Two elderly patients with cognitive problems

*Inter-hospital Geriatric Meeting*

Presentation: Dr. Chan Fei
Supervisor: Dr. James Luk
Case History (1)

- M/84  Chow
- Lives with family
- Walks unaided and basic activities of daily living independent
- Retired civil servant
- Non-smoker and non-drinker
- No known drug allergy
Past medical History

- History of hypertension and BPH
- FU in GOPD
- On minipress 0.5mg BD
Chief complaint (7/09)

- Presented to QMH for dizziness and mild left side weakness for one week
- Sudden onset and dizziness related to head movement, no vertigo/tinnitus/hearing impairment

- No slurring of speech / no limb numbness
- No head injury, no fever

- Decreased gait stability but no fall
- Also subjective worsening of short term memory for 2 months e.g. forgot to close door
Physical Examination

- GC satisfactory and fully orientated
- BP 174/82 mmHg, no cyanosis/pallor/ankle edema
- Chest /CVS/ abdomen unremarkable

- CNS: Cranial nerves normal, normal tone, no cogwheel rigidity, no tremor, no nystagmus
  
- Power 4+/5 over left side, 5/5 over right side
- Left side intention tremor +ve
- Jerks and sensory normal
- Gait : small steps and unsteady
- No postural BP drop
Progress

- WBC 6.42x 10^9/L
- Hb 13.4g/dL
- Plt 144 x10^9/L
- Clotting normal
- Na/K 140/3.7 mmol/L
- Urea 12.2 mmol/L
- Creatinine 130 umol/L
- LFT and CK all normal
- Takeover by Geriatric team
- Mini-mental State Examination 28/30
- CXR: bilateral apical fibrosis
- ECG: SR, 76 bpm, Q wave over inferior lead and lead V1-3
- CT brain: mild cerebral atrophy, periventricular white matter ischaemic change
Progress

Private MRI brain + MRA carotid artery booked:
1. Bilateral periventricular hyperintensities and multiple punctate and early confluent hyperintense foci in bilateral centrum semiovale and subcortical white matter, in keeping with small vessel disease
2. Small chronic lacunar infaract in left pons
3. No significant artery stenosis found

- Treated as lacunar infaract
- Possible risk factors:
  1. HT
  2. Old age
  3. Q wave seen in ECG

In view of memory loss, Vitamin B12/folate, VDRL, TFT also checked and all normal

Fasting glucose 5.2 mmol/L
Start on cartia and zocor
Progress

- While planned for stroke rehabilitation
- Noted increased dullness, and repeat MMSE 12/30
- Patient also developed fever 38.4°C
- Repeat physical examination unremarkable

- WBC 10.36 (Neutrophil 9.3) x 10^9/L
- Derranged liver function test:
  - ALP 120 U/L
  - ALT/AST 368/246 U/L
  - GGT 61 U/L
Progress

- Septic work up done: blood culture, CXR, urine and sputum culture, NPA for virus
- CRP 9.45mg/dl, ESR 112 mm/hr
- Urgent USG abdomen showed normal liver, spleen and kidney, a small liver cyst noted
- Discussed with patient’s daughter: agreed for lumbar puncture exam to rule out chronic CNS infection e.g. TB
Progress

- LP done uneventful
- Opening pressure 23cm H2O
- Total cell count 0 x 10^6/L
- CSF / blood glucose 3.4 / 6.5 mmol/L
- CSF protein 0.38 g/L (0.12 – 0.60 g/L)
- Gram stain and culture / cryptococcal antigen test / fungal smear and culture / AFB smear and PCR / HSV PCR / cytology all negative

- EEG booked
Progress

- Started on zinnat for positive nitrites on urine microscopy, treated as UTI (with history of BPH)
- Stopped zocor
- Fever, WBC and liver function test showed improving trend

- Working diagnosis:
  1. Lacunar infaract
  2. Delirium secondary to sepsis (? UTI)

