

Feature

Creutzfeldt-Jakob Disease: Are routine practices/standard precautions enough?

A 66 year-old man presents to the emergency room at a local hospital; his family has brought him there because of ongoing and accelerating confusion with features of dementia, as well as uncontrollable jerking. They have noticed distinct personality changes over the past year to the point where he is no longer the man they knew. Investigations at the hospital are all negative, including peripheral blood work and CSF analysis. A

logical link between the development of the disease and participation in the ritual funeral feasts in which endocannibalism was practiced. The disease had an incubation period of from 10 to 15 years with the vast majority of cases occurring within 25 years of exposure. Since the cessation of the practice of the funeral feasts in New Guinea, no cases of kuru have been documented. This link showed the transmissibility of prion diseases through ingestion of infected tissues.

CJD in that patients tended to be younger, with more neuropsychiatric symptoms and sensory disturbances. Since 1995 to October 2008, there have been 167 definite or probable cases of nvCJD in the UK. The outbreak of the disease was linked to an ongoing outbreak of bovine spongiform encephalopathy (BSE) or “mad cow” disease, a prion disease within the cattle population of the UK. The outbreak of BSE peaked in the early 1990’s and has declined steadily since that time, largely due to a number of measures in the UK including banning feeds for ruminant animals containing ruminant proteins.

Prion diseases are characterized by the accumulation of abnormal isoforms of host proteins, usually within the central nervous system, resulting in dysfunction of the neurons.

C.T. head scan showed non-specific atrophic changes. The patient went on to develop aspiration pneumonia and passed away soon after. Post-mortem brain examination revealed histological changes consistent with prion disease, and further testing confirmed Creutzfeldt-Jakob disease. The hospital approaches the Infection Control Practitioner for advice as no “special” precautions were taken for the patient.

In the 1920’s the neurologists Creutzfeldt and Jakob separately described a series of patients with neurological deterioration and the term Creutzfeldt-Jakob disease was coined. However, it wasn’t until the 1990’s that prions were implicated as the causative agent of the disorder. In 1995, cases of a new variant form of Creutzfeldt-Jacob disease (nvCJD) were reported in the United Kingdom. The nvCJD differed from classical or sporadic cases of

Transmission of CJD and Infection Prevention and Control

Despite the concern regarding the transmission of CJD and nvCJD (or transmissible spongiform encephalopathies), these diseases are not spread from person to person. In the hospital setting, routine practices/standard precautions should be adequate to prevent any nosocomial disease.

In experimental studies, no infectivity of bodily fluids have been found, including urine,

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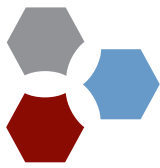
Overview of Prion Diseases

In the 1950’s, a group of atypical, slow infections were described in sheep, including scrapie, a chronic encephalitis that had been recognized in animals since the 18th century. Following the discovery of kuru, a neurodegenerative disorder endemic to the natives of Papua New Guinea, scientists and physicians noted the similarities of the disease with sheep scrapie and began to search for a transmissible cause. Kuru became the first of the prion diseases to be characterized. This group of disorders consists of Creutzfeldt-Jakob disease (CJD), new-variant Creutzfeldt-Jakob disease (nvCJD), as well as a number of rare, familial prion diseases including Gerstmann-Sträussler-Scheinker syndrome (GSS), and fatal familial insomnia (FFI). Prion diseases are characterized by the accumulation of abnormal isoforms of host proteins, usually within the central nervous system, resulting in dysfunction of the neurons.

The study of kuru revealed an epidemio-

Table 1

Infectivity Category	Tissues & Fluids	
High Infectivity	Brain Spinal cord Eye	
Low Infectivity	CSF Kidney Liver Lung Lymph nodes/spleen Placenta	
No Detectable Infectivity	Adipose tissue Adrenal gland Gingival tissue Heart muscle Intestine Peripheral nerve Prostate Skeletal muscle Testis Thyroid gland	Tears Nasal mucous Saliva Sweat Serous exudates Milk Semen Urine Faeces



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faeces, saliva, semen, mucous, milk, tears, or sweat. Although there has been some recent concern in the UK regarding three probable cases of blood-transfusion related nvCJD, in Canada there have been no documented cases of nosocomial transmission with exposure to blood in surgical or clinical contexts.

