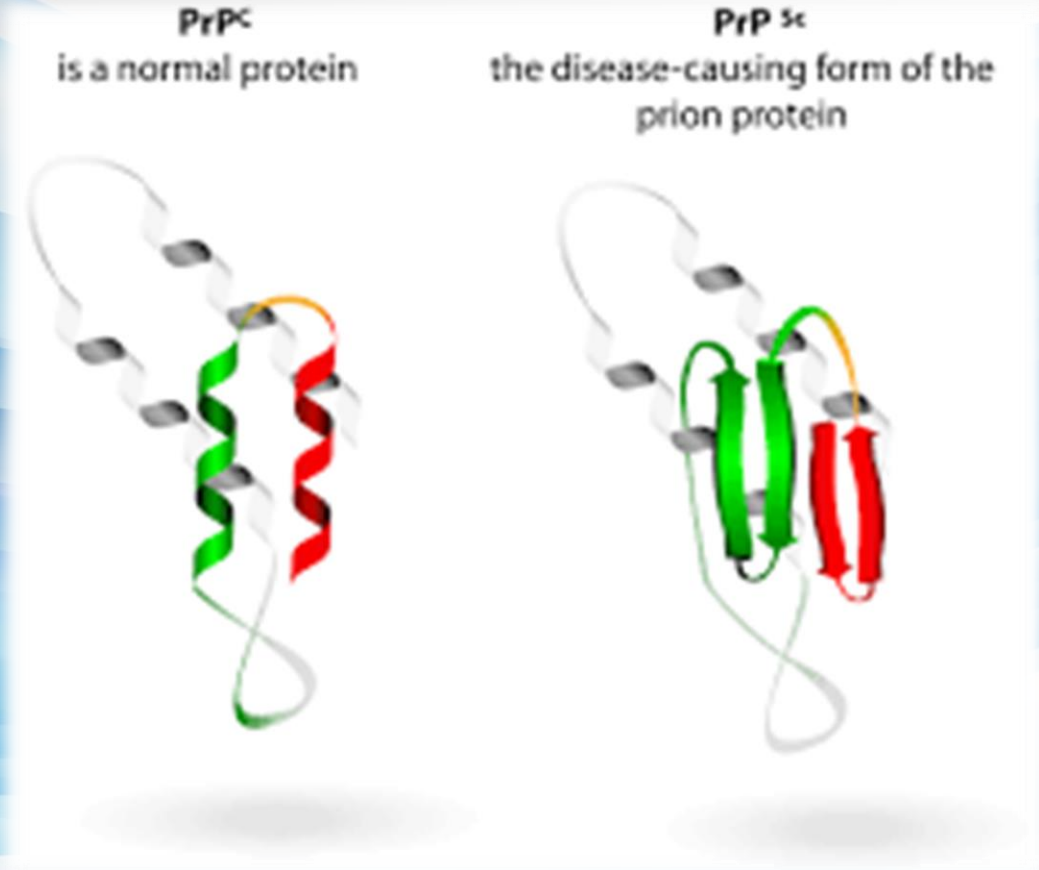

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CJD a Prion Disease

Helen Shaw, MLT, CIC
Manager, Infection Prevention & Control at BWH

MY PROMISE TO EMILY

I promise you and your family I will:

- **Respect you** as an individual on a unique healthcare journey
- **Take time** to address your concerns and calm your fears
- **Involve you** whenever decisions are being made about you
- Be your **advocate**

