NPDPSC

The Importance of an Autopsy

- **A brain autopsy is strongly recommended in all cases where a clinician suspects prion disease**
  - Only method currently available to confirm prion disease
  - Only way to determine the type of prion disease (e.g. sporadic CJD, familial CJD, variant CJD)
  - May provide the opportunity to detect new and emerging types of human prion diseases
  - Helps provide information and increase the understanding of prion diseases
    - Goal to develop successful treatments in the future

Source: NPDPSC

NPDPSC Services

- If you would like the Center to make autopsy arrangements please have the family call the Autopsy Coordinators at 216-368-0587. The Center will collect some basic information and have the family send written consent to perform the autopsy.
- Information on laboratory testing protocols and referral forms that must accompany samples can be found on the NPDPSC website: http://www.cjdsurveillance.com

Source: NPDPSC

National Prion Disease Pathology Surveillance Center
Cases Examined[1]

(August 14, 2012)

Year	Total Referrals[2]	Prion Disease	Sporadic	Familial	Iatrogenic	vCJD
1996 & earlier	50	32	28	4	0	0
1997	114	68	59	9	0	0
1998	88	52	44	7	1	0
1999	123	74	65	8	1	0
2000	145	103	89	14	0	0
2001	210	120	110	10	0	0
2002	248	149	125	22	2	0
2003	266	168	137	31	0	0
2004	326	187	164	22	0	1[3]
2005	344	194	157	36	1	0
2006	382	196	166	28	0	2[4]
2007	377	213	185	28	0	0
2008	396	232	206	26	0	0
2009	423	256	212	43	1	0
2010	414	257	216	41	0	0
2011	411	258	214	43	0	0
2012	233	133	105	19	0	0
TOTAL	**4550**[5]	**2692**[6]	**2282**	**391**	**6**	**3**

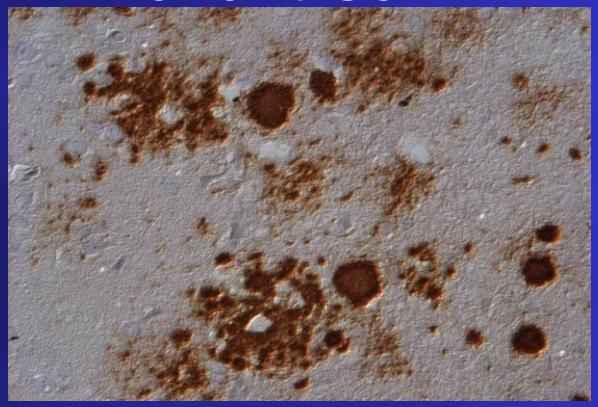
[1] Listed based on the year of death or, if not available, on year of referral; [2] Cases with suspected prion disease for which brain tissue and/or blood (in familial cases) were submitted; [3] Disease acquired in the United Kingdom; [4] Disease was acquired in the United Kingdom in one case and in Saudi Arabia in the other case; [5] Includes **8** cases in which the diagnosis is pending, and **18** inconclusive cases; [6] Includes **10** (**9** from **2012**) cases with type determination pending in which the diagnosis of vCJD has been excluded. The Sporadic cases include **16** cases of sporadic Fatal Insomnia (sFI) and **42** cases of Variably Protease-Sensitive Prionopathy (VPSPr) and 2224 cases of sporadic Creutzfeldt-Jakob disease (sCJD).

