ISSN 1188-4169

Canada Communicable Disease Report

FAX

Vol. 25-1

Date of publication: 1 January 1999

Contained in this FAX issue: *(No. of pages: 6)*

Official page numbers:

For reference purposes, citing should refer to the page numbers of the printed copy and not to those of the FAX copy (F-#).

International Notes

HUMAN TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHIES

This article presents the conclusions and recommendations of the WHO Consultation on the global surveillance, diagnosis, and therapy of human transmissible spongiform encephalopathies which was held in February 1998 in Geneva.

Creutzfeldt-Jakob disease (CJD) is a rare and fatal human neurodegenerative condition characterized in most cases by a rapidly progressive dementia, myoclonus, and a periodic electroencephalogram (EEG). It is classified as a transmissible spongiform encephalopathy (TSE) because it causes characteristic spongy degeneration of the brain and can be transmitted to laboratory animals. TSEs also affect a range of animal species including sheep, goats, cows, deer, mink, and cats in non-experimental conditions. CJD is by far the most common human TSE. It occurs sporadically in about 85% of cases, and is inherited in 10% to 15% of cases; the remaining cases are iatrogenic. The other human prion diseases are Gerstmann-Sträussler-Scheinker disease and fatal familial insomnia, both extremely rare hereditary disorders, and kuru, a disease seen in Papua New Guinea and acquired via ritualistic cannibalism. CJD occurs worldwide, but as systematic surveillance has only been undertaken in very few countries, its incidence in much of the world is currently unknown.

Bovine spongiform encephalopathy (BSE), a TSE affecting cattle, was first reported in the United Kingdom in 1986, and over 170,000 cases have been reported since then in that country alone. Relatively small numbers of cases have also been reported in native-born cattle in Belgium, France, Ireland, Luxembourg, the Netherlands, Portugal, and Switzerland. Cases have also been reported in Canada, Denmark, the Falkland Islands, Germany, Italy, and Oman, but solely in animals imported from the United Kingdom.

In March 1996, the occurrence in the United Kingdom of 10 cases of an apparently new clinicopathologic variant of CJD (nvCJD) was announced. The temporal and geographic association with the BSE epidemic raised the possibility of a causal link. Evidence supporting this hypothesis has subsequently accumulated: (1) neuropathologic features similar to those of nvCJD are seen in macaque monkeys inoculated intracerebrally with brain material from confirmed cases of BSE; (2) transgenic mice (mice carrying only a human prion protein [PrP] gene) have now been shown to be susceptible to BSE; and (3) the biologic strain of the nvCJD agent (as defined by transmission characteristics of inbred strains of mice) and molecular ‘strain’ (as defined by the PrP glycosylation pattern) closely resemble the pattern in several animals which were naturally or experimentally infected with the BSE agent, but differ from those identified in sporadic CJD.

As the size of the human population exposed and suspectible to the BSE agent in the United Kingdom is not known, and there are uncertainties relating to the potential length and distribution of the incubation period, accurate prediction of the future number of nvCJD cases is not possible. Populations in other countries may also have been exposed to the infective agent as a result of importation of live cattle and/or cattle products or byproducts from BSE-affected countries. Thus, the possibility of a significant and perhaps geographically diverse nvCJD epidemic occurring over the next 2 decades cannot be dismissed at present.

At the time of the Consultation, a total of 24 cases of nvCJD had been reported to WHO, 23 in the United Kingdom and one case in France. Strong evidence indicates that nvCJD is linked with BSE and that the possibility of many more nvCJD cases

occurring in the future cannot be dismissed. Furthermore, and adding to the seriousness of the situation, no treatment is known to prevent the occurrence of CJD or halt disease progression. While there was concern at the possibility of a significant epidemic of nvCJD over the next 10 to 15 years, there was no lack of ideas for potential therapeutic interventions which had to be seriously evaluated. By considering them at the Consultation, WHO intended to stimulate the relevant bodies to further support research aimed at the early identification of an effective therapy. The conclusions and recommendations of the Consultation were as follows.

