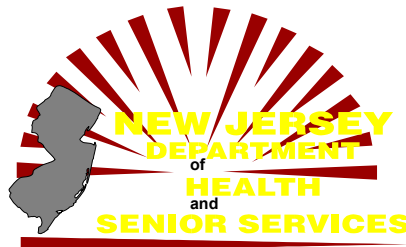


# Evaluation of a Suspected Cluster of CJD in New Jersey

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# Transmissible Spongiform Encephalopathies (TSEs)

- Bovine spongiform encephalopathy (BSE)
- Scrapie (sheep)
- Chronic wasting disease (deer, elk)
- Creutzfeldt-Jacob disease (CJD) (humans)

# CJD Classification

- Variant (vCJD)
- Classic
  - Sporadic
  - Familial
  - Iatrogenic

**TABLE. Clinical and pathologic characteristics distinguishing variant Creutzfeldt-Jakob disease (vCJD) from classic CJD — United Kingdom (UK) and United States, 1979–2001**

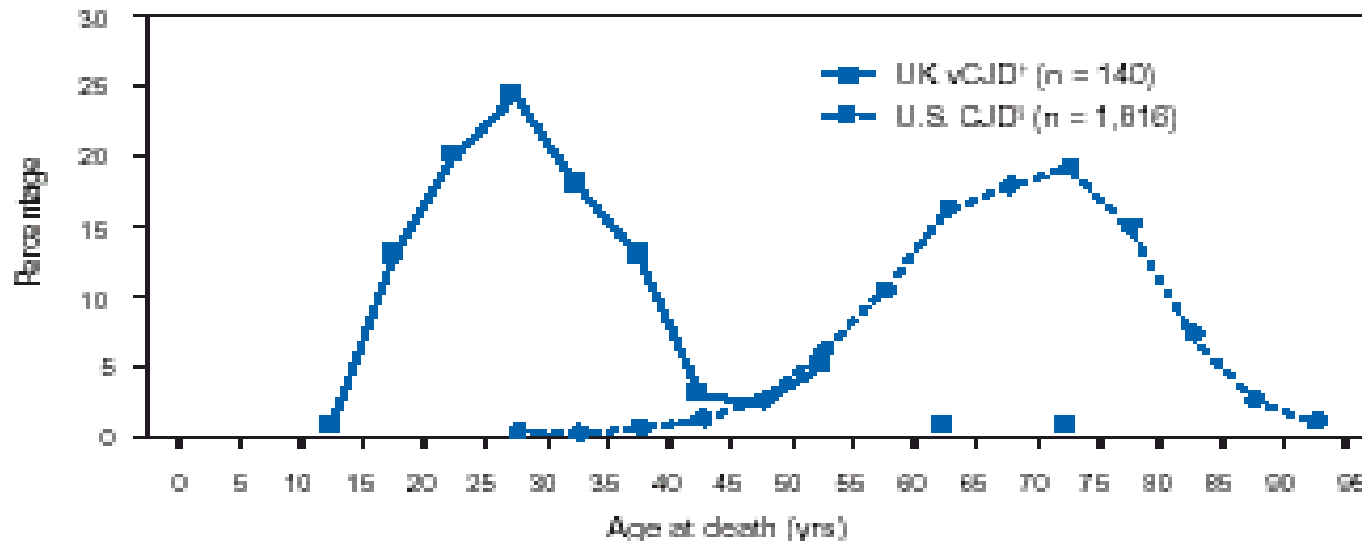
Characteristic	UK vCJD	U.S. classic CJD
Median age at death (yrs)	28 (range: 14–74)	68 (range: 23–97)*
Median illness duration (mos)	13–14	4–5
Clinical presentation	Prominent psychiatric/behavioral symptoms; painful sensory symptoms; delayed neurologic signs	Dementia; early neurologic signs
Periodic sharp waves on EEG	Absent	Often present
“Pulvinar sign” on MRI†	Present in >75% of cases	Not reported
Presence of “florid plaques” on neuropathology	Present in great numbers	Rare or absent
Immunohistochemical analysis of brain tissue	Marked accumulation of Pr <sup>Pres</sup> §	Variable accumulation
Presence of agent in lymphoid tissue	Readily detected	Not readily detected
Increased glycoform ratio on immunoblot analysis of Pr <sup>Pres</sup>	Present	Not present
Genotype at codon 129 of prion protein	Methionine/Methionine	Polymorphic

\* Surveillance data 1979–2001.