- Transfer to Grantham Hospital Geriatric Unit for further management and stroke rehabilitation
Progress in GH

- Fever and LFT normalized
- MMSE 28/30 → 12/30 → 18/30 → 16/30
- (27/7) (31/7) (12/8) (18/8)

- Poor rehabilitation progress
- Elderly mobility scale 2/20
- Mainly chairbound and ADL dependent
- Progressive dysphagia needed thickener
- Also noted occasional limb twitching

- HIV: - ve
- CEA/AFP/PSA: normal
Progress in GH

- EEG report traced: almost continuous bifrontotemporal sharp waves, sometime periodic. Change consistent with epileptiform discharge or Creutzfeldt-Jakob disease (CJD)

- Transfer back to QMH for repeat EEG +/- consider lumbar puncture for 14.3.3 protein
Summary

1. M/84, good pre-morbid function
2. Rapidly progressive dementia over 6 weeks
3. Rapid worsening of mobility from walks unaided to chairbound
4. Myoclonus occurred at late stage
Case History (2)

• M/67
• Ex-smoker and non-drinker
• Retired worker in transport industry
• Lives with wife in mainland since retired
• Basic ADL and instrumental ADL independent
• NKDA
Case History

- Good past health
- Family history unremarkable
- Not on any long term medications
Chief complaint
( Nov 2008, presented to private doctor)

1. C/o progressive slowness of motion for 4 months
2. His wife also noted that he had on and off limbs “shaking” for 4 months, gradually increased in severity
3. No limb weakness/slurring of speech
4. No fall/ incontinence
Chief complaint
Case History: chief complaint

- No fever / Headache / skin rash
- No weight loss/ poor appetite
- No herbs/OTC medications
- No Head injury

- Memory impairment (mainly short term memory) for 4 months
- Blurring of vision (exact duration not sure)
Investigations in private hospital:

- MMSE 13/30
- CBP, LRFT normal
- ESR 4
- VDRL: non reactive
- TFT / vitamin B12/ folate: All normal
Investigations in private hospital:

Report of private MRI brain with contrast:

1. Extensive hyper-intense signal seen at cortical grey matter on diffusion and T2 involving bilateral posterior temporal, occipital, parietal and superior part of temporal lobes. Changes are bilateral and symmetrical.

2. The white matter and deep grey matter nuclei are not involved

3. Likely encephalitis

4. Refer to QMH for further work up
Physical Exam in QMH

- Alert, orientated to place/person but not to time
- No fever and BP / P stable
- No pallor/jaundice/cyanosis/edema
- Chest /abd /CVS : unremarkable
- CNS: CN normal, bilateral power full, plantar reflex were down-going, no cogwheel rigidity
Further Investigations:

- ANA / ANCA: negative
- AFP 3ng/ml
- CEA 1.2ng/ml
- PSA 1.5ng/ml
- LDH 179 U/L
- HIV : -ve

LP performed:
- Opening pressure: 16mmH2O
- TCC : 0
- CSF glucose/ serum RG: 3.7/5.6mmol/L
- CSF protein: 0.5g/L
- CSF for bacteria and fungal culture, VDRL, TB PCR, HSV PCR, VDRL and cryptococcus Ag all negative
Further Investigations:

*Whole body FDG PET-CT with contrast:*

1. Few mild hyper-metabolic lymph nodes in mediastinum and hilar region, non-specific and could be reactive in nature
2. Otherwise unremarkable

- MRI brain repeated:
MRI brain:
MRI brain report:

- Abnormal signals seen involving the cortical gray at both frontal and both parietal-occipital regions.
- Findings are compatible with early CJD. DDx include chronic HSV infection.
EEG result

• 1st EEG: irregular slowing of background activities to theta range without periodic sharp waves, which can be seen in early CJD (but not specific). To repeat EEG in 6-12 weeks if CJD is suspected.