CJD clinical diagnosis: criteria for probable sporadic CJD

The clinical diagnosis of CJD is currently based upon the combination of progressive dementia, myoclonus, and multifocal neurologic dysfunction, associated with a characteristic period EEG. However in nvCJD, most growth hormone-related iatrogenic cases and up to 40% of sporadic cases do not have the characteristic EEG appearance. This hampers clinical diagnosis, and hence surveillance, and illustrates the need for additional diagnostic tests. Advances in CJD diagnostics have occurred in the past 2 years, in particular the assay for 14-3-3 protein in cerebrospinal fluid (CSF), which appears to have a high sensitivity and specificity for sporadic CJD diagnosis. The following criteria for probable sporadic CJD were proposed:

progressive dementia;

and

at least **two** out of the following four clinical features

- myoclonus
- visual or cerebellar disturbance
- pyramidal/extrapyramidal dysfunction
- akinetic mutism;

and

- a typical EEG during an illness of any duration;

and/or

- a positive 14-3-3 CSF assay and a clinical duration to death < 2 years;

and

- routine investigations should not suggest an alternative diagnosis.

Results from a recent study suggest that the detection of high signal from the basal ganglia on T2 and proton-density-weighted magnetic resonance imaging (MRI) supports the diagnosis of sporadic CJD. These abnormalities can be particularly prominent if a fluid attenuated inversion recovery sequence or diffusion-weighted images are obtained. The Consultation recommended that further research be conducted into the use of MRI in human TSE.

EEG interpretation

No widely agreed and validated definition of a diagnostic EEG tracing is available, leading to potential inconsistencies in case ascertainment between centres. To enhance CJD surveillance, a workable definition of a diagnostic EEG is required. It was proposed that the following criteria devised by Steinhoff and Knight be adopted now and results be further evaluated:

- strictly periodic activity
  - variations in intercomplex intervals are no higher than 500 ms
  - periodic activity is continuous for at least one 10-second period;
- bi- or triphasic morphology of periodic complexes;
- duration of majority of complexes 100 ms to 600 ms;
- periodic complexes may be generalized or lateralized but not regional or asynchronous.

New variant CJD: definition of a suspect case

New variant CJD cannot be diagnosed with certainty on clinical criteria alone at present. However, on the basis of the 23 neuropathologically confirmed cases, the diagnosis of nvCJD should be considered as a possibility in a patient with a progressive neuropsychiatric disorder with at least five out of the six clinical features given in Table 1. The suspicion of nvCJD is strengthened by the criteria given in Table 2. A patient with a progressive neuropsychiatric disorder and five out of the six clinical features in Table 1 and all of the criteria in Table 2 should be considered as a suspect case of nvCJD for surveillance purposes.

Pathologic diagnosis

The Consultation discouraged the use of cerebral biopsy in living patients except to make an alternative diagnosis of a treatable disease. It concurred with the previous WHO recommendation that instruments used for neurosurgery on patients with CJD should be destroyed. If reuse is unavoidable, instruments must be immersed in 1N NaOH or fresh undiluted hypochlorite for at least 1 hour, cleaned, and then autoclaved at 134° C for 1 hour.

A definite diagnosis of CJD, including nvCJD, is established only by neuropathologic examination. The Consultation recommended that autopsy be strongly encouraged in any suspect case of CJD. Where autopsy is not possible or permitted, postmortem biopsy of the brain should be sought.

Experience to date of the use of palatine tonsillar biopsy in CJD diagnosis is limited. Because the abnormal isoform of PrP has been detected in tonsillar tissue from patients with nvCJD but not patients with sporadic CJD, analysis of tonsillar tissue may provide potential diagnostic information in nvCJD, but requires further postmortem evaluation.

HEALTH CANADA · SANTÉ CANADA

Notifiable Diseases Summary (Preliminary) · Sommaire des maladies à déclaration obligatoire (Provisoire)

New Cases Reported from 1 July - 30 September 1998 · Nouveaux cas déclarés du 1 juillet - 30 septembre 1998