† High signal in the posterior thalamus.

§ Protease-resistant prion protein.

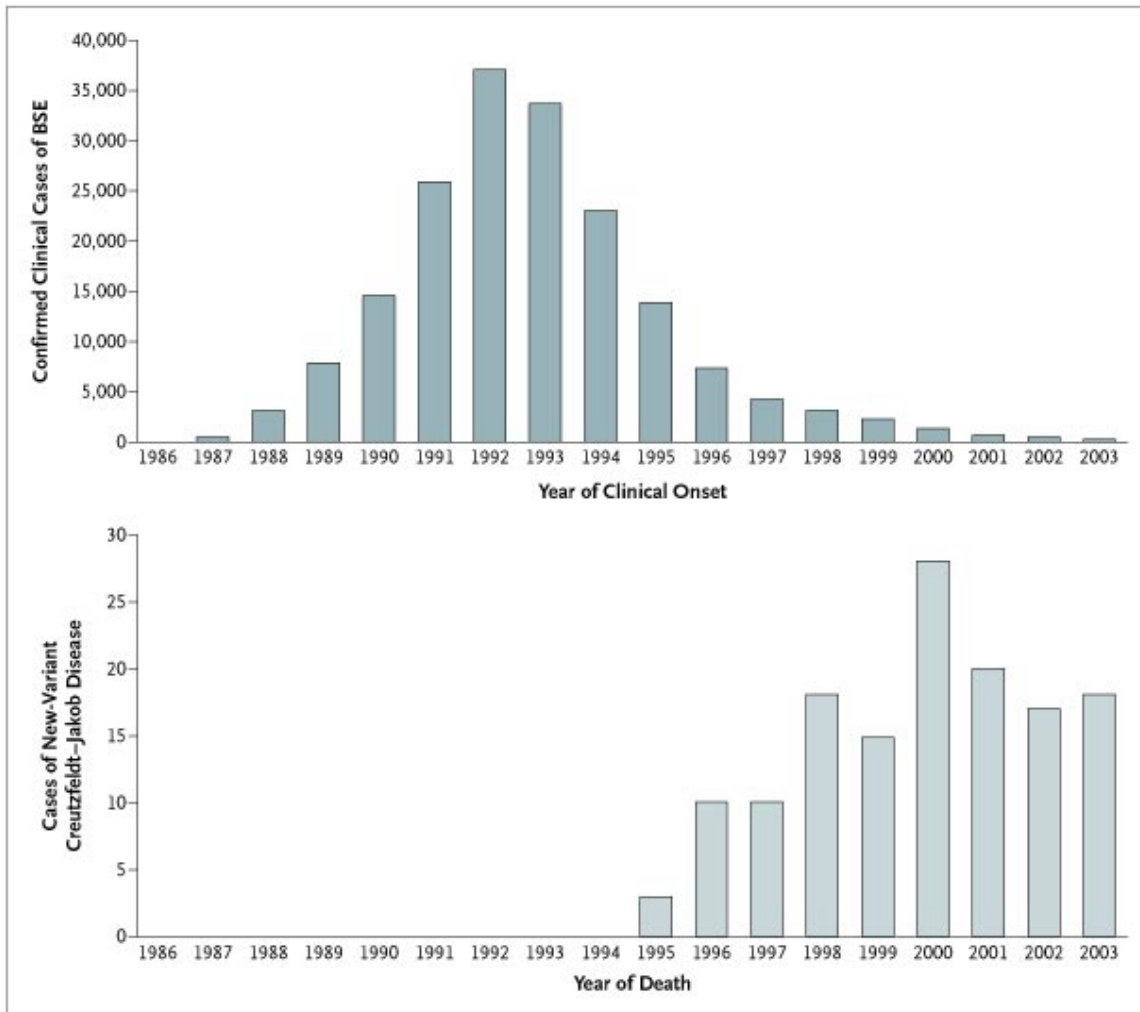
**FIGURE. Percentage distribution of deaths caused by variant Creutzfeldt-Jakob disease (vCJD) in the United Kingdom (UK) and deaths caused by CJD in the United States, by age at death, 1995–2003\***



\* Excludes blood transfusion–associated vCJD and pituitary hormone- or dural graft–associated CJD.