NPDPSC, 2012: http://www.cjdsurveillance.com/pdf/case-table.pdf

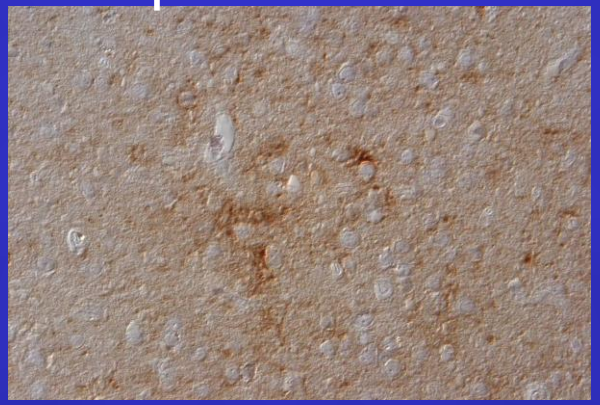
Diagnostic Testing Recap

- Positive 14-3-3 and Tau protein tests on CSF are suggestive of prion disease, but not confirmatory
- Disease can only be confirmed by a brain autopsy or in some cases a brain biopsy

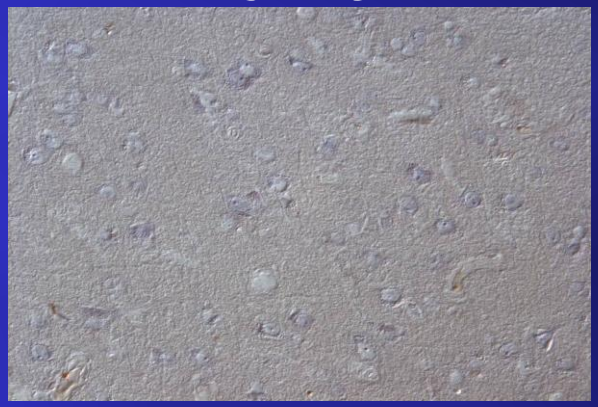
Variant CJD



Sporadic CJD



Normal

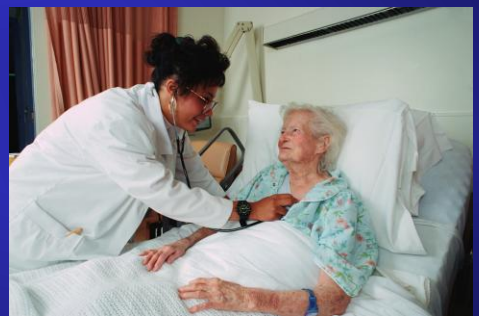


Source: J. Ironside, National CJD Surveillance Center, Edinburgh, and P. Gambetti, National Prion Disease Pathology Surveillance Center, Cleveland, OH September 2003.

Infection Control

Patient Care

- **Normal clinical contact does not pose a threat to health care workers**
  - **Prion diseases are not contagious in the typical sense**
    - **Not transmitted by respiratory or airborne droplets, through bodily fluids, or sexual contact**
- **Standard precautions should be used during routine clinical contact**
  - **Gloves should be used for blood draws**
- **Private rooms are not required**



WHO Infection Control Guidelines for TSEs, 1999

Occupational Exposure

No cases of human prion disease are known to have occurred through occupational exposure

Occupational Exposure

- **Contamination of unbroken skin with internal body fluids or tissues:**
  - Wash with detergent and warm water (avoid scrubbing), rinse, and dry
- **Needle sticks or lacerations:**
  - Encourage bleeding; wash (avoid scrubbing) with warm soapy water, rinse, dry and cover with a waterproof dressing. Further treatment should be appropriate to the type of injury
  - Report the injury to your occupational health service
- **Splashes into the eye or mouth:**
  - Irrigate with either saline (eye) or tap water (mouth); report accordingly
- **Health and safety guidelines mandate reporting of injuries, and records should be kept for no less than 20 years for prion disease exposures**

WHO Infection Control Guidelines for TSEs, 1999

Tissue Infectivity Levels

Infectivity Category	Tissues, Secretions, and Excretions
High Infectivity	Brain, Spinal Cord, Eye, Blood*
Low Infectivity	CSF, Kidney, Liver, Lung, Lymph node/ spleen, Placenta

*Blood transfusions have been linked to the transmission of Variant CJD, but not Sporadic CJD