Disease / Maladie	ICD-9 / CIM-9	Canada†			Newfoundland / Terre-Neuve			Prince Edward Island / Île-du-Prince-Édouard			Nova Scotia / Nouvelle-Écosse			New Brunswick / Nouveau-Brunswick			Quebec / Québec		
		J-S J-S	Cum. 98	Cum. 97	J-S	Cum. 98	Cum. 97	J-S	Cum. 98	Cum. 97	J-S	Cum. 98	Cum. 97	J-S	Cum. 98	Cum. 97	J-S	Cum. 98	Cum. 97
AIDS-Sida	042-044			260						2			3			2			99
Amoebiasis - Amibiase	006	307	942	1367			5	1	2	1	3	19	14			5	45	135	179
Botulism - Botulisme	005.1	1	3	5														1	
Brucellosis - Brucellose	023	3	7	10															
Campylobacteriosis - Campylobactériose	008.41	4189	9121	9677	98	174	77	21	36	33	80	175	173	93	223	175	690	1980	2219
Chancroid - Chancre mou	099.0																		
Chickenpox - Varicelle	052	1789	7045	22874	52	382	532				1	21	304	1	1	2			
Chlamydia, genital - Chlamydiose génitale	099.81*	6384	20891	21956	102	282	251	41	112	105	307	915	829	256	700	567	1064	4137	4308
Cholera - Choléra	001	1	1																
Diphtheria - Diphtérie	032			1															
Giardiasis - Giardiase	007.1	1096	3013	4090	12	38	31	3	5	5	30	68	69	17	58	102	165	550	644
Gonococcal Infections - Infections gonococciques[1]	098	948	2942	3327		2	2		1	1	18	67	79	7	15	29	77	273	408
Gonococcal Ophthalmia neonatorum - Ophtalmie gonococcique du nouveau-né	098.4	6	16	45													1	2	1
Haemophilus influenzae B (all invasive) - (invasive) à H. Influenzae B[2]	320.0,038.41*	8	35	40									1					10	6
Hepatitis A - Hépatite A	070.0,070.1	177	735	1391	1	2	3		1			8	12		3	2	22	133	395
Hepatitis B - Hépatite B	070.2,070.3	413	1201	1544		1	2				14	30	25		6	12	133	451	614
Hepatitis C - Hépatite C		3310	11815	13255	11	29	24	14	16		87	289	260	52	134	95	373	1785	831
Hepatitis non-A, non-B - Hépatite non-A, non-B																			
Legionellosis - Legionellose	482.41	24	52	56				1	1		1	3	1	2	2		6	12	12
Leprosy - Lèpre	030	1	3	4															
Listeriosis (all types) - Listériose (tous genres)	027.0,771.22*	14	31	32	1	1				1				1	1				
Malaria - Paludisme	084	77	220	909								1			1	1	23	72	128
Measles - Rougeole	055	3	16	557			9						2	1	2	4		3	3
Meningitis, pneumococcal - Méningite à pneumocoques	320.1	13	31	26			1			2			1						
Meningitis, other bacterial - Autres méningites bactériennes[3,4]		10	39	136		3	2					1	2		1	3			70
Meningitis/Encephalitis viral - Méningite/encéphalite virale[5]		250	333	188					1	1		3	1	2	2		4	16	109
Meningococcal Infections - Infections à méningocoques	036	17	87	155		1	3		1		1	2	1		2	2	5	25	45
Mumps - Oreillons	072	18	76	240							1	2	2		1	2	2	6	19
Paratyphoid - Paratyphoïde	002.1-002.9	4	9	14														3	3
Pertussis - Coqueluche	033	1648	3201	2746	15	35	31	9	13	45	26	39	69	73	176	36	681	1345	504
Plague - Peste	020																		
Poliomyelitis - Poliomyélite	045																		
Rabies - Rage	071																		
Rubella - Rubéole	056	3	61	3440									2			1		1	6
Congenital Rubella - Rubéole congénitale	771.0		1	1															
Salmonellosis - Salmonellose[6]	003	1606	4463	4429	38	149	34	10	29	27	57	146	84	43	115	96	211	779	846
Shigellosis - Shigellose	004	379	944	1108			2			7	1	7	7	4	11	9	36	196	319
Syphilis, Congenital - Syphilis congénitale	090	2	3																
Syphilis, Early Latent - Syphilis, latente récente	092		3	8														1	2
Syphilis, Early Symptomatic - Syphilis, symptomatique récente	091	51	116	33							1	2	1						2
Other Syphilis - Autres syphilis	090,092-097	62	163	239							1	6	7		4	3	4	19	23
Tetanus - Tétanos	037		1	2									1						
Trichinosis - Trichinose	124	13	19	1															
Tuberculosis - Tuberculose	010-018	171	589	686	1	4	6					3	2				31	155	187
Typhoid - Typhoïde	002.0	11	37	38												1	4	11	10
Verotoxigenic E. coli - E. coli vérotoxinogènes	008.01*	538	961	801	4	4	1	4	11	8	21	70	6	19	30	13	128	261	240
Yellow Fever - Fièvre jaune	060																		