† Noniatrogenic UK vCJD deaths, including UK-related nonresident cases, 1995–2003.

§ Noniatrogenic U.S. deaths, 1995–2001.



From: Donnelly, NEJM 2004; 350: 539-542.

**Figure.** Creutzfeldt-Jakob Disease Age-Adjusted and Age-Specific Death Rates, and Deaths by Age United States, 1979-1998



From: Gibbons et al: JAMA 2000; 284:2322-2323.

# Number of CJD Cases in New Jersey

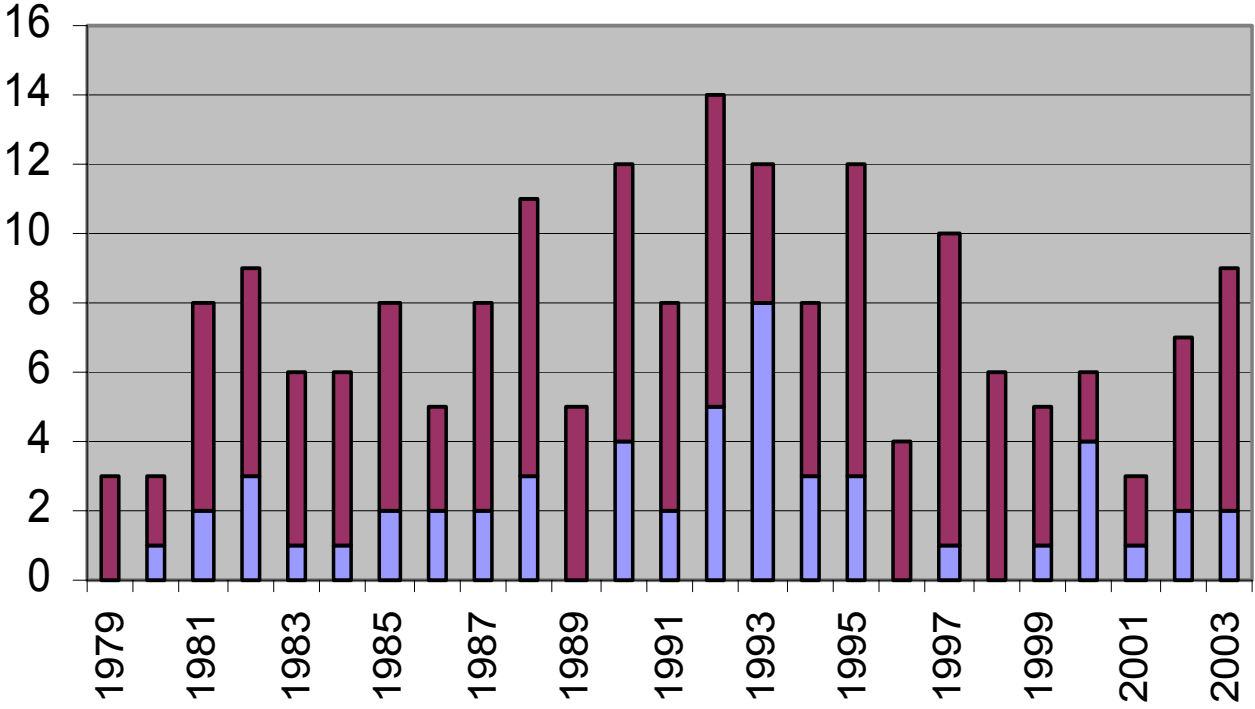
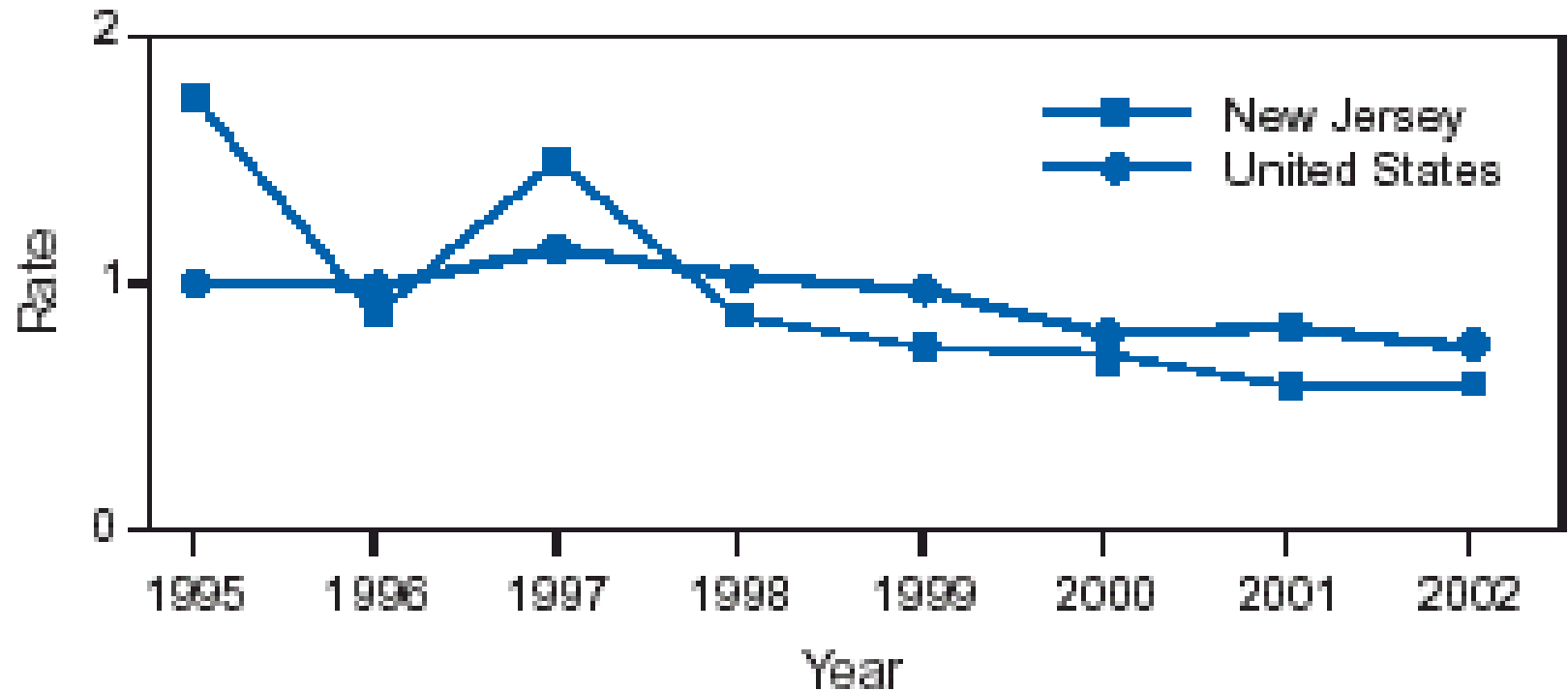