Infectivity

- The route of exposure must be considered along with the infectivity of a tissue when determining the risk of infection
  - The use of contaminated neurosurgical equipment that is not properly disinfected carries a much higher risk of infection compared to getting CSF on intact skin
  - Transmission seems to require the direct inoculation of the brain or nervous system with infective tissue

Disinfection

- Prions are not inactivated by conventional chemical or physical decontamination methods such as:
  - Disinfectants
  - Tissue fixatives
  - Ionizing and UV radiation
  - Infectivity may persist after standard autoclaving (121 C for 15 minutes)

Infection Control- Instruments

Instruments used on neurosurgery patients with no clear diagnosis should be processed using the guidelines for prion decontamination

Disposable Instruments

- Single use, disposable instruments/ clothing should be used whenever possible during surgery involving a confirmed or suspected case of prion disease
- Disposable non-permeable material should be used to cover all work surfaces
- Single use instruments and materials should be disposed of by incineration

Non-disposable Instruments

- Instruments should be placed in a leak-proof container labeled with prion contamination warning and cleaned according to WHO guidelines as soon as possible after use
- Instruments should be kept moist until cleaned
- Avoid mixing instruments used on tissues with different levels of infectivity
- Re-use durable items only after prion decontamination methods
- Instruments must be decontaminated by prion methods before processing through automated machines/ washer, and the washers (or other equipment) should be run through an empty cycle before any further routine use

Decontamination

- Heat-resistant instruments
- Methods listed from more (1) to less (3) stringent

1. Immerse in a pan containing 1N sodium hydroxide (NaOH); heat in a gravity displacement autoclave at 121°C for 30 min; clean; rinse in water; and subject to routine sterilization.

2. Immerse in 1N NaOH or sodium hypochlorite (20,000 ppm available chlorine) for 1 hour; transfer instruments to water; heat in a gravity displacement autoclave at 121°C for 1 hour; clean; and subject to routine sterilization.

3. Immerse in 1N NaOH or sodium hypochlorite (20,000 ppm available chlorine) for 1 hour; remove and rinse in water, and then transfer to open pan and heat in a gravity displacement (121°C) or porous load (134°C) autoclave for 1 hour; clean; and subject to routine sterilization.

