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原文作者姓名：	Zarina Bali, Rishi K. Bali & Saurabh Nagrath
通訊作者學校：	Dayanand Anglo Vedic (centenary) Dental Collage, Yamunanagar, Haryana, India
報告者姓名(組別)：	謝宗叡 Intern G組
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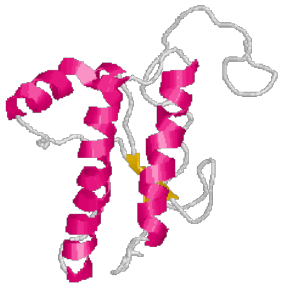
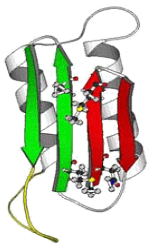
內文：

Introduction

- The term “prion” has been used since 1982 to differentiate it from infectious agents that contain nucleic acids (e.g.viruses and bacteria).
- Prion diseases are caused by the transformation of normal cell glycoprotein into a conformationally-altered isoform (PrP, prion precursor protein).
  - Conformationally-altered isoform (PrP) : partial resistance to proteolytic degradation and detergent insolubility



Normal Prion(α-helix)



Disease Prion(β-sheet)

Materials and methods

Search strategy for the identification of studies

- Using Science Direct, Hinari, Pubmed Ovid Medline, In-Process & Other Non-Indexed Citations, and, Cochrane Database of Systematic Reviews

Methods of the review

- Up to 2008, approximately 215 articles were identified in the English language literature using the search strategy.
- Articles that did not address the characteristics, the risk of transmission, and infection-control considerations in dentistry for prions were excluded.
- Further articles were identified by reviewing the references and bibliographies of articles.

General clinical aspects of prion disease

- Peculiar features of prion disease
  - Co-existence of infectious, genetic, and sporadic forms
- Up to May 2010, a total of 2617 cases of CJD were reported, with 1494 deaths.(Creutzfeldt–Jakob disease (CJD) surveillance unit, UK)
- Scrapie affecting sheep and goats was the first prion disease to be described.
- Human prion disorders are classified into CJD, Gerstmann–Straussler–Scheinker (GSS) syndrome, and Kuru.

Inherited prion diseases

- Approximately 15% of all human prion diseases
  - comprise GSS syndrome and a group of other familial human prion diseases(fatal familial insomnia)
- All mutations are inherited in an autosomal dominant manner.

Acquired prion diseases

- Kuru
  - Incurable, degenerative, neurological disorder(brain disease) that is a type of transmissible spongiform encephalopathy found in humans
  - Occurred in Papua New Guinea in the middle of the 20th century.
  - Long incubation period(several years)
  - Characteristics : cerebellar ataxia, preceded by headaches, joint pains, and shaking of the limbs, with the clinical stage lasting an average of 12 months.
- Classic CJD (Sporadic CJD (sCJD))
  - Spontaneous transformation of normal prion proteins into abnormal prions
  - Rapidly progressive and always fatal (within 1 year of the onset of illness)
  - Approximately 0.50–0.68 cases per 1 million people per year in Europe
  - Most common in the 45–75 age group, with the peak age of onset being 60–65
- Variant CJD (vCJD)
  - First described in 1996 in the UK
  - Rare and fatal transmissible spongiform encephalopathy (TSE)
  - Affects younger patients (average age is 29 year-old)
  - Relatively longer duration of illness (median of 14 months)
  - Strongly linked to exposure, probably through food
- Iatrogenic CJD (iCJD)
  - Accidental transmission of the causative agent via contaminated surgical equipment
  - Cornea or dura mater transplants
  - Administration of human-derived pituitary growth hormones
  - Less than 5% of CJD cases are iatrogenic

Dental implications of prion disease

Oral manifestations

- Dysphagia and dysarthria due to pseudobulbar palsy
- In patients with vCJD, there can be
  - Orofacial dysesthesia or paresthesia
  - Loss of taste and smell
- The mouth would seem to be rarely affected in patients with prion disease.

Infectivity and transmission risk from oral cavity

- Possible route of transmission from the brain to the oral tissues and vice versa
  - Neuronal degeneration with probable prion protein accumulation in the trigeminal ganglia of patients with sCJD
- Dental pulp originates from the richly-innervated tissue of the neural crest
→ The dental pulp of people infected with vCJD, sCJD might be infectious???
- There is evidence that infected laboratory animal models build up some level of infectivity in the oral tissues (including dental pulp, gingiva, tongue musculature, salivary glands, and trigeminal ganglion).
- There is little data to indicate that prions are transmitted within the dental clinic setting.

