EPI-NEWS **NATIONAL SURVEILLANCE OF COMMUNICABLE DISEASES**

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CREUTZFELDT - JAKOB DISEASE 1998

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Creutzfeldt-Jakob disease (CJD) belongs to the group of human prion diseases giving rise to fatal spongiform encephalopathies. CJD occurs spontaneously with an incidence of about 1:1,000,000 population per annum and is divided into sporadic, familial and iatrogenic types. In addition, variant CJD (vCJD) was described in 1996 and is thought to be the human form of mad cow disease or bovine spongiform encephalopathy (BSE; EPI-NEWS 10/97). In the UK a total of 52 patients have so far been reported to have died of definite or probable vCJD. In addition, two cases of vCJD have been reported in France and one in Ireland.

Notified cases

The first report from the Danish surveillance of CJD was published in EPI-NEWS 13-14/99. This report dealt with all notified cases whose illness started in 1996 or 1997 and comprised 13 cases. As the final classification of the disease can normally only be established after death, future reports will be based on the year of death. Thus Table 1 shows the number of patients notified with suspected CJD who died in 1998. Figures for 1997 are also included for comparison.

Table 1. No. of notified cases of Creutzfeldt-Jakob disease

Year of death	1997	1998
Definite cases	8	3
Probable cases	2	2
Possible cases	1	0
vCJD cases	0	0
Total	11	5

Table 1 shows that a total of 11 patients with suspected CJD died in 1997; three men and eight women. In 1998 five patients died, three men and two women. The median age for patients dying in 1997 and 1998 was 65 and 70 years, respectively, with a range of 40-88 years. All the cases were classified as sporadic CJD. So far no case of vCJD has been detected in Denmark. Eight cases from 1997 and three from 1998 could be classified as definite on the basis of neuropathological investigations. In the five instances in which no post mortem was carried out, the cases were classified from the clinical criteria given overleaf.

The median duration of illness was six and seven months, respectively, for cases from 1997 and 1998, with a range of 2-22 months. Provisional figures for 1999 suggest that the incidence of sporadic CJD is remaining similar to that reported for 1998.

Diagnostic criteria and classification

CJD and related spongiform encephalopathies have been officially notifiable since 1997, and must be notified on suspicion alone. In addition, Denmark is participating in a European project which is collecting data to monitor the incidences of the different human prion diseases, with special emphasis on vCJD. In connection with this project a uniform set of criteria has been established for the diagnosis and classification of cases of CJD. These criteria, for the respective classifications of sporadic CJD and vCJD, are given overleaf.
The stated criteria for vCJD have recently been adopted by the British surveillance system for this disease.
Sporadic CJD occurs mainly in the middle-aged or elderly. The typical picture is one of rapidly progressive dementia with myoclonus. Most patients have characteristic EEG changes and the disease progresses rapidly to akinetic mutism and death within a few months. Cases of vCJD typically occur in younger persons and takes the form of progressive neuropsychiatric symptoms followed by cerebellar symptoms and ataxia. A progressive dementia then supervenes. Myoclonus occurs in a majority of these patients, and the duration of illness is typically longer than for sporadic CJD.
For either sporadic CJD or vCJD, the diagnosis can only be made with certainty after post mortem neuropathological examination.
As part of the European project, a Danish expert group has been established to follow and advise on national surveillance of the disease.

Diagnostic investigations

EEG and MRI are regarded as useful investigations for establishing the probable diagnosis of CJD. As stated overleaf, determination of protein 14-3-3 in the cerebrospinal fluid will provide additional evidence of possible sporadic CJD. 14-3-3 is a normal brain protein that leaks into the cerebrospinal fluid during the course of the disease. It is also possible to perform genetic investigations for the familial variant of the disease.
Because the diagnosis, as previously stated, can only be finally established by neuropathological examination no matter what type of CJD is concerned, it is especially important to carry out a post mortem examination including the brain on patients dying from suspected CJD. Special precautions must be rigorously observed when handling tissues from suspected CJD patients. These have been published in the National Board of Health Circular no. 55 of 7 May 1997.

Risk of BSE-infected foods in Denmark

In the UK, 400,000-500,000 infected cows probably entered the food chain before the risk of infection was acknowledged. Since then about 178,000 infected cows have been destroyed. It must be assumed that a considerable number of people were exposed to infection from this meat.
At present 52 cases of vCJD are known to have occurred in the UK. There are no signs of a rising tendency.
On the basis of a single case of mad cow disease in Denmark, the risk of Danish consumers' being exposed to infected meat is thought to be quite small. However, this risk can not be precisely quantified.
(J. Duus Johansen, E. Smith, Dept. of Epidemiology)

AMBULANCE HYGIENE RECOMMENDATIONS

The Ambulance Hygiene Committee, which includes representatives from the Copenhagen Fire Brigade and the Falck Rescue Service, has initiated the updating of hygiene recommendations for patient transport in ambulances. CAS and the Department of Epidemiology have revised the recommendations, which deal with modes of infection and the precautionary measures to be adopted for certain diseases. The recommendations can be downloaded from the Statens Serum Institut/CAS home page.
(A. H. Christiansen, Dept. of Epidemiology)

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Diagnostic criteria and classification of Creutzfeldt-Jakob disease

As applied in the European surveillance programme

Sporadic CJD

I. Progressive dementia

II. A Myoclonus
B Visual or cerebellar problems
C Pyramidal/extrapyramidal features
D Akinetic mutism

III. Typical EEG changes

IV. Demonstration of 14-3-3 protein in the cerebrospinal fluid

Definite CJD:	Neuropathologically confirmed
Probable CJD:	Either: Criterion I, plus two manifestations listed under II, plus III
	Or: Possible CJD plus criterion IV
Possible CJD:	Criterion I, plus two manifestations listed under II, plus duration of illness < 2 years

Variant CJD

I. A Progressive neuropsychiatric disorder
B Duration of illness > 6 months
C Routine investigations do not suggest an alternative
D No history of potential iatrogenic exposure

II. A Early psychiatric symptoms
B Persistent, painful sensory symptoms
C Ataxia
D Myoclonus or chorea or dystonia
E Dementia

III. A Abnormal EEG, but not typical for sporadic CJD
B High signal in thalamus on MRI

Definite vCJD:	Neuropathologically confirmed
Probable vCJD:	Criterion I, plus four manifestations listed under II, plus III
Possible vCJD:	Criterion I, plus four manifestations listed under II

(Department of Epidemiology