In guidelines set out by the World Health Organization (WHO), no specific risk is attributed to either healthcare workers, relatives of patients, or the community with normal clinical interaction with patients diagnosed with CJD. Isolation of patients is not required and they do not need contact or other precautions, but simply routine practice/standard precautions. No special precautions or procedures are recommended for cleaning of feeding implements, beds, linens, and the like. Biomedical waste from the patients (i.e. syringes, blood-soaked gauze) should be disposed of using standard precautions for infectious waste, according to hospital and regional guidelines. Canadian guidelines published by Health Canada in 2002 echo the WHO

recommendations, specifically following routine practice for care of patients with CJD.

Transmission of CJD and Surgical Procedures

Cases of iatrogenic spread, while rare, have been documented. CJD has been shown to have been transmitted through contaminated dural grafts, corneal transplantation, liver transplantation, cadaveric human growth hormone injections and contaminated neurosurgical equipment. In addition, there have been six cases of transmission through contaminated neurosurgical equipment, all before 1980. Although transmission through dural grafts and cadaveric growth hormone have virtually disappeared due to changes in procedures of preparing such materials, surgical instruments can still pose a risk if used for surgery on high infectivity tissues in a patient with CJD (see table 1). Unfortunately, standard sterilization techniques employed in hospitals will not be effective in removing the infectivity of the prion. Commonly used disinfection and sterilization procedures including alcohol, formaldehyde, phenolics, ethylene

oxide, boiling, and dry heat (<300°C) are all ineffective for sterilization.

Surgical equipment used for patients with known or suspected CJD should be decontaminated through a multi-step process. The WHO recommendations include: immersion in sodium hydroxide with autoclaving at 121°C for 30 minutes to one hour, then cleaning and routine sterilization following the initial treatment. Other recommended protocols include soaking instruments for one hour in NaOH or sodium hypochlorite then heating in an autoclave to 121°C for one hour, then cleaning and subjecting to routine sterilization. Many methods of sterilization recommended by the WHO and CDC (Centres for Disease Control) can cause damage, either cosmetic or severe, to surgical equipment, as well as to the autoclave itself. Steam from the NaOH can also be hazardous.

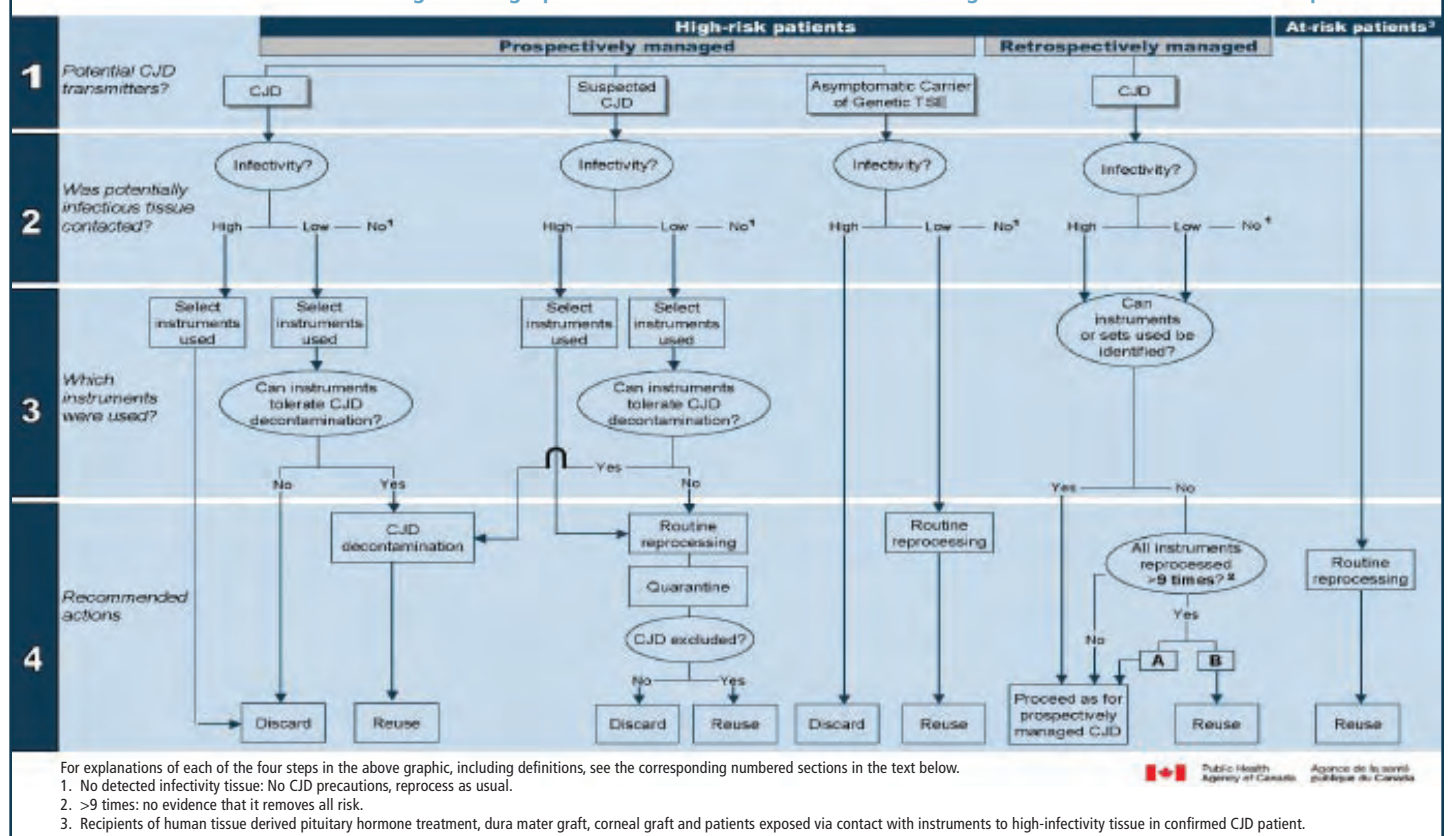
Work surfaces can also be contaminated with CJD and should be cleaned by flooding with 2N NaOH or sodium hypochlorite for one hour and then subjected to routine cleaning.