(1) Includes all 098 categories except 098.4.
(2) Includes buccal cellulitis or epiglottitis 464.3 in a child <5 years with no other causative organisms isolated.
(3) Includes encephalitis.
(4) All other categories except Haemophilus 320.2, Listeriosis 027.0, Meningococcal 036, Pneumococcal 320.1 and Tuberculosis 013.0.
(5) All categories except Measles 055, Mumps 072, Poliomyelitis 045, Rubella 056 and Yellow Fever 060
(6) Excludes Typhoid 002.0 and Paratyphoid 002.1 to 002.9.
* ICD-9 codes used in the list may be incomplete. All 5 digit codes are unofficial and are for LCDC surveillance purposes only.
† May not represent national total if data from the provinces are incomplete.

(1) Comprend toutes les rubriques 098, sauf 098,4.
(2) Comprend cellulite buccale ou épiglottite 464,3 chez un enfant < 5 ans chez qui aucun autre microorganisme causal n'a été isolé.
(3) Comprend encéphalite.
(4) Toutes les autres rubriques sauf à Haemophillus 320,2, listériose 027.0, à méningocoques 036, à pneumocoques 320,1 et tuberculeuse 013,0.
(5) Toutes les rubriques, sauf rougeole 055, oreillons 072, poliomyélite 045, rubéole 056 et fièvre jaune 060.
(6) Sauf typhoïde 002,0 et paratyphoïde 002,1 à 002,9.
* Les codes de la CIM-9 figurant dans la liste ne sont peut-être pas pomplets. Quant aux codes à 5. chiffres, ils ne sont pas officiels, ayant été établis uniquement aux fins de la surveillance du LLCM.
† Il se peut que ce chiffre ne représente pas le total national si les données provenant des provinces sont incomplètes.

New Cases Reported from 1 July - 30 September 1998 - Nouveaux cas déclarés du 1 juillet - 30 septembre 1998