World Health Organization

Infection Control Guidelines

http://whqlibdoc.who.int/hq/2000/WHO_CDS_CSR_APH_2000.3.pdf

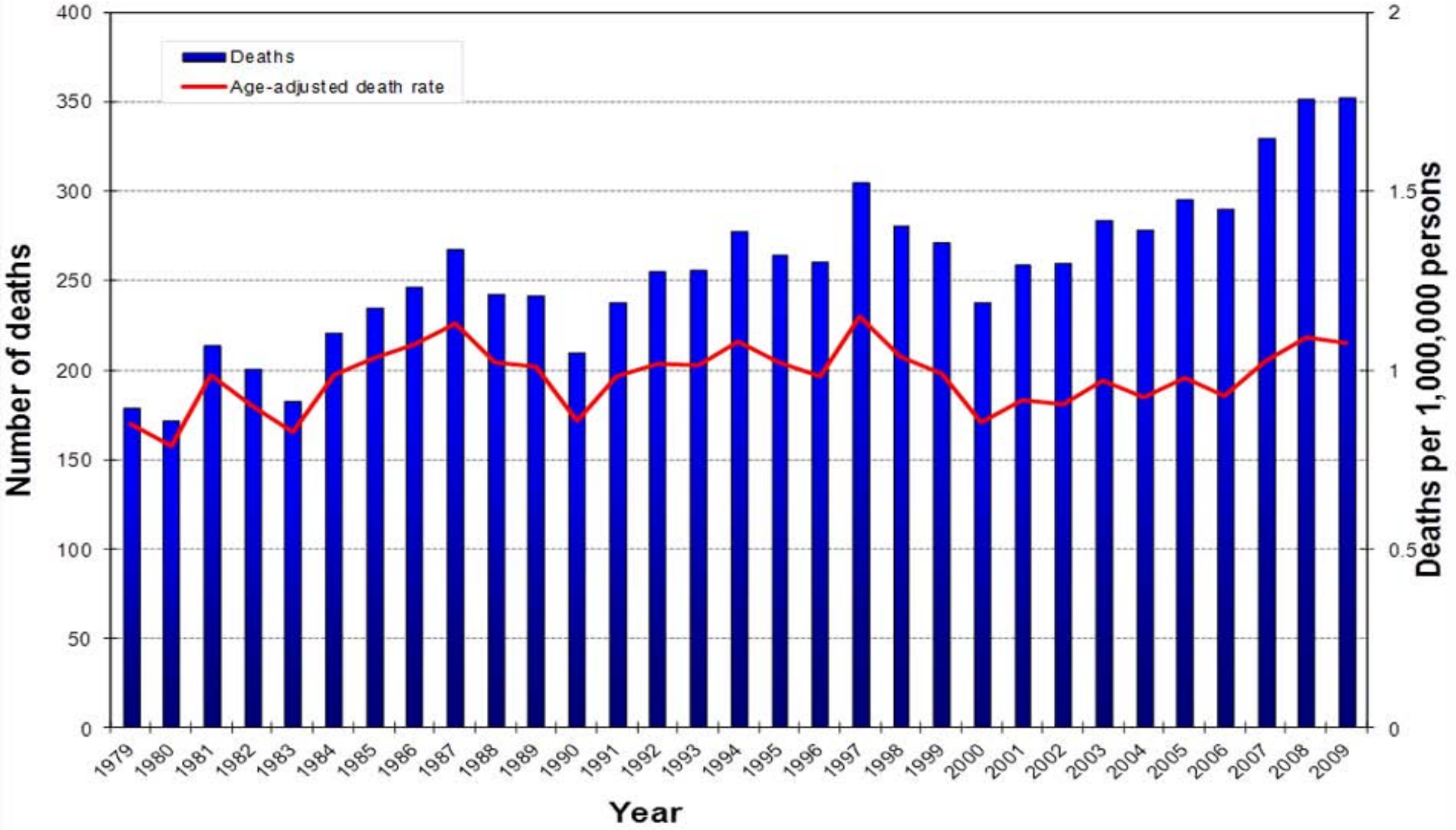
Prion Disease Surveillance

Sporadic CJD Incidence United States

- **Current CDC estimates:**
  - Over 55 years old: 1 case per million people per year
  - 55 years old or younger: 1 case per 10 million people per year
  - 65-79 age group: 5 cases per million people per year
    - Highest rate among any age group

Based on the population of the United States we would expect to see ~ 300 cases of CJD per year

Creutzfeldt-Jakob disease deaths and age-adjusted death rate, United States, 1979-2009*



* Deaths obtained from the multiple cause-of-death data for 1979-1998 are based on ICD-9 codes, and those beginning in 1999 are based on ICD-10 codes with available computerized literal death certificate data. Death information was also obtained from other surveillance mechanisms; includes familial prion disease. Rates are adjusted to the US standard 2000 projected population.

Source: CDC- http://www.cdc.gov/ncidod/dvrd/cjd/index.htm

Prion Disease Case Definitions

- **Sporadic CJD**
  - **Definite:**
    - Diagnosed by standard neuropathological techniques, and/or
    - IHC confirmed protease-resistant prion protein, and/or
    - Western blot confirmed protease-resistant prion protein, and/or
    - Presence of scrapie associated fibrils
  - **Probable:**
    - Progressive dementia; and at least two out of the following four clinical features:
      - Myoclonus
      - Visual or cerebellar signs
      - Pyramidal/extrapyramidal signs
      - Akinetic mutism

    And
    - Routine investigations should not suggest an alternative diagnosis
    - Atypical EEG during an illness of any duration, and/or
    - A positive 14-3-3 or Tau CSF assay and a clinical duration to death of <2 years