• 2nd EEG (8 weeks later): background slowing with almost persistent bilateral sharp and slow waves, suggestive of CJD.
Progress of patient

• Follow up in Feb 2009
• Noted gradually worsening of functional status
• Poor in balance and fall twice
• Confused speech and lost temper easily
• Occasionally incontinence
Progress of patient

- Follow up in May 2009
- Already bed-ridden and ADL all dependent
- Non-communicable
- Frequent choking during feeding
- Clinically admitted for nasogatric tube insertion and placement arrangement
Summary

- M/67, good past health
- From the date of first symptoms to bedbound: 11 months
- Myoclonus occurred early in the disease
Sporadic Creutzfeldt-Jakob disease (sCJD)
Introduction

• Prion diseases are neurodegenerative diseases that have long incubation periods and progress rapidly once clinical symptoms appear.

• Five human prion diseases are currently recognized:
  1. kuru,
  2. Creutzfeldt-Jakob disease (CJD),
  3. variant Creutzfeldt-Jakob disease (vCJD),
  4. Gerstmann-Straussler-Scheinker syndrome (GSS), and
  5. fatal familial insomnia (FFI)
Introduction

- Kuru (means shivering): endemic in Papua New Ginea, transmitted from person to person by ritual cannibalism

- Gerstmann-Straussler-Scheinker syndrome (GSS), and fatal familial insomnia (FFI) – very rare, few cases per 100 million population per year
Human prion diseases take
1. **sporadic (sCJJD),**
2. **iatrogenic (ICJJD),**
3. **variant (vCJJD) and**
4. **familial (fCJJD) forms**

The vast majority of CJD cases are
1. sporadic (85 to 95 percent),
2. 5 to 15 percent are fCJJD;
3. iCJJD and vCJJD accounts for fewer than 1 percent
EPIDEMIOLOGY

- Approximately one case of sporadic CJD occurs per 1,000,000 population per year with a worldwide distribution.
- The mean age for the onset of disease is between 57 and 62 years.
- Patients with vCJD and iCJD tend to be much younger.
- Up to Feb 2008, about 40 cases of CJD were detected in Hong Kong since 1996, including one case of variant CJD probably acquired in the United Kingdom.

1. Mortality from Creutzfeldt-Jakob disease and related disorders in Europe, Australia, and Canada, neurology 2005
2. Department of health, HK, 2008
Variant Creutzfeldt-Jakob disease

- Up to 2008, total 203 cases of vCJD reported worldwide
- human exposure to high concentrations of Bovine Spongiform Encephalopathy-contaminated products in the late 1980s (1981-1996)

- The average age at onset in vCJD is 29 years, contrasting to the sCJD average age of 65 years
  - vCJD typically presents with psychiatric symptoms e.g. depression, with later emergence of more typical CJD features, i.e. dementia and neurological signs

- The mean duration of illness for vCJD is longer than for sCJD (14 versus 5 months)
Iatrogenic Creutzfeldt-Jakob disease

- Iatrogenic CJD (iCJD) has followed administration of:
  1. cadaveric human pituitary hormones (growth hormone and gonadotrophin)
  2. dural graft transplants,
  3. use of dural mater in radiographic embolization procedures,
  4. corneal transplants,
  5. liver transplants, and
  6. the use of contaminated neurosurgical instruments or stereotactic depth electrodes

- Dural graft transplants and use of cadaveric pituitary hormones account for the vast majority of cases of iCJD

- Human growth hormone-related iatrogenic Creutzfeldt-Jakob disease with abnormal imaging, Arch Neurol. 2006 Feb
Pathophysiology

• Primary pathogenic agent in Creutzfeldt-Jakob disease is the abnormal prion protein

• Prion protein cellular (PrPC), a membrane-bound glycophosphatidylinositol-anchored protein found in the brain

• The normal function of PrPC is not sure, probably play a role in copper homeostasis,

• Gene (PRNP gene) encoding prion protein is located on short arm of chromosome 20

• A strong link between mutations in the PRNP gene and familial CJD

• Codon 129 (Methionine or valine) of the PRNP gene also related to the individual susceptibility to CJD
• The abnormal prion “auto”-enzymatic fashion to convert normal host prion into the abnormal isoform.