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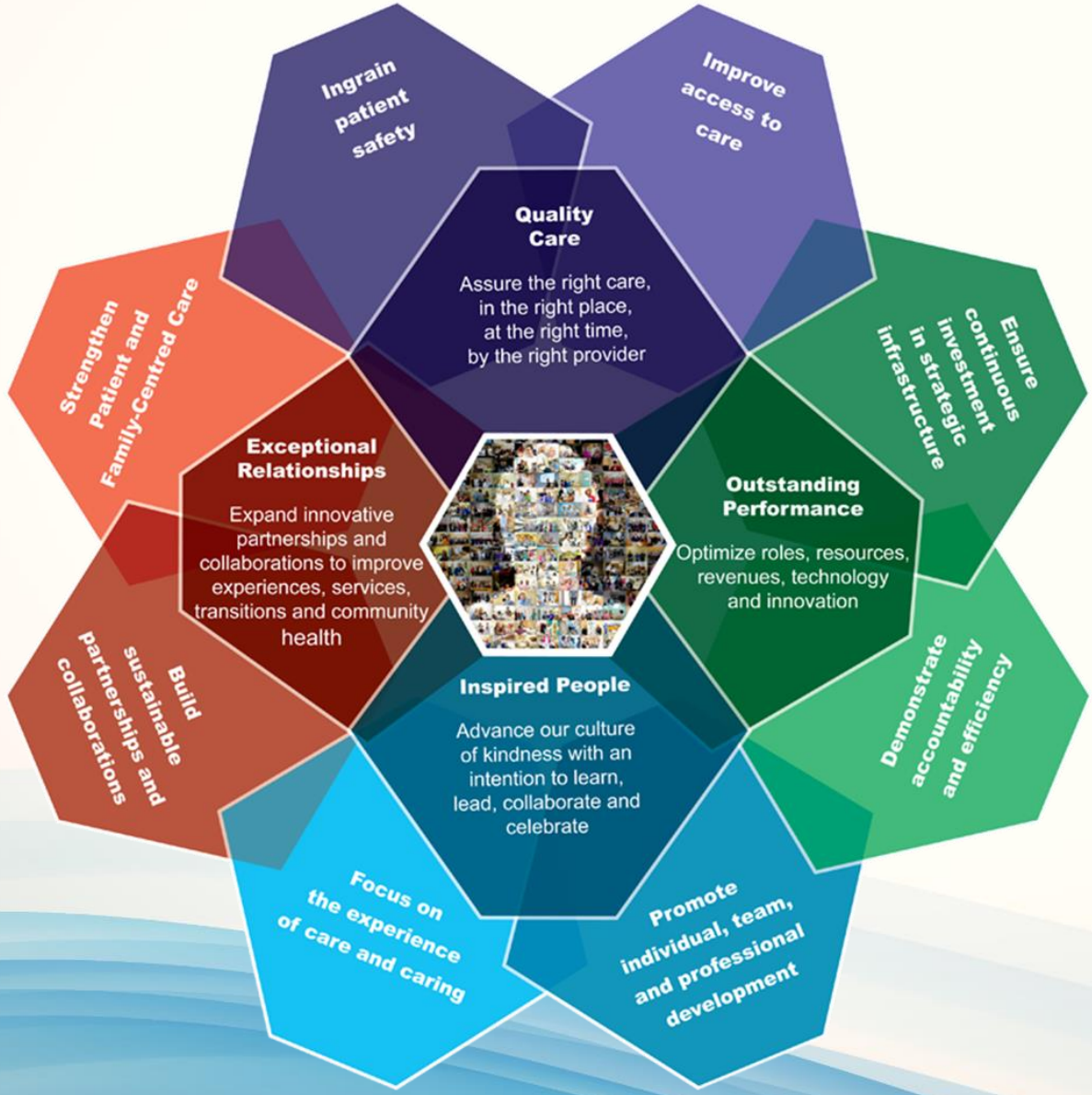
Kaleidoscope of Care



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Objective

1. What is CJD
2. How CJD presents
3. Guidelines for managing CJD in Health Care
4. Protocol for managing instruments

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Prions as Infectious Agents

- Prion protein (PrP) in its normal form is present in all humans & animals
- No nucleic acid
- In CJD, PrP is an altered, self-propagating form and becomes an infectious particle
- Can then infect non-mutated forms of the protein which initiates disease process
- Abnormal proteins aggregate which contributes to widespread neuron loss
- NOTE: the exact disease mechanism is still not known

https://www.schulich.uwo.ca/publichealth/cases/2017_Case_5.pdf

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Bovine Prion Disease – Mad Cow Disease (Bovine Spongiform Encephalopathy or BSE)

First described in Britain in 1986 and recognized as BSE in 1988

Cattle are considered natural hosts

Transmission in cattle believed to be through ingestion of contaminated feed: beef in cattle feed as a cheap protein included brains & spinal cords of dead cattle

Prions not easily destroyed by cooking or other common sterilization methods

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Human Prion Disease - CJD

- CJD Epidemic (Papua New Guinea 1957) mostly in women
  Related to customs around deceased family members where a woman ate the brains of her dead brother.
- Variant CJD Identified in 1994 in UK in unexpectedly young persons in a widespread outbreak
- Canadian CJD Surveillance System established by Health Canada in 1998 in response
- First case in Canada in 2002

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Types of Prion Disease

Classic or sporadic(sCJD)

- Endemic throughout the world with no known external source of infection
- Median age of death (USA) 68
- Not related to Mad Cow Disease
- 5-15% appears inherited mutations of prion protein gene and these are known as genetic CJD
- Diagnosis – CSF, EEG, confirmed with brain tissue biopsy or autopsy

Variant (vCJD)

- Median age of death (UK) 28
- Related to consuming infected beef or contact with contaminated tissue as a result of a medical procedure including use of contaminated neurosurgical equipment, human growth hormone, dura mater & corneal grafts
- Incubation period unknown

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How Does CJD Present Itself?