FIGURE 1. Creutzfeldt-Jakob disease death rates\*, by year — New Jersey and United States, 1995–2002†



\* Per 1 million persons.

† From CDC's multiple cause-of-death file; 2002 data are preliminary.

# Reported Suspected Sporadic CJD

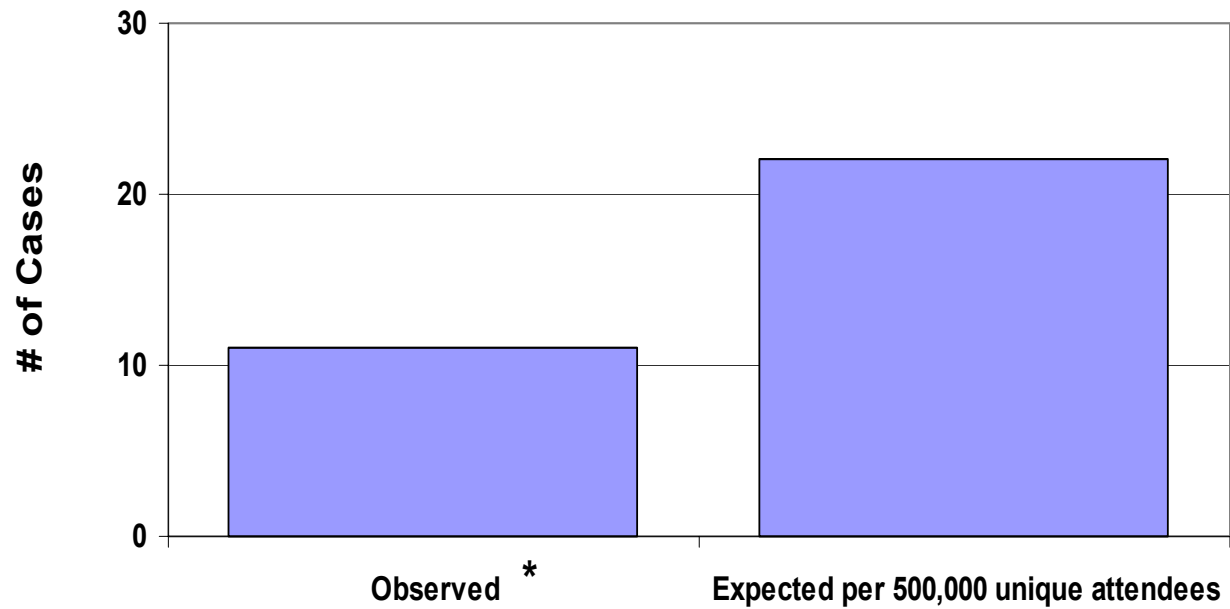
## Garden State Racetrack, 1995 – 2004

<b>Status</b>	<b>Number</b>	<b>NJ</b>	<b>Non-NJ</b>
<b>Definite</b>	5	4	1
Probable	6	2	4
Possible	2	0	2
Excluded	4	1	3

Non-NJ: PA, VA, MD, CT, DE

# CJD Cases at Garden State Racetrack

Observed vs. Expected (>55 years of age)  
1993 - 2004

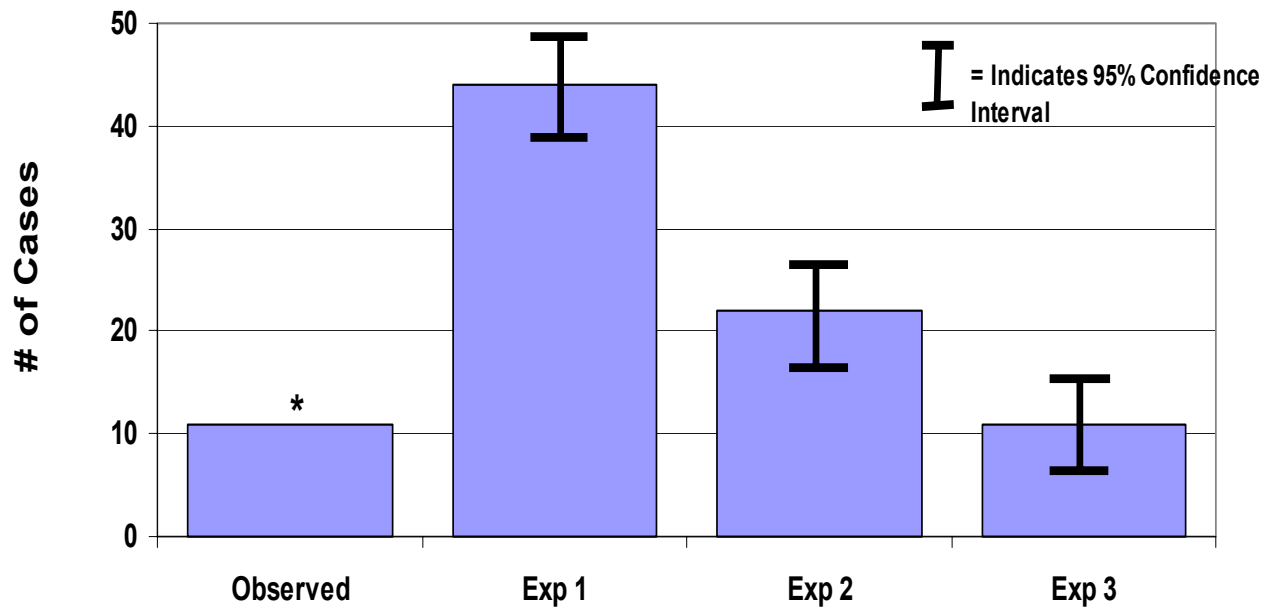


\*1993 - April. 2004

# CJD Cases at Garden State Racetrack

## Observed vs. Expected (>55 years of age)

### 1993 - 2004



\*1993 - April, 2004

Exp 1 = Expected # deaths per 1,000,000 unique attendees  
Exp 2 = Expected # deaths per 500,000 unique attendees  
Exp 3 = Expected # deaths per 250,000 unique attendees