Disease Maladie	ICD-9 CIM-9	Ontario			Manitoba			Saskatchewan			Alberta			British Columbia Colombie-Britannique			Yukon			Northwest Territories Territoires du Nord-ouest		
		J-S J-S	Cum. 98	Cum. 97	J-S J-S	Cum. 98	Cum. 97	J-S J-S	Cum. 98	Cum. 97	J-S J-S	Cum. 98	Cum. 97	J-S J-S	Cum. 98	Cum. 97	J-S J-S	Cum. 98	Cum. 97	J-S J-S	Cum. 98	Cum. 97
AIDS-Sida	042-044	_	_	101	_	_	_	_	_	2	_	_	34	_	_	17	_	_	_	_	_	_
Amoebiasis - Amibiase	006	140	430	729	11	39	32	16	38	34	6	33	56	84	241	309	1	2	3	_	3	_
Botulism - Botulisme	005.1	1	1	_	_	1	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	5
Brucellosis - Brucellose	023	2	5	1	_	_	_	_	_	_	_	_	4	_	_	_	_	_	_	1	2	5
Campylobacteriosis - Campylobactériose	008.41*	1619	3299	4044	86	179	175	105	219	177	454	861	584	932	1958	1998	4	6	9	7	11	13
Chancroid - Chancre mou	099.0	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_
Chickenpox - Varicelle	052	2093	6573	16757 7797	828	2181	1855	579	1754	1665	1513 750	6105 3284	4921 3733	_	_	_	32 56	66 134	126 129	190 308	470 819	232 717
Chlamydia, genital - Chlamydiose génitale	099.81*																					
Cholera - Choléra	001	1	1	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	1	_	_	_
Diphtheria - Diphtérie	032																					
Giardiasis - Giardiase	007.1	343	1026	1849	53	127	_	75	174	169	84	262	346	302	683	848	8	10	18	4	12	9
Gonococcal Infections - Infections gonococciques[1]	098	343	1153	1439	107	325	318	91	255	224	84	302	386	163	431	339	2	5	_	56	113	102
Gonococcal Ophthalmia neonatorum - Ophtalmie gonococcique du nouveau-né	098.4	5	14	37	_	_	_	_	_	_	_	_	7	_	_	_	_	_	_	_	_	_
Haemophilus influenzae B (all invasive) - (invasive) à H. Influenzae B[2]	320.0,038.41*	_	4	6	_	1	2	5	13	20	3	7	5	_	_	_	_	_	_	_	_	_
Hepatitis A - Hépatite A	070.0,070.1	54	179	353	6	29	51	8	29	182	7	59	162	79	286	229	_	1	2	_	5	_
Hepatitis B - Hépatite B	070.2,070.3	13	48	141	2	12	9	26	66	34	15	56	52	208	524	652	1	2	_	1	5	3
Hepatitis C - Hépatite C		919	3571	4994	_	_	_	170	554	463	434	1502	961	1235	3852	5561	10	60	48	5	23	18
Hepatitis non-A, non-B - Hépatite non-A, non-B		_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_
Legionellosis - Legionellose	482.41	14	28	33	_	_	1	_	_	_	_	5	9	_	_	_	_	_	_	_	1	_
Leprosy - Lèpre	030	_	1	3	1	1	_	_	_	_	_	1	1	_	_	_	_	_	_	_	_	_
Listeriosis (all types) - Listériose (tous genres)	027.0,771.22	8	23	29	_	_	_	3	5	2	1	1	_	_	_	_	_	_	_	_	_	_
Malaria - Paludisme	084	27	81	402	1	6	17	1	2	5	13	26	74	12	30	282	_	_	_	_	1	_
Measles - Rougeole	055	1	5	22	_	_	_	1	2	21	_	1	233	_	2	263	_	1	_	_	_	_
Meningitis, pneumococcal - Méningite à pneumocoques	320.1	_	_	_	2	5	2	1	4	1	3	9	8	3	8	10	_	_	1	4	5	_
Meningitis, other bacterial - Autres méningites bactériennes[3,4]		_	9	38	_	_	3	1	4	5	9	19	12	_	_	_	_	_	_	_	2	1
Meningitis/Encephalitis viral - Méningite/encéphalite virale[5]		_	1	11	99	111	20	43	56	5	91	123	29	10	19	11	_	_	_	1	1	1
Meningococcal Infections - Infections à méningocoques	036	7	33	58	1	5	7	_	1	12	2	14	21	_	1	_	_	1	_	1	1	6
Mumps - Oreillons	072	5	24	53	_	_	2	3	14	6	4	16	26	3	12	129	_	1	1	_	_	_
Paratyphoid - Paratyphoïde	002.1-002.9	4	5	5	_	1	2	_	_	_	_	_	4	_	_	_	_	_	_	_	_	_
Pertussis - Coqueluche	033	380	648	663	143	230	73	59	185	241	165	332	493	97	196	566	_	_	7	_	2	18
Plague - Peste	020	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_
Poliomyelitis - Poliomyélite	045	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_
Rabies - Rage	071	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_
Rubella - Rubéole	056	1	13	25	1	20	3382	_	_	11	1	24	28	_	3	3	_	_	2	_	_	_
Congenital Rubella - Rubéole congénitale	771.0			1		1																
Salmonellosis - Salmonellose[6]	003	757	2024	2120	74	147	129	84	188	171	145	432	422	175	428	475	2	5	7	10	21	18
Shigellosis - Shigellose	004	92	253	307	68	169	85	48	85	72	64	110	71	66	110	226	_	1	2	_	2	1
Syphilis, Congenital - Syphilis, congénitale	090	_	_	_	_	_	_	_	_	_	2	3	_	_	_	_	_	_	_	_	_	_
Syphilis, Early Latent - Syphilis, latente récente	092	_	2	3	_	_	_	_	_	1	_	_	2	_	_	_	_	_	_	_	_	_
Syphilis, Early Symptomatic - Syphilis, symptomatique récente	091	2	3	9	_	_	_	_	_	_	1	2	5	47	109	16	_	_	_	_	_	_
Other Syphilis - Autres syphilis	090,092-097	51	112	162	_	_	_	_	_	_	6	21	44	_	_	_	_	1	_	_	_	_
Tetanus - Tétanos	037	_	1	1	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_
Trichinosis - Trichinose	124																			13	19	1
Tuberculosis - Tuberculose	010-018	53	174	151	_	_	_	_	_	_	_	_	_	83	229	313	1	2	1	2	22	26
Typhoid - Typhoïde	002.0	7	21	22	_	1	2	_	_	_	_	2	3	_	2	_	_	_	_	_	_	_
Verotoxigenic E. coli - E. coli vérotoxinogènes	008.01*	125	251	328	35	67	53	21	32	32	116	149	114	65	86	_	_	_	_	_	_	6
Yellow Fever - Fièvre jaune	060	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_	_

