Creutzfeldt-Jakob Disease Guidelines: A Regional Approach

By C. Bauce, J. Berenyi, C. Canaon, C. Emery, C. Mindorff, J. Tyndall and A. Young

Introduction and Background

Creutzfeldt-Jakob Disease (CJD) is an infectious, progressive, degenerative neurological disorder. In addition to Standard Precautions, **Unique CJD Precautions** must be adhered to as the infectious agent is difficult to destroy. These precautions go beyond Additional Precautions. The goal is to isolate all items used on brain tissue, dura and cerebral spinal fluid (CSF) where the causative agent is harbored.

CJD is a transmissible but not a contagious disease. Daily patient care is not a risk for health care workers, except during exposure to central nervous tissue or fluids during neurological procedures, interventional neuroradiology (e.g. stereotactic brain biopsy) and autopsies.

With the restructuring of Chedoke-McMaster and the Hamilton Civic Hospitals in Hamilton Ontario, the process of standardization of policies and procedures began for the operating rooms. It soon became apparent there was a need for comprehensive regional guidelines for CJD. As a result, the Hamilton-Burlington CJD Working Group was formed. The group, which consisted of representatives from Infection Control, Operating Room staff, Surgical and Neurological nurse clinicians, and Central Processing, has composed the Creutzfeldt-Jakob Disease Guidelines based on the following approach. Infectious Disease physicians, pathologists and neurosurgeons were consulted in the process.

Consensus on the guidelines was through critical review of the current evidence of risk of transmission in a Canadian healthcare environment; current published standards of practice in North America, the United Kingdom and Australia; and principles of laboratory and patient care safety as well as expert opinion.

Comprehensive guidelines were developed to identify and care for the patient with CJD throughout the hospital stay. The intent of the guidelines was to ensure appropriate care for people affected by CJD, reduce the opportunity for transmission of CJD while providing care and to ensure a consistency of response if an exposure or potential exposure should occur. These guidelines include:

- Identification of High Risk Cases.(Appendix I).
- Risk assessment and Pre-op Checklist. (Appendix II).
- Patient care Area Protocol (Appendix III).
- Operating Room Protocol, case cart list and Biopsy Tray, contents of Devon Kwik Kit, contents of Baxter Custom Tray. (Appendix IV + a,b,c)

Abstract

Creutzfeldt-Jakob Disease (CJD) is an infectious, progressive, degenerative neurological disorder. Unique CJD Precautions must be adhered to as the infectious agent is difficult to destroy.

A regional group in Hamilton-Burlington Ontario developed CJD guidelines based on critical review of the current evidence of transmission in a Canadian healthcare environment, current published standards of practice in North America, the United Kingdom and Australia; and principles of laboratory and patient care safety as well as expert opinion.

Authors

Cathy Bauce, BA, RN, MEd,Nurse Clinician, Surgical Services, Joseph Brant Memorial Hospital, Burlington, ON.

Janina Berenyi, RN, CPN(C), Neuro Resource, Surgical Program, St. Joseph's Hospital, Hamilton.

Cellie Canaon, RN, Service Resource Nurse, Neurosurgery, Surgical Program, Hamilton Health Sciences Corp., Hamilton General Hospital, Hamilton.

Chris Emery, BSc, MSc, Manager, Central Processor, Customer Support, Hamilton Health Sciences Corporation,HendersonGeneral Hospital, Hamilton.

Catherine Mindorff, RN, MSc(Cand), Infection Control, CIC, Hamilton Health Sciences Corporation, Hamilton General Hospital, Hamilton, ON.

Judi Tyndall, RN, CPN(C),Education & Development Clinician, OR/SDS, Surgical Program, Hamilton Health Sciences Corporation, Hamilton.

Alaine Young, RN, BA, CPN(C), Education and Development Clinician - OR, Surgical Program, Hamilton Health Sciences Corporation, McMaster University Medical Centre, Hamilton, Ontario.