Potential of transmission in health-care workers

Community transmission

- No evidence to show that CJD or any other amyloidosis is transmissible from person to person by normal contact, airborne droplets, or sexual contact.
- Long incubation period of amyloidosis(e.g. vCJD)

- Premature to infer that they are not transmitted from one person to another by social contact

Transfusion of blood

- There have been 4 instances of possible transmission of vCJD infection through blood transfusions.(Donors were at a preclinical phase of the disease at the time of donation)

Occupational exposures and patient safety

- Theoretically, it is possible that health-care workers might acquire TSE from patients through inoculation injuries.
- In case of a needle stick injury while performing dental procedures on a TSE patient, the WHO common-sense actions are recommended:
  (a) Contamination of unbroken skin with internal body fluids or tissues: Wash with detergent and abundant quantities of warm water (avoid scrubbing), rinse, and dry. Brief exposure (l min, to 0.1N NaOH or a 1:10 dilution of bleach) can be considered for maximum safety)
  (b) Needle sticks or lacerations: Gently encourage bleeding; wash (avoid scrubbing) with warm soapy water, rinse, dry, and cover with a waterproof dressing. Institute further treatment (e.g. sutures) as per the type of injury. Report the injury according to normal procedures for your hospital or health-care facility/laboratory
  (c) Splashes into the eye or mouth: Irrigate with either saline (eye) or tap water (mouth); report according to normal procedures for your hospital or health-care facility/laboratory
  (d) Health and safety guidelines mandate reporting of injuries, and records should be kept for no less than 20 years.

Optional precautions for major dental work

(a) Use single-use items and equipment (e.g. needles and anesthetic cartridges)
(b) Reusable dental broaches and burs that might have become contaminated with neurovascular tissue should either be destroyed after use (by incineration) or alternatively decontaminated by a method recommended by the WHO
(c) Schedule procedures involving neurovascular tissue at the end of day to permit more extensive cleaning and decontamination.

General measures for cleaning instruments and environment

(a) Instruments should be kept moist until cleaned and decontaminated
(b) Instruments should be cleaned as soon as possible after use to minimize drying of tissues, and blood and body fluids on to the item
(c) Avoid mixing instruments used on no detectable infectivity tissues with those used on high- and low- infectivity tissues
(d) Recycle durable items for reuse only after TSE decontamination
(e) Instruments to be cleaned in automated mechanical processors must be decontaminated before processing through these machines, and the washers (or other equipment) should be run through an empty cycle before any further routine use
(f) Cover work surfaces with disposable material, which can then be removed and incinerated; otherwise clean and decontaminate underlying surfaces thoroughly
(g) Be familiar with and observe safety guidelines when working with hazardous chemicals, such as sodium hydroxide (NaOH, soda lye) and sodium hypochlorite (NaOCl, bleach)
(h) Observe manufacturers' recommendations regarding care and maintenance of equipment

Dental management of patients at high risk for CJD

- Proper medical history (including a risk assessment for CJD) should be obtained from all patients before all dental procedures
- It is important to differentiate between symptomatic patients and asymptomatic patients
- For a high-risk patient
  - Dental procedures without the involvement of the neurovascular tissues : General infection control
  - Dental procedure involves exposure to neurovascular tissues : More rigorous infection control should be followed
- Treat suspected or confirmed CJD patients
  - The patient should be appointed at the end of the day to prevent cross-infection and allow more extensive cleaning

Proper infection control in treating high-risk patients

- Prion agents resist conventional sterilization methods, such as steam autoclaving or by ethylene oxide gas.
- Human sCJD prions were more than 100000 times more resistant to inactivation than hamster prions.
- The most stringent protocol for heat-resistant instruments(WHO)
  - Immerse the instruments in sodium hydroxide (1 N NaOH) and heat in a gravity displacement autoclave at 121℃for 30 min; clean; rinse in water and subject to routine sterilization
- The protocol for surfaces and heat-sensitive, reusable instruments(WHO)
  - Flood with 2 N NaOH or undiluted sodium hypochlorite (NaOCl); let stand for 1 h; mop up and rinse with water

Conclusion

- There is no evidence to show that sCJD is transmissible
- vCJD is transmissible during the preclinical stage of the disease
- Clinical sterilization and cleaning should be of the highest standard possible
- Medical history should be obtained from all patients before all dental procedures
- Dental professionals should maintain up-to-date standards of knowledge of infection control and decontamination protocols

題號	題目
1	下列哪一種疾病和prion較無關係? (A) Creutzfeldt-Jakob disease (B) Crohn's disease (C) Gerstmann-Straussler-Scheinker syndrome (D) Scrapie
答案(B)	出處：Prion diseases : risks, characteristics, and infection control considerations in dentistry
題號	題目
2	下列何者不是Human prion disease? (A) Kuru (B) Creutzfeldt-Jakob disease (C) Scrapie (D) Fatal familial insomnia
答案(C)	出處：Prion diseases : risks, characteristics, and infection control considerations in dentistry