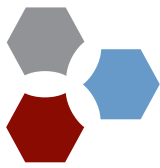
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Figure 1

Quick Reference Guide – CJD

Risk Assessment Tool – Decision algorithm- graphic version. Recommendations for management instruments used on CJD patients





Thanks

There are many volunteers who contribute to the on-going operation of the OSMT and they are an invaluable resource. The Board of Directors and staff would like to take this opportunity to thank the people who offered their time, knowledge, and commitment in 2008.

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This includes surfaces that may have been exposed to CSF (cerebrospinal fluid) of patients with CJD, such as the biosafety hood in a microbiology lab. Canadian guidelines for decontamination of surgical equipment used on patients with CJD include first thorough cleaning, soaking in 1N NaOH for one hour, or substituting with 2% NaOCl (20,000 ppm available chlorine), rinsing then sterilizing in a prevacuum-method autoclave at 134°C for 60 minutes. Recognizing the need for decision-making around surgical equipment, Health Canada has created a decision-making algorithm for use when dealing with a patient having either known or suspected CJD (see figure 1).

Perhaps the most important component in preventing the transmission of CJD through surgical equipment and procedures is the identification of patients at risk of CJD. These patients include recipients of dura mater grafts, cadaveric human growth hormone and corneal implants (especially within the 1990's) and members of families with a heritable form of transmissible spongiform encephalopathy. In addition, patients who present with an acute or severe dementia who are to undergo brain biopsies should be considered to possibly have CJD. In those patients, quarantine of surgical equipment until the diagnosis can be confirmed or refuted is recommended. Many institutions have policies to destroy such equipment if the diagnosis of TSE is confirmed to avoid the need for arduous decontamination procedures. Screening of patients undergoing

neurosurgical, spinal or eye surgery involving retinal or optic nerve for either risk factors or symptoms of CJD has been suggested in many institutions in Canada.

Conclusion

Although the outbreak of nvCJD appears to be waning, cases of sporadic and familial CJD still occur; precautions to prevent spread, particularly through surgical procedures, must be followed. Routine Practices/Standard Precautions for patient interactions in hospital are still the gold standard for prevention of nosocomial transmission, but special measures must be taken with surgical equipment, especially those used for neurosurgery or brain biopsy on patients with an acute, unexplained dementia or epidemiological links putting them at risk for disease. ❖

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