Appendix I

Creutzfeldt-Jakob Disease (CJD) Protocol

Identification of High Risk Cases

The risk of acquiring CJD therefore depends on the patient and the type of procedure. The following are considered high risk patients when performing neurosurgical procedures, or autopsies:

- patients with known or suspected CJD
- patients with a familial history of CJD
- patients with a history of cadaveric pituitary growth hormone therapy
- patients with a history of human dural engraftment
- patients with rapidly progressive dementia

Appendix II

Creutzfeldt-Jakob Disease (CJD)

Risk Assessment Tool/Pre-op Check List

Risk assessment for identification of possible CJD patients includes the following questions:

1. Does the patient have known/suspected CJD or exhibit any of these classic clinical symptoms?

☐ Rapid progressive presenile dementia

☐ Myoclonus (myoclonic jerks)

☐ Progressive motor dysfunction (dementia with lower motor neuron findings)

☐ Cerebellarataxia

☐ Abnormal EEG patterns with periodic complexes of repetitive triphasic spikes and slow waves at 1Hz[9]

2. Is there a family history of CJD?

Yes ☐ No ☐

3. Is there a family history of any other inheritable spongiform encephalopathy (Gertstman-Strausaler-Scheinker or Fatal Familial Insomnia)?

Yes ☐ No ☐

4. Has the patient ever received any human pituitary growth hormone therapy?

Yes ☐ No ☐

5. Does the patient have a history of receiving any human dural engraftment?

Yes ☐ No ☐

6. Does the patient have a history of receiving a human corneal transplant?

Yes ☐ No ☐

7. Does the patient have a rapidly progressive dementia not yet diagnosed?

Yes ☐ No ☐

8. Has the patient received notification from their Family Physician that they have received blood products from a CJD donor?

Yes ☐ No ☐

When the quartet of dementia, myoclonus, periodic EEG activity and rapid progression is present, the diagnosis of CJD is almost certain.

Diseases that must be ruled out are: Alzheimer's, Parkinson's, and Familial Myoclonic Dementia. When high risk is suspected, notify the surgeon and Infection Control.

Appendix III

Creutzfeldt-Jakob Disease Protocol for Patient Care Areas

Preamble:

There is no evidence that normal social contact can transmit CJD. The infectious agent is concentrated within the central nervous system consisting of the brain, spinal cord, cranial nerves, and cerebrospinal fluid.

Patients at Risk: Patients with:

- known or suspected CJD
- familial history of CJD
- history of cadaveric pituitary growth hormone therapy
- history of human dural engraftment
- rapidly progressive dementia

Protocol: ***NOTIFY INFECTION CONTROL***

Standard precautions (universal precautions) are used for general care. Confine, contain any contaminant. **The isolation of CJD patients is not necessary.**

If there is a possibility of contamination with cerebrospinal fluid, do the following:

- Obtain the pathological waste disposal box(es) from the Lab (or as designated by each site). Line the pathological waste disposal box with two yellow garbage bags. A dedicated system must be arranged to ensure the waste is taken directly to be incinerated.
- Dispose of any equipment used to deal with cerebrospinal fluid or brain tissue (ie., lumbar puncture tray, neuro drains) in biohazard waste and place in special pathological waste disposal box for incineration. Label bag "DO NOT OPEN".
- Dispose of contaminated linen and garbage in a double yellow garbage bag and label bag "DO NOT OPEN".
- Dispose of needles and other sharp articles in sharps containers. Send immediately for incineration.

Environmental Cleaning:

Same as for regular cleaning.

Appendix IV

Creutzfeldt-Jakob (CJD)

Protocol for the Operating Room

Identification of High Risk Cases:

All professionals who provide pre-operative preparation for neurosurgical patients are accountable for including an assessment for CJD.

Brain biopsies are considered a high risk procedure when performed on: patients with dementia not yet diagnosed (NYD).

By limiting the number of instruments used in a high risk case (e.g. mini craniotomy set for brain biopsies), substituting disposable equipment for reusable wherever possible, and incinerating contaminated equipment in histologically confirmed cases, the risk of transmission is drastically reduced.

Once the patient has been identified as high risk for CJD, the surgeon will:

- notify the O.R. booking clerk
- notify the pathologist (as appropriate).

The booking clerk, once notified of CJD potential will:

- ensure CJD precautions are put on the O.R. list and notify the O.R. charge nurse
- ensure special CJD pick card generated
- notify admitting or day surgery (if appropriate to facility).

The O.R. charge nurse, once notified of CJD potential will:

- notify Hamilton General O.R. to obtain the CJD kit. This kit contains:

⇒ pathological waste disposal box for contaminated instruments

⇒ signage stating CJD precautions (sign and labels)

⇒ specific instrument set for dementia NYD biopsy set (CJD precautions) (see Appendix I, II, III, IV)

⇒ DO NOT OPEN label

- notify Infection Control
- notify Central Processing/O.R. to ensure correct case cart is picked
- notify ward or SDS, PACU/ICU as appropriate

Appendix IV

Day of Surgery:

1. Central Processing/O.R. will assemble the following equipment prior to procedure:

Equipment for disposal

- YELLOW garbage bags (incinerated)
- disposable sterile gowns
- disposable boot covers
- disposable suction tips
- disposable regular cautery

2. O.R. Preparation:

Important points to remember:

- use disposable equipment wherever possible (e.g. Frazier suctions)
- DO NOT use power tools (no MIDAS REX), gigli saw may be used instead. All tissue, instruments and equipment that comes in direct contact with brain, nerve tissue or CSF is considered potentially "contaminated" with CJD. All contaminated items must be separated and isolated for quarantine and placed in a dedicated controlled monitored area (i.e., CSR).
- Items will be separated which will require a sterile table for "clean" instruments (i.e. those items that do not come in contact with any tissue that penetrates dura) and a separate sterile mayo

tray used for all instruments used on dura and brain tissue (considered "contaminated with CJD").

- Set up the pathological waste cardboard box (provided by Hamilton General) and line box with TWO yellow bags.
- Inform Environmental Services of case. Remove all other garbage bags and leave only double-lined yellow garbage bag and ONE sharps container in the room.

During The Procedure:

1. Ensure that instrument tray/container is removed and placed on the clean case cart prior to initiation of surgery.
2. The initial instruments used to reach dura (e.g. the periosteal elevator, the Hudson Brace, the Burr and Perforator or Gigli saw) should be wiped off for gross blood and tissue and placed on the "clean" back table. This can remain sterile to avoid contamination with items used once dura is entered.
3. The remaining instruments used once dura has been entered will need to remain on the separate mayo tray as "contaminated". These include but are not limited to: the cone retractor, the dura hook, the metz scissors, the tissue forcep(s), the cautery and tip, the needle driver used to close dura, etc.
4. All specimens must be fixed in formalin. The specimen should be covered with ten times its volume of formalin and then placed in a plastic bag and labelled "Query CJD". Take the specimen directly to the Pathology Department - Histology.
5. In the event that CJD is suspected or in the differential diagnosis, a frozen section is required to be sent to Pathology.

End of Case:

1. "Clean" instruments from the back table are processed as usual - placed in a basin for routine cleaning. Handled with clean gloves only.
2. All "contaminated" instruments from mayo tray are wiped off with water to remove gross contaminants and blood. ENSURE THAT INSTRUMENTS ARE DRY as they may/will be quarantined until results of pathology determine whether CJD is present.
3. These "contaminated" instruments are then placed in the pathological waste disposal box in the CJD kit provided by Hamilton General O.R.
4. The circulating nurse will secure the lid of this box and tape and label with the appropriate CJD precaution sign.
5. The sealed box with "contaminated" instruments and "clean" instruments go on the case cart. Cover cart with plastic for transport as usual.
6. Drapes in contact with possibly infected tissue or fluids must be disposed of in the yellow double-lined garbage bags.
7. The scrub nurse will suction up all fluids into the suction liner. The suction liner is placed directly into the double-lined yellow bag.
8. The cart is sent to appropriate decontamination area.
9. All disposable items should be placed in the YELLOW double-lined garbage bag (includes suction liner, cautery pencils, drapes, gloves and sharps container). This double-lined bag should be libelled "For Incineration - DO NOT OPEN". This bag must go directly to area where incineration occurs or to area designated for incineration.
10. Brain tissue/CSF specimens shall be treated as "RUSH for Permanent" specimens for pathology and identified with the "Creutzfeldt-Jakob Disease" label. These items are HAND DELIVERED ASAP to the lab.
11. The Charge nurse at the front desk will notify the lab that specimen is on the way.
12. The O.R. informs Central Processing that the case cart **is on the way.**

Environmental Cleaning:

Any hard surfaces that may have blood/CSF on them may be decontaminated with IN Sodium Hydroxide (NaOH) for 60 minutes. Do not dilute.

In dealing with surfaces contaminated with high risk tissue or fluids, absorbent material (e.g., paper towels) should be placed over the entire surface in a controlled fashion soaked with 2 M sodium hydroxide or sodium hypochlorite (20,000 ppm available chlorine). The absorbent **material** should be left in place for at least one hour with repeated wetting. The area should be carefully wiped dry by using personal protective practice measures and equipment. All paper towelling should by disposed into a dedicated, impermeable waste container or bag to be incinerated. The area can then be cleaned with a detergent germicide.

Routine Cleaning for rest of OR room.

Central Processing: (once case cart received)

- Remove labelled "CJD precaution" sealed container from case cart.
- Clean all items on case cart as per usual routine **except** sealed container
- Quarantine the box of instruments in designated cupboard until surgeon/pathology confirms diagnosis
- If negative for CJD (testing minimum 72 hours), Central Processing Coordinator to retrieve the instruments from the designated cupboard and process as routine through Decontam.
- If positive for CJD, the Central Processing Coordinator will contact appropriate source for pick up of sealed container for incineration.

Contingency Plan: (for post-procedure identification of CJD)

This section will be activated when the O.R. protocol has been breached.

1. Inform administrative line (ie., Administrator, Risk Manager, Infection Control Practitioner).
2. Generate patient exposure register if contaminated instruments have been reprocessed and circulated to other patients.

It is not possible to retrieve O.R instruments used on a CJD case if the diagnosis was missed and the instruments have gone in the general instrument reprocessing cycle.

Notification Process

Any confirmed diagnosis of spongiform encehpalopathy, specifically Creutzfeldt Jakob disease is to be reported by the pathologist or neurologist to: Creutzfeldt-Jakob Surviellance System
Division of Blood-borne Pathogens,
Bureau of infectious Diseases
Laboratory Centre for Disease Control
Health Canada
PL3005A, 511-11 Holland Avenue
Ottawa, Ontario KIA OL2
Tel. toll free 1-888-489-2999

Appendix IV a

Amount	Item **CJD Biopsy Tray**
1	Baby Weitliner
1	Curette
2	Curved mosquitoes
1	Sharp hook
1	#8 Kilner suction tip
1	Biopsy brain needle
1	Elevator Adson
1	Kerrison rongeur
1	Bayonet forceps non-tooth

Appendix IV b

Devon KWIK-KIT™
MINI-PLUS KIT ™
Reorder#7440-SJH 31147153

Contents

1ea	Kidney Basin 800cc
1ea	Heart Sponge Bowl™ 1500cc
3ea	Medicine Cup 120cc w/lid
1ea	Needle Counter BG II™ #1940
1ea	Bowl 480cc
1ea	Cautery Tipolisher™ #3200
1ea	Flexible Ruler
9ea	Labels #183
2ea	Lite Glove® * #3611
1ea	Marker #160
6ea	Round Prep Balls
1ea	Surgical Blade #15
1ea	Surgical Blade #20
1ea	Suture Bag #3570
1ea	Wrap 30X30 CSR Fold

Lot Number 82591

Appendix IV c
Baxter
Custom Tray
ER Laceration Tray
Re-Order # AC-S-LAC 1

Contents

1ea	Insert
1ea	Fenestrated Drape
2ea	Towel
5ea	Gauze, 2x2[3]
10ea	Gauze, 4x4[3]
1ea	Needle, 27x1.5
1ea	Needle, 18x1.5
1ea	Needle, 25x5/8
1ea	Syringe
1ea	Needleholder[1]
1ea	Scissors[1]
1ea	Forceps[1]
1ea	Hemostat[1]
2ea	Medicine Cup
1ea	Tray
1ea	Wrap

LOT# 596 Exp. 1/03

[3] China [1] Pakistan

WARNING: This product contains natural latex rubber which may cause reactions to some individuals.

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