• Although prior protein cellular (PrPc) and the abnormal protease resistant scrapie form of PrP (PrPsc) have identical amino acid sequences (chemically identical but conformational different)

• How the first PrPsc appears in the host remains a mystery

• Prions are resistant to normal sterilization methods, organic solvents, formalin fixation, irradiation, and heat.
PrPSc is distinguished from PrPC by several characteristics:

1. unique 3-D conformational structure,
2. enhanced stability,
3. greater hydrophobic character,
4. protease resistance,
5. insolubility after detergent extraction,
6. deposition in lysosomes,
7. and polymerization into rod-like structures with the characteristics of amyloid.
The main histologic features of prion disease are:

1. *Spongiform change*
2. *Neuronal loss (particularly of cortical layers) without inflammation*
3. *Accumulation of the abnormal prion protein*
High power photomicrograph of classic CJD demonstrating the typical size variation of CJD vacuoles. The vacuoles that create the spongiform change in CJD.
CLINICAL FEATURES OF sCJD

- Two main clinical manifestations of sCJD are:

  1. *rapidly progressive mental deterioration*

  2. *myoclonus*
CLINICAL FEATURES OF sCJD

- Mental deterioration may be manifest as dementia, behavioral abnormalities, and deficits involving higher cortical function

1. Concentration, memory, and judgment difficulties are frequent early signs
2. Mood changes such as apathy and depression are common.
3. Sleep disturbances, particularly hypersomnia are also common.
4. With disease progression, dementia becomes dominant
CLINICAL FEATURES OF sCJD

1. Myoclonus, especially provoked by startle, is present in more than 90 percent of patients
2. May present only at late stage of disease
3. Extrapyramidal signs such as hypokinesia and cerebellar signs, occur in approximately two-thirds of patients
4. Corticospinal tract involvement develop in 40 to 80 percent of patients, including hyperreflexia, extensor plantar responses (Babinski sign), and spasticity
Risk factors of sCJD

1. Family history of CJD
2. Health workers working with brain specimens
3. Medical history of psychosis
4. History of multiple surgical procedures
5. Invasive dental treatment
Differential diagnosis of Rapidly Progressive Dementia (RPD)
Rapidly progressive dementia

- 825 RPD cases referred, many with a presumptive diagnosis of CJD.
- Diagnostic breakdown of this group was determined as 54% prion disease (37% probable or definite sporadic, 15% genetic, and 2% acquired), 28% undetermined or possible prion disease.
- 18% who had other non-prion conditions, many of which were treatable.
- The diagnostic breakdown of these non-prion RPDs was 26% neurodegenerative, 15% autoimmune, 11% infectious, 11% psychiatric, and 9% miscellaneous other, whereas 28% still were undetermined.

- Michael D et al. Memory and aging center, San Francisco
Differential diagnosis of rapidly progressive dementias

**Neurodegenerative**
- CJD (sporadic, iatrogenic, familial)
- AD
- DLB
- FTD
- CBD
- Progressive supranuclear palsy (PSP)

**Infectious**
- Viral encephalitis, including herpes simplex virus
- HIV dementia
- Progressive multifocal leukoencephalopathy
- Subacute sclerosing panencephalitis (young adults)
- Fungal infections (immunosuppression [eg, central nervous system (CNS) aspergillosis])
- Syphilis
- Parasites
- Lyme disease (rarely encephalopathy)
- Balamuthia
- Whipple’s disease