Prion Disease Case Definitions

- **Sporadic CJD cont'd**
  - **Suspect:**
    - Progressive dementia; and at least two of the following four clinical features:
      - Myoclonus
      - Visual or cerebellar signs
      - Pyramidal/extrapyramidal signs
      - Akinetic mutism

    And

    - Atypical EEG or no EEG
    - A duration of <2 years

Prion Disease Case Definitions

- **Familial CJD**
  - Definite or probable CJD **and** definite or probable CJD in a first degree relative, **and/or**
  - Neuropsychiatric disorder and disease-specific prion protein gene mutation
  - Note: Gerstmann-Sträussler-Scheinker (GSS) syndrome and fatal familial insomnia (FFI) should be reported in a similar manner as familial CJD
- **Iatrogenic CJD**
  - Progressive cerebellar syndrome in a recipient of human cadaver-derived pituitary hormone; **or**
  - Sporadic CJD with a recognized exposure risk, e.g. antecedent neurosurgery with dura mater implantation

Prion Disease Case Definitions

- **Variant CJD (in the United States)**
  - **Definite:**
    - Diagnosed by neuropathologic examination of brain tissue
    - The following confirmatory features should be present:
      - Numerous widespread kuru-type amyloid plaques surrounded by vacuoles in both the cerebellum and cerebrum (florid plaques); and
      - Spongiform change and extensive prion protein deposition shown by IHC throughout the cerebellum and cerebrum

Prion Disease Case Definitions

- **Variant CJD (in the United States) Cont'd**
  - **Suspect:**
    - Routine investigations of the patient do not suggest an alternative, non-CJD diagnosis
    - Current age or age at death <55 years old
    - Duration of illness of over 6 months
    - Dementia; and the development ≥4 months after illness onset of at least two of the five neurologic signs: (If persistent painful sensory symptoms exist, the ≥4 months delay in the development of the neurological signs is not required)
      - poor coordination, myoclonus, chorea, hyperreflexia , visual signs
    - Psychiatric symptoms at illness onset and/or persistent painful sensory symptoms (frank pain and/or dysesthesia)
    - A normal or an abnormal EEG, but not the diagnostic EEG changes often seen in sporadic CJD
    - No history of receipt of cadaveric human pituitary growth hormone or a dura mater graft
    - No history of CJD in a first degree relative or prion protein gene mutation in the patient

Michigan Prion Disease Surveillance

- **Reporting**
  - On December 1st, 2004 all spongiform encephalopathies (prion diseases) were added to the reportable disease list in Michigan
  - Physicians, hospitals and laboratories are required to report cases of prion disease
    - Reporting is NOT a violation of HIPAA
  - Please contact the local health department where the patient resides if you have a suspected or confirmed case of prion disease

Michigan Prion Disease Surveillance

- **Surveillance Methods**
  - Required reporting of all suspect or confirmed cases of prion disease from physicians, health care facilities, and laboratories
  - Follow-up on test results received from National Prion Disease Pathology Surveillance Center
  - Detailed medical record review in cases ≤ 55 years old
  - Detailed surveillance questionnaire to be completed by the physician and/or family
  - Death certificate review