SYMBOLS

. Not reportable
.. Not available
_ No cases reported

SIGNES

. À déclaration non obligatoire
.. Non disponible
_ Aucun cas déclarés

SOURCE:
Division of Disease Surveillance
Laboratory Centre for Disease Control
Health Canada
Ottawa, Ontario K1A 0L2
Tel.: (613) 957-0334

SOURCE:
Division de la surveillance des maladies transmissibles
Laboratoire de lutte contre la maladie
Santé Canada
Ottawa (Ontario) K1A 0L2
Tél.: (613) 957-0334

Continued on page F-5

Continued from page F-2

Table 1
Clinical features for definition of a suspect case of nvCJD

- Early psychiatric symptoms
- Early persistent paresthesia/dysesthesia
- Ataxia
- Chorea/dystonia or myoclonus
- Dementia
- Akinetic mutism

Genetic analysis

Screening cases of CJD for the mutations associated with the heriditary forms of disease raises ethic and logistic concerns. Written consent for genetic testing is considered mandatory in many countries but may be culturally unacceptable in others. The Consultation recommended that genetic counselling of patients and/or their families should be performed prior to any PrP gene analysis and that ideally written consent, or at least documented oral consent, should be obtained. The genetic counsellor should be provided with information on the genetics of the human TSE to be used when seeking consent. Because of the low PrP gene mutation detection rate in sporadic CJD, it is recommended that at present only those patients with a family history of a TSE be considered for PRP gene analysis as part of WHO's surveillance activities. Analysis could be performed at one of the proposed WHO collaborating centres (see below).

All suspect cases of nvCJD should undergo PrP gene analysis (if consent is obtained) to exclude a mutation and, for research purposes, to identify codon 129 status.

Geographic attribution of cases and proposed network of WHO collaborating centres

When a diagnosis of CJD is made, the initial geographic attribution should be the country of residence at the onset of clinical disease. Final attribution should be decided on a case-by-case basis.

As part of WHO's activities to promote the global surveillance of human TSE, the Consultation recommended that collaborating centres be established to aid in diagnosis and training.

Communication of information

The communication of any important new information to the public benefits from planning. This is particularly the case when the information is complex and has the potential to cause great concern. The Consultation recommended that each national authority plan a strategy for disseminating information that may result from CJD surveillance.

Therapy

The Consultation concluded that at present there is no available therapy known to alter the underlying disease process for any human TSE. Animal and in vitro studies have demonstrated that a number of therapeutic compounds have the potential for interfering with the underlying disease process. Although some compounds are known to delay the onset of disease (in some cases beyond the animal's natural life span), no compound is known that can cure a clinically affected animal.

Future evolution of new variant CJD

The Consultation noted that the possibility of a significant epidemic of nvCJD occurring within the next 10 to 15 years could not be dismissed and therefore emphasized that the early identification of an effective therapy was of paramount importance. Such a treatment would also offer hope to those individuals who are at risk of developing familial or iatrogenic disease.

Table 2
Criteria for definition of a suspect case of nvCJD

- Absence of a history of potential iatrogenic exposure
- Clinical duration > 6 months
- Age at onset < 50 years
- Absence of a PrP gene mutation
- EEG does not show the typical periodic appearance
- Routine investigations do not suggest an alternative diagnosis
- An MRI showing abnormal bilateral high signal from the pulvinar on axial T2- and/or proton density-weighted images

Further research

The Consultation stressed the pressing need for further research into the molecular properties of the TSE agent that could lead to potential disease-modifying compounds. In parallel, efforts should be made to identify presymptomatic diagnostic tests, to enable any future therapy to be used as early as possible in the disease course.