**Toxic/metabolic**
- Vitamin B12 (cyanocobalamin) deficiency
- Vitamin B1 (thiamine) deficiency
- Niacin deficiency
- Folate deficiency (dementia rare)
- Uremia
- Wilson’s disease
- Portosystemic encephalopathy
- Acquired hepatocerebral degeneration
- Porphyria
- Bismuth toxicity
- Lithium toxicity
- Mercury toxicity
- Arsenic toxicity
- Electrolyte abnormalities
Differential diagnosis of rapidly progressive dementias

**Autoimmune**
- Hashimoto's encephalopathy (HE)
- Paraneoplastic (autoimmune) limbic encephalopathy (PLE)
- Nonparaneoplastic autoimmune (eg, anti-voltage-gated potassium channel [VGKC] antibodies mediated)
- Lupus cerebritis
- Other CNS vasculitides
- Sarcoid

**Neoplasms related**
- Nonautoimmune paraneoplastic conditions
- Metastases to CNS
- Primary CNS lymphoma (PCNSL)
- Intravascular lymphoma
- Lymphomatoid granulomatosis
- Gliomatosis cerebri

**Endocrine abnormalities**
- Thyroid disturbances
- Parathyroid abnormalities
- Adrenal diseases
VITAMINS mnemonic causing rapidly progressive dementias

- Vascular
- Infectious
- Toxic-metabolic
- Autoimmune
- Metastases/neoplasms
- Iatrogenic
- Neurodegenerative
- Systemic
Progress of patient 1

- Transfer back to QMH
- EEG repeated: similar changes as last EEG but periodic sharp waves appear much more abundant, compatible with CJD with progressive disease.
- Shall we repeat LP for CSF 14-3-3 protein?
Diagnosis of sCJD

- Brain biopsy remains the gold standard diagnostic test
- Protein markers: some reports suggested abnormal CSF protein serve as a marker for CJD
- One abnormal protein, the 14-3-3 protein, initially thought as a useful diagnostic test for sCJD
14-3-3 protein

• "false positive" elevations in CSF 14-3-3 have been noted with a variety of neurologic diseases:
  1. herpes simplex encephalitis,
  2. hypoxic encephalopathy,
  3. cerebral metastases,
  4. paraneoplastic disease, and
  5. metabolic encephalopathies

• Further studies suggested that the protein may be a marker of brain cell death

• The 14-3-3 protein detectable in the cerebrospinal fluid of patients with prion-unrelated neurological diseases is expressed constitutively in neurons and glial cells in culture, Eur Neurol 1999;41(4):216-25
14-3-3 protein

- subsequent case review series have found the sensitivities and specificities of range 53 to 88 percent only

- In one analysis of 1032 assays for the 14-3-3 protein, a positive result was likely found in the early stage of the disease

- Detection of CSF 14-3-3 protein in CSF should be considered an adjunctive rather than absolute test for the diagnosis of prion diseases. A negative test does not exclude the diagnosis.

Imaging

- Head CT is generally normal in sCJD

- Magnetic resonance imaging (MRI) is the most helpful in the diagnosis of CJD

- Abnormally increased T2 and fluid-attenuated inversion recovery (FLAIR) signal intensity in the putamen and head of the caudate are the most common finding on conventional MRI sequences in patients with CJD
MRI of a woman with pathologically proven sporadic CJD. Initial diffusion-weighted MRI (DWI) shows some hyperintensity in the bilateral caudate and putamen, (Figure B). DWI also shows slight hyperintensity in the medial frontal lobe cortices (Figure C). MRI two weeks later shows more apparent increased signal intensity within the bilateral basal ganglia on T2 (Figure D) and especially FLAIR images (Figure G) with characteristic posterior progression and involvement of the putamen. DWI images show increased signal intensity within the left basal ganglia (Figure E) and medial frontal lobe cortices (Figure F)
sporadic Creutzfeldt-Jakob disease

- MRI findings usually appear as bilateral increased signal intensity in the basal ganglia, corpus striatum, or thalamus, visualized best on DWI, and are less evident on fluid-attenuated inversion recovery (FLAIR) or T2-weighted scans
Diagnostic utility of MRI

• An analysis of 1036 patients with pathologically confirmed CJD found that MRI studies had an overall sensitivity of 46 percent.
• However, this study found a high specificity of MRI findings of 93 percent.
• In another study in Japan, found that the sensitivity of diffusion-weighted images (DWI) for the diagnosis of CJD was 92 percent.

