Creutzfeldt-Jakob Disease

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Risk

- Iatrogenic transmission (actual and theoretical)
  - Blood products: blood transfusion (3 reported cases). Greater risk with pooled products (since 1999 UK has imported blood fractions) or massive blood transfusion. Risk mitigated by deferral of recipients as donors (initiated: 1998 in France, 2005 in UK) and appropriate blood transfusion policies
  - Organ donation
  - Tissue grafts: Human dura mater (168 reported cases) or corneal grafts (3 reported)
  - Human pituitary hormones: 180 reported cases
  - Human gonadotrophin: 4 reported cases
  - Neurosurgical instruments
- Transmission via food chain (vCJD and BSE); causal link found through epidemiology, biochemical, and transmission studies
- Other transmission: Potentially through vaccines; however, bovine-derived products no longer in use

Perioperative Risks

- Risk of transmission through surgical instruments: through neurosurgical instruments (4–5 cases reported) or stereotactic intracranial probes (2 reported)
- Risk of transmission through blood transfusion: Pooled products pose a greater risk (3 cases reported)

Worry About

- Management of pts with dementia or neuropsychiatric issues
- Aspiration risk
- Nutritional status and frailty; resultant risk of poor wound healing and pressure sores
- Risk of transmission through surgical instruments

Overview

- Rare neurodegenerative disease
- Prion disease, also known as TSE
  - Four types: Sporadic (sCJD), inherited, iatrogenic, variant (vCJD)
  - Worldwide incidence 1-2:1,000,000 people per y
  - All are transmissible
- Fatal; treatment is supportive and management focused on preventing transmission
- Definitive diagnosis is histopathologic with 100% sensitivity and specificity

Etiology

- PrP^{c} occurs normally in human and animal cells; prion protein gene found on chromosome 20
- Prion disease results from accumulation of PrP^{Sc}, which may be by internal (inherited) or external (ingestion, iatrogenic) exposure
- PrP^{Sc} results in an autocatalytic process in which normal prion is converted to abnormal prion
- Animal forms of prior disease include scrapie and BSE (or "mad cow disease," resulting in vCJD, first diagnosed in 1996 in UK)

Usual Treatment

- Management is supportive for pt and family; no specific treatments exist, although various experimental options have been trialed

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Restricted mouth opening if PEG	Previous airway management problems	Airway examination	
CV	Risk of CJD as recipient of blood products	Blood transfusion history		
RESP	Recurrent pneumonia	Aspiration		CXR
GI	Swallowing difficulties Aspiration risk	Recurrent pneumonia	PEG in situ	
CNS	Psychiatric and behavioral problems (vCJD) Dementia, memory problems (sCJD > vCJD) Neurologic symptoms (vCJD > sCJD)	Psychiatric history, behavioral history from medical notes and carers Peripheral pain syndrome (vCJD) Previous human dura mater graft or human pituitary hormones/gonadotropin Family history of CJD	Neurologic examination: Ataxia Extrapyramidal and pyramidal signs Myoclonus Dysarthria Confusion	MRI: pulvinar or hockey stick sign (vCJD) CSF: positive for 14-3-3-protein, tau, S100 beta EEG: sharp wave complex (sCJD) General slowing (vCJD)
MS	Poor nutritional status Contractures		Frailty Contractures of limbs Weight/height and BMI	

Key References: Porter M, Leemans M: Creutzfeldt-Jakob disease, *Contin Educ Anaesth Crit Care Pain* 13(4):119–124, 2013; World Health Organization: Variant Creutzfeldt-Jakob disease. <http://www.who.int>, 2012.

Perioperative Implications

- Prevention of transmission is the focus; prion proteins cannot be destroyed by current sterilization techniques. WHO guidance should be adhered to.

Preoperative Preparation

- Early identification of at-risk pt and appropriate communication with and preparation of theater team.
  - Surgical procedure: Consider if there is a high risk of transmission and whether tissues to be handled are moderate or high infectivity risk. Of note, vCJD is found in lymphoreticluar tissues (e.g., tonsils), as well as cerebral and spinal tissues. This has implications for endoscopic procedures and instruments. Decide if single-use disposable instruments can be safely used for the procedure. Single-use instruments should be of the same standard and quality as multiuse instruments and cost effective.
  - Decide if surgical instruments can be safely destroyed.
  - Consider if new surgical instruments are required. In UK, children born after January 1, 1997, have a separate pool of surgical instruments.
  - Tracking system for all surgical instruments must be used.
  - When disease status is unknown or pt is considered at risk, and tissue of high risk of infectivity has been handled, surgical instruments should be quarantined until diagnosis is confirmed or excluded.
  - WHO surgical checklist can be used to highlight risk assessment.
  - System in place for linking pts with surgical equipment use.
- Decision on management of surgical instruments is based on risk of transmission.

Airway

- Prion protein may be found on lymphoreticular tissue.
  - Single-use laryngoscopes wherever possible.
  - Single-use disposable airway devices (e.g., supraglottic airways).
  - Fiber optic scopes: Single-use fiber optic scopes are now available and should be used. Where this is not possible, a tracking system should be in place so that all equipment is traceable.

Postoperative Period

- Pt isolation is not required.
- Both inhalation and IV induction and maintenance of anesthesia are appropriate and should be determined by pt-specific factors.
- Ventilators do not need to be quarantined, but single-use filters should be used as normal.
- Dispose of suction in the conventional manner.
- Sharps should be disposed of in the conventional safe manner.
- Routine postop management of pt, paying particular attention to prevention of pressure sores and maintenance of nutritional intake, as well as analgesia. Pts may not be able to express their needs.
- Pts who are identified to be "at risk" should have this communicated to them and their family with appropriate support services in place.

Anticipated Problems/Concerns

- Not correctly identifying a pt at risk and the possible iatrogenic transmission of CJD to other members of the public
- Postop confusion and delirium following general anesthesia
- Preop frailty and poor oral intake with resultant risk of pressure sores and surgical site infection