Health Canada Santé Canada

Creutzfeldt-Jakob Disease Surveillance System in Canada

Update: CJD in Canada

May 2002

Highlights

- The 14-3-3 test must be used in conjunction with the overall clinical signs and symptoms of CJD.
- 121 confirmed cases of CJD identified by CJD-SS since inception. Incidence of CJD in Canada is as expected at 0.92 and 1.02 per million population.
- A comparison is made between the signs and symptoms of classical and variant CJD.

for further information
1-888-489-2999

New Web site
http://www.hc-sc.gc.ca/pphb-dgspsp/bbp-apdh/cj_e.html

BULLETIN!!!

Due to the recent use of new testing methods and techniques for 14-3-3 CSF protein analysis, we have noticed an increase in sensitivity of these techniques, resulting in a greater number of positive results. This phenomenon is being seen worldwide and is currently being assessed. It is therefore strongly recommended that the 14-3-3 protein analysis not be used as a screening test in the evaluation of Creutzfeldt-Jakob Disease patients, but rather as a test to be interpreted in conjunction with the overall clinical signs, symptoms and other investigations.

CJD Surveillance System Update

As of December 31, 2001 we have had 290 referrals to CJD-SS (Creutzfeldt-Jakob Disease Surveillance System) with 121 cases of definite (proven by pathology) CJD. As reflected in figure A, there has been an ever increasing number of referrals to the System since it's inception in 1998. We believe this is attributable to the increasing public awareness of this disease as well as the increased reporting by physicians to CJD-SS. As seen in figure B, 1999 and 2000 are the years for which we have the most complete data thus far. The incidence is 0.92 for 1999 and 1.04 for 2000. This incidence rate is comparable with international incidence rates of 0.5 to 1/million of population. Of the 290 referrals we have been able to obtain genetic sequencing on 86. As a result of this information we have assisted in the diagnosis of 10 familial CJD cases including 6 GSS. Since we began doing the 14-3-3 testing on CSF in our laboratory in 1998 there have been 110 specimens processed. We feel we have been effective in assisting physicians in diagnosing patients with probable CJD. This successful recruitment rate for the CJD Surveillance System is thanks to the overwhelming support from physicians caring for patients with CJD as well as families affected by this devastating disease.

Figure A

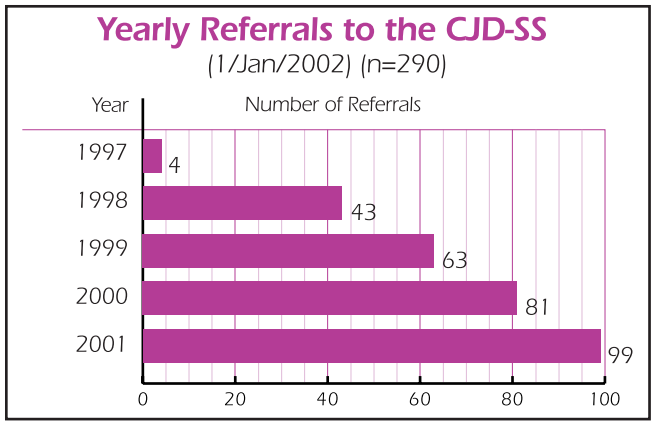
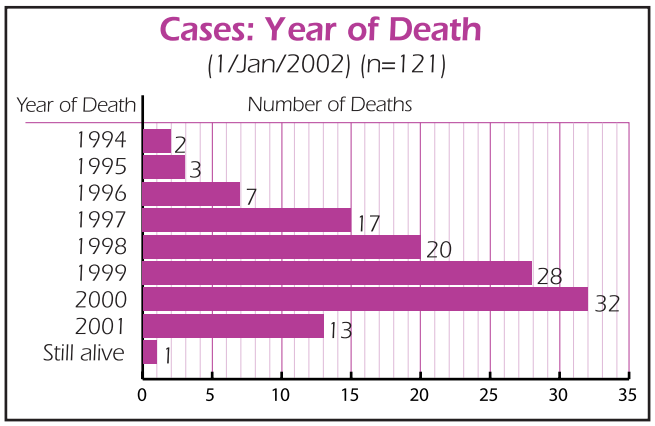


Figure B



Classical CJD and Variant CJD: Similar names, different conditions

As there seems to be some difficulty with the diagnosis of CJD versus vCJD we have decided to highlight, in this issue, the differences between CJD and vCJD as they relate to signs, symptoms and investigations.

	Classical CJD	Variant CJD (vCJD)
Age Range	onset 45 - 75 years of age	onset 12 - 74 years of age
Average Age	60 years of age	28 years of age
Average Duration	4 - 6 months	8 - 38 months
Symptoms (in order of progression)	1 - rapid progressive dementia 2 - involuntary movements (myoclonus, chorea, dystonia) 3 - akinetic mutism 4 - extrapyramidal symptoms 5 - ataxia 6 - pyramidal signs 7 - cortical blindness	1 - psychological symptoms (anxiety, depression, withdrawal) 2 - ataxia (with persistent dysesthesia) 3 - involuntary movements (myoclonus, chorea, dystonia) 4 - dementia 5 - akinetic mutism
CT	cerebral/cerebellar atrophy	usually normal
MRI	putamen and caudate hyperintensity cerebral/cerebellar atrophy	bilateral pulvinar signal (77%)
CSF	normal	normal (can have high protein)
14-3-3 (CSF)	positive (>80%)	positive (50%)
EEG	pseudoperiodic sharp waves (50-70%)	normal (or non-specific low waves)
DNA (at codon 129)	met/met 71% met/val 13% val/val 16%	met/met 100%
Pathology - brain	Both classical and variant CJD: Spongiform changes, neuronal loss, astrocytosis, protease resistant PrP	
	PrP amyloid plaques NOT seen	amyloid plaques
Pathology - tonsil	normal	positive immuno-cytochemistry for PrPsc