- Determinants of diagnostic investigation sensitivities across the clinical spectrum of sporadic Creutzfeldt-Jakob disease. Brain 2006, Sept
Electroencephalogram

- A characteristic EEG pattern of periodic synchronous bi- or triphasic sharp wave complexes (PSWC) can be detected up to 95 percent of patients with sCJD.

- The PSWCs is characterized by the following features:
  1. Strictly periodic cerebral potentials, the majority with a duration of 100 to 600 milliseconds and an inter-complex interval of 500 to 2000 milliseconds.
  2. Generalized and lateralized complexes permitted.
  3. At least five repetitive intervals with a duration difference of <500 milliseconds required to exclude semiperiodic activity.

- The mechanism of PSWCs is not known, but similarity of PSWCs in sCJD was noted in the EEG pattern of preterm newborns. One postulation is that cortical degeneration due to sCJD may eroding the normal physiologic sleep architecture.

Electroencephalography

- In a series of autopsy confirmed (n=150) or autopsy excluded (n=56) cases of CJD were studied
- Objective EEG criteria for sCJD showed a sensitivity and specificity of 64 and 91 percent, respectively,
- Positive and negative predictive values were 95 and 49 percent.
- If applying combined EEG criteria with clinical criteria yielded an overall specificity of 98 percent
- PSWCs are not found in patients with new variant CJD (vCJD)

sporadic Creutzfeldt-Jakob disease

WHO criteria for probable sporadic Creutzfeldt-Jakob disease

1. Progressive dementia and
2. At least two of the following four:
   - A. Myoclonus
   - B. Visual or cerebellar signs or symptoms
   - C. Pyramidal or extrapyramidal signs or symptoms
   - D. Akinetic mutism

Probable CJD is defined by these criteria and periodic slow wave complexes in EEG or 14-3-3 CSF protein in patients who have disease duration for less than 2 years.

Possible CJD is defined by these criteria and the absence of EEG or CSF changes.
Progress of Patient 2

• Clinically admitted for started nasogastric tube feeding

• PEG not considered as limited life span and possibility of contamination of surgical equipment

• Not qualify for Disability Allowance as not stay in Hong Kong for one year

• Arranged Emergency placement by MSW
Progress of Patient 1

- Patient gradually develop severe dysphagia and complicated with aspiration pneumonia

- Started on nasogastric tube feeding

- Family planned to arrange old age home for him
Old Age Home arranged by patient’s daughter……

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<thead>
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<th>Particulars of Resident</th>
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<tr>
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<td>醫院／診所編號：</td>
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<tr>
<th>Part II</th>
<th>History of Major Illnesses</th>
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<td>(1)</td>
<td>Any history of major illnesses/operations?</td>
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<td>Any evidence of infectious or contagious disease?</td>
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Transmissible spongiform encephalopathy
How infectious is sCJD?

- According to available epidemiological studies, there is no evidence that CJD can be transmitted through normal social contact, sexual intercourse or by transmission from mother to child.

- History of preceding transfusion does not increase the risk of developing CJD.

- No definite cases of transfusion-associated iCJD are known to have occurred.

- One study reviewed 436 recipients of 36 blood donors who subsequently developed CJD. After 2096 person years of follow-up, no recipient had developed CJD.

References:
- Department of health and aging, Australia.
How infectious is sCJD?

- However, low levels of infectivity have been noted when whole blood, serum, or buffy coat derived from patients with sCJD is inoculated into animals.