"...characterized by the accumulation of prion proteins in the brain & neural tissue degeneration, giving the tissue a spongy appearance (PHAC, 2012)

Cattle

- Initial neurological signs of apprehension, fear, easily startled, depression
- Exciteable, hypereflexia (over reaction to stimuli), hypermetria (loss of control of voluntary muscle movement); cardiac rhythm problems; heightened sensitivity of senses.
- Results in a spongy degeneration of brain & spinal cord
- No treatment or vaccine

Human

- **vCJD - acquired** (14 month avg) initial anxiety, agitation, depression, delusions & hallucinations
- 6 months later: ataxia (lack of muscle coordination affecting movement & speech) & eventual uncontrolled twitching, jerking as well as rapidly progressive dementia
- **sCJD – sporadic** (<6 months)**;** rapid decline of memory, cognition & muscle coordination & onset of dementia.
- At time of death , patients are completely immobile and mute.
- No treatment available

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Impact to Brain of CJD

https://www.google.ca/search?q=creutzfeldt-jakob+disease+pictures&sa=X&biw=1286&bih=648&tbm=isch&source=iu&ictx=1&fir=1jF2FIeMZfpDUM%253A%252CDdBEuG97qUqOWM%252C_&usg=AI4_-kS1l3WPRUikqS6Vx-Y_yR_BwbLjFA&ved=2ahUKEwjB5bO5_t3dAhUM7YMKHaszDnsQ9QEwA3oECAQQCg#imgrc=1jF2FIeMZfpDUM:

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How many cases occur?

- Worldwide - one to two cases per 1,000,000 persons (NCJDRSU, 2016)
- Canada – 40-50 new cases per year; since 1994 there have been over 800 cases of confirmed CJD in Canada with a majority of cases in Ont & Que.
- Decrease in variant CJD since beef has been controlled for BSE and protocols for health care have been implemented

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How CJD (prion disease) spreads

- Prion disease from one species (e.g. cattle) can infect another species (e.g. humans) through consummation of contaminated cattle products
- Specific surgical procedures
- No evidence that CJD can spread via social contact, sexual contact, mother-to-child, blood transfusion, or through routine health care

http://www.cchem.berkeley.edu/molsim/teaching/fall2008/prions/Site/About_Prions_2.html
https://www.slideshare.net/GBISHAJIT/bovine-spongiform-encephalopathy-by-bishajit

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The risks in Healthcare

High Infectivity Tissues

- Brain
- Spinal cord
- Dura mater
- Pituitary gland
- Posterior eye
- Spinal cord & ganglia
- Trigeminal ganglia

Low Infectivity Tissues

- Cornea
- Kidney
- Liver
- Lung
- Lymph nodes
- Placenta
- Spleen

https://www.schulich.uwo.ca/publichealth/cases/2017_Case_5.pdf

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Risks at Individual Organizations

Based on the types of surgeries/procedures done

Procedures presenting the greatest risk:

- Dura mater grafts
- Corneal grafts
- Any surgery involving neuro tissue

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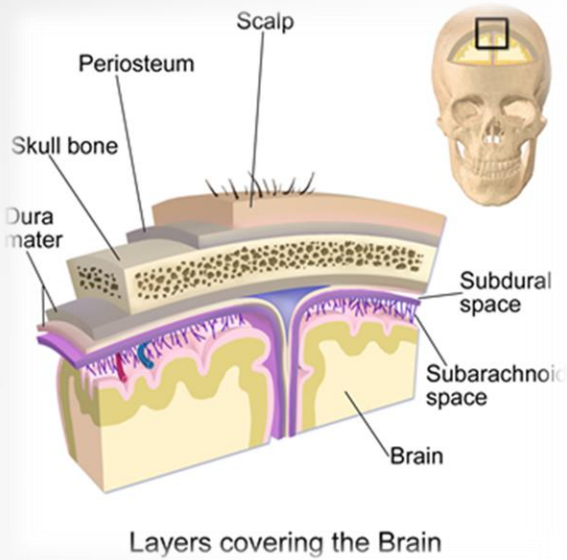
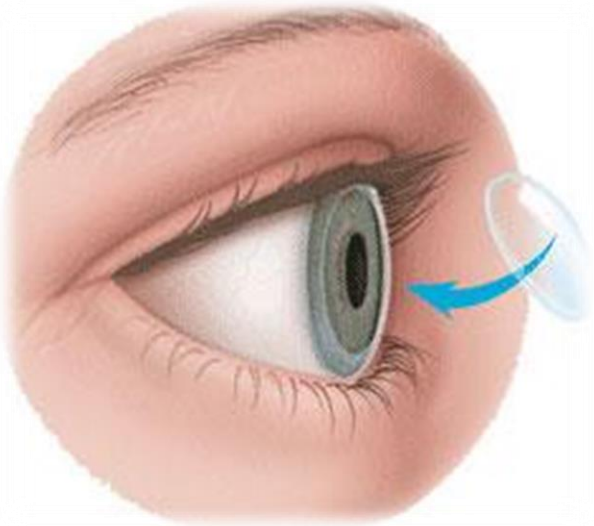


Layers covering the Brain



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Infection Control related to CJD

- Government of Canada regulates the Canadian beef industry to minimize risk of BSE contamination
- Infection Control guidelines issued in the Creutzfeldt-Jakob Disease Quick Reference Guide (PHAC, 2007) found at

http://www.phac-aspc.gc.ca/nois-sinp/pdf/cjd-eng.pdf

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WHO Guidelines

- World Health Organization has developed guidelines which are the basis of guidelines developed by other organizations:
- Public Health Agency of Canada (PHAC)
- Centre for Disease Control (CDC)
- and ultimately hospitals and MDRDs

http://www.who.int/csr/resources/publications/bse/whocdscsraph2003.pdf

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Reprocessing Surgical Instruments Used on Suspected or Confirmed CJD Patients

WHO Guidelines

Prions are resistant to routine sterilization methods.

If single-use instruments are not available, maximum safety is attained by destruction of re-usable instruments. Where destruction is not practical, re-usable instruments must be handled as per Table 5 and must be decontaminated as per Section 6 and Annex III.

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PHAC

Prospective Management:

- Surgical instruments that have been in contact with high risk tissue from suspected CJD persons, such as the brain, spinal cord, cornea, retina, pituitary gland, dura mater, and CSF should be reprocessed routinely and quarantined until diagnosis confirmed/ruled out
- Any surgical instruments that have contacted high risk tissue in a confirmed case of CJD should be discarded.

Retrospective management:

- If instruments have been identified , manage as for prospective CJD
- If instruments not identifiable, if instruments reprocessed at least 9 times, reuse; if less than 9 times manage as prospective CJD