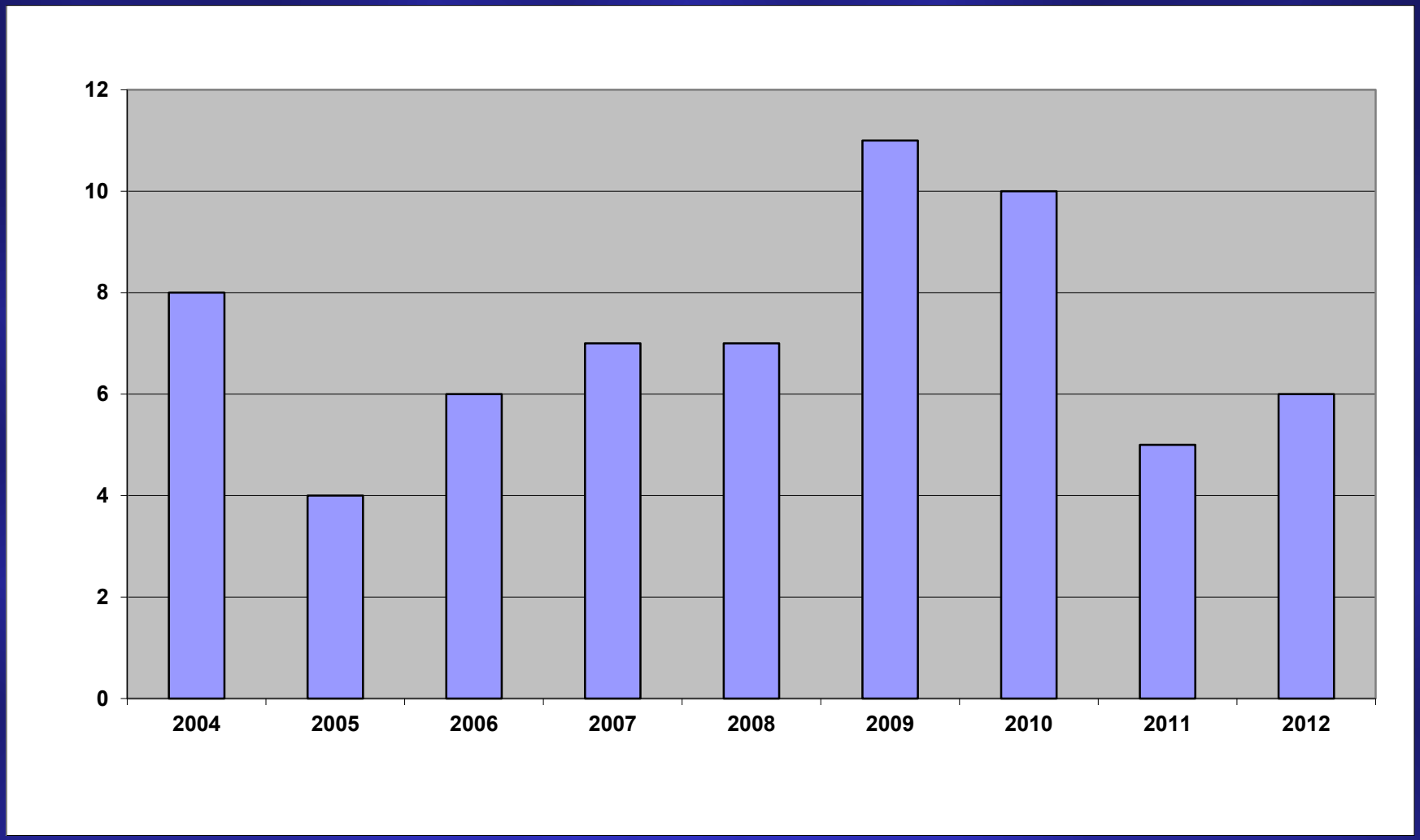
Michigan Prion Disease Statistics

- Based on the population of Michigan it is expected that ~10 cases of prion disease would occur per year

- Timely reporting by physicians is essential to determining the types of prion diseases present in the U.S. and the true prevalence of prion disease in the population

Confirmed* Prion Disease Cases in Michigan 2005-2012

(as of 9/01/12)



*This table includes only cases where a brain autopsy or biopsy was done and testing confirmed the presence of prion disease

Questions?

Please contact the Michigan Department of Community Health Communicable Disease Division at: 517-335-8165

In order to better understand the audience for this presentation, please email the following information to Shannon Johnson, MDCH Prion Disease Epidemiologist (johnsons61@michigan.gov):

- Your name
- Your job title
- Your affiliation, e.g. hospital, private practice, health department, etc.
- Comments or suggestions about the presentation

This information will only be used to help improve the presentation. Thank you for your assistance.