- While it is theoretically possible that CJD can be transmitted by blood, there is no confirmed epidemiological or clinical evidence to support such claims.

- Transfusion-associated vCJD has been reported.
What should you know about the Health History Enquiry in Blood Donation?
Q24. Have you spent a total of three or more months in the UK between 1 January 1980 and 31 December 1996?
Have you spent a total of five or more years in France between 1 January 1980 and the present?
Have you spent a total of five or more years in Europe between 1 January 1980 and the present?
Have you received blood transfusion in the UK between 1 January 1980 and the present?
Have you worked or lived for a total of six or more months at US Military bases in Europe between 1 January 1980 and 31 December 1996?

Q25. Have you received bovine insulin injection since 1 January 1980?

The above questions aim at assessing the risk of donors who might have been exposed to vCJD or human equivalent of Mad Cow Disease because of their residence during 1980 to 1996. As it has been demonstrated in the animal model and there has been suspected human vCJD case transmitted through blood transfusion, theoretical risk exists. A “yes” answer to the above questions will mean deferral from blood donation for prospective blood donors.
Infections in health care workers

- CJD infection in health care workers is very rare

- Special precautions should be employed in the handling of cerebrospinal fluid as well as biopsy tissue

- Several cases have since been reported, a link to a prior exposure is difficult to establish given the long incubation period and the dependence on the recall of coworkers

- Case-control studies have not found a significant association between CJD and health care profession

- Is there evidence for exogenous risk factors in the aetiology and spread of Creutzfeldt-Jakob disease? QJM. 2000 Sep;93(9):617-31
Progress of Patient 1

- While pending suitable placement
- Patient develop another episode of hospital acquired pneumonia
- Sputum grew methicillin resistant Staphylococcus aureus (MRSA)
- Finally succumbed despite treatment with intravenous vancomycin
Prognosis of CJD

• Abrupt onset, dramatic course of neurological degeneration, rapidly progressive degenerative disease and death in months to 2 years

• A cohort of 88 patients with definite or probable sporadic CJD
• Median survival from symptom onset is \textbf{66 – 421 days}
• Shortest survival in classic sCJD phenotype and longest for affective variant CJD

• Arch Neurology 2009, Feb;66(2):208
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<tr>
<th>風險分類</th>
<th>感染</th>
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<tr>
<td>第1類</td>
<td>不屬於下述第2、3類所列的傳染病</td>
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### Category 2

#### Danger of Infection

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<th>Embalming</th>
<th>Hygienic preparation in funeral parlour</th>
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<td>殯儀館內瞻仰遺容</td>
<td>防腐處理</td>
<td>殯儀館內裝身及化粧</td>
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<tr>
<td><strong>Must</strong></td>
<td><strong>Allowed</strong></td>
<td><strong>Not allowed</strong></td>
<td><strong>Not advisable</strong></td>
</tr>
<tr>
<td>必須</td>
<td>可以</td>
<td>不可以</td>
<td>不宜</td>
</tr>
</tbody>
</table>
## Danger of Infection

<table>
<thead>
<tr>
<th>Category</th>
<th>Bagging</th>
<th>Viewing in funeral parlour</th>
<th>Embalming</th>
<th>Hygienic preparation in funeral parlour</th>
</tr>
</thead>
<tbody>
<tr>
<td>Category 1</td>
<td>Bagging:入屍袋</td>
<td>Viewing in funeral parlour:殯儀館內瞻仰遺容</td>
<td>Embalming:防腐處理</td>
<td>Hygienic preparation in funeral parlour:殯儀館內裝身及化粧</td>
</tr>
<tr>
<td></td>
<td>Not necessary:不需要</td>
<td>Allowed:可以</td>
<td>Allowed:可以</td>
<td>Allowed:可以</td>
</tr>
</tbody>
</table>

### Category 2

- In handling dead bodies, Standard Precautions are required.
- In addition, the following precautions are also required:

<table>