Source: *WHO Weekly Epidemiological Record, Vol 73, No 47, 1998.*

SURVEILLANCE FOR CREUTZFELDT-JAKOB DISEASE IN CANADA

Health Canada conducts active surveillance for Creutzfeldt-Jakob disease (CJD) through the CJD Surveillance System (CJD-SS). Information collected from the surveillance system will be used to determine if there is any risk of developing CJD as a result of receiving a blood/blood product transfusion or following tissue transplantation. As a member of an international project team, CJD-SS Canada also conducts surveillance for new variant CJD (nvCJD). To date, 48 cases reported in 1997 and 1998 have been enrolled in the study. This includes some cases that have turned out not to be CJD.

Over the next several years, CJD-SS will continue to use active surveillance methods to seek out and investigate all cases of CJD occurring in Canada. Although we expect to be notified of cases primarily through neurologists, neuropathologists, and geriatricians, we ask that **any** physician aware of a case of CJD contact the surveillance system at our toll free number **1-888-489-2999**.

CJD occurs at a worldwide rate of between 0.5 and 1 case per million population per annum. There is an even distribution by sex, and the peak age of onset is between 60 and 65 years of age. Cases in persons < 30 years of age are rare. In Canada, the epidemiologic pattern and rate of CJD remains steady and consistent with the epidemiology in the rest of the world[(1)]. Current information on the epidemiology of CJD in Canada is derived from published Statistics Canada mortality data for the years 1979 to 1996 (CJD was not listed as a cause of death before 1979).

Overall, 421 deaths attributed to CJD were recorded in Canada in the 18-year period from 1979 to 1996, ranging from 14 to 34 per year, with a 1:1 male-to-female ratio. Eighty percent of deaths occurred in persons at least 60 years old, and 50% occurred in those 60 to 69 years of age, corresponding to the peak age of onset for sporadic-type CJD[(1)]. Thirteen deaths (3%), spanning the years 1979 to 1996, were reported in persons 30 to 44 years of age. No CJD deaths have been reported in persons < 30 years old in Canada.

New variant CJD has not been reported in Canada. As of the end of October 1998, 34 cases of nvCJD have been reported worldwide: 33 from the United Kingdom, and one case from France.

Reference

1. Will RG. *Prion disease surveillance.* In: Baker HF, Ridley RM, eds. *Methods in molecular medicine: prion diseases*. Totowa NJ: Humana Press, 1996:128.

Source: *E Stratton, MSc, Division of Blood-borne Pathogens, Bureau of Infectious Diseases, LCDC, Ottawa, ON.*

Our mission is to help the people of Canada maintain and improve their health.

Health Canada

The Canada Communicable Disease Report (CCDR) presents current information on infectious and other diseases for surveillance purposes and is available through subscription. Many of the articles contain preliminary information and further confirmation may be obtained from the sources quoted. Health Canada does not assume responsibility for accuracy or authenticity. Contributions are welcome (in the official language of your choice) from anyone working in the health field and will not preclude publication elsewhere.

Scientific Advisors	Dr. John Spika	(613) 957-4243
	Dr. Fraser Ashton	(613) 957-1329
Editor-in-Chief	Eleanor Paulson	(613) 957-1788
Assistant Editor	Nicole Beaudoin	(613) 957-0841
Desktop Publishing	Francine Boucher	

Submissions to the CCDR should be sent to the Editor-in-Chief, Laboratory Centre for Disease Control, Tunney's Pasture, Address Locator 0602C2, Ottawa, Ontario K1A 0L2.

To subscribe to this publication, please contact:

Canadian Medical Association	Tel. No.:	(613) 731-8610 Ext. 2307
Member Service Centre		or (888) 855-2555
1867Alta Vista Drive	FAX:	(613) 236-8864
Ottawa, ON Canada K1G 3Y6		

Annual subscription: $83.00 (plus applicable taxes) in Canada; $109 (U.S.) outside Canada.

This publication can also be accessed electronically via the Internet using a Web browser at <http://www.hc-sc.gc.ca/hpb/lcdc>. It also can be accessed at any time from any fax machine using LCDC's FAX*link* Service by calling 1-613-941-3900.