http://www.phac-aspc.gc.ca/nois-sinp/pdf/cjd-eng.pdf

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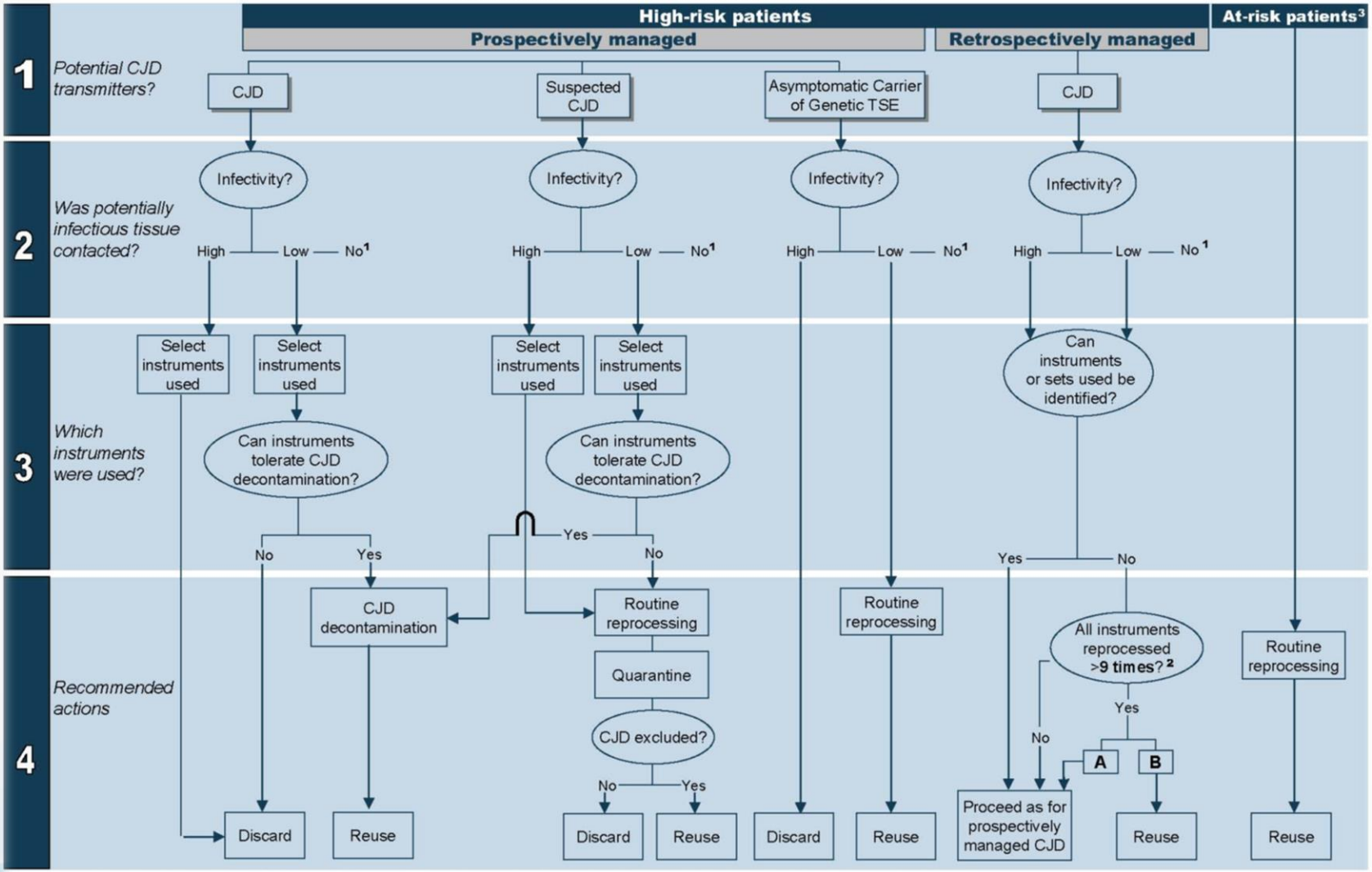
Public Health Agency of Canada | Agence de la santé publique du Canada

Quick Reference Guide - CJD

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



For explanations of each of the four steps in the above graphic, including definitions, see the corresponding numbered sections in the text below.

1 No detected infectivity tissue: No CJD precautions, reprocess as usual.

2 >9 times: no evidence that it removes all risk.

3 Recipients of human tissue derived pituitary hormone treatment, dura mater graft, corneal graft and patients exposed via contact with instruments to high-infectivity tissue in a confirmed CJD patient.

http://www.phac-aspc.gc.ca/nois-sinp/pdf/cjd-eng.pdf

CDC

One of the three most stringent chemical and autoclave sterilization methods outlined in Annex III of the WHO guidelines …should be used to reprocess heat-resistant instruments that come in contact with high infectivity tissues (brain, spinal cord, and eyes) and low infectivity tissues (cerebrospinal fluid, kidneys, liver, lungs, lymph nodes, spleen, olfactory epithelium, and placenta) of patients with suspected or confirmed CJD.

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MDR at BWH

Infection Prevention & Control/ MDR Policy # 9-4-70

(includes risks of known asymptomatic carriers of the genetic form)

- Isolation not required, although all persons providing care should be aware
- Use disposable instruments if possible
- Preferred CJD decontam:

  Clean; soak in 1N NaOH solution for 1 hour: rinse; sterilize in a prevacuum method autoclave at 134C for 60 minutes
- Cannot use on instruments that contain plastic or electronic devices
- If an instrument cannot be processed, it can be quarantined until diagnosis is confirmed. Discard if CJD confirmed.

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Consequences of not decontaminating CJD

- Additional CJD cases, but may not know about them for many years
- Possible CJD outbreak

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The Lighter Side

https://www.youtube.com/watch?v=Uy7s6gmgNhU

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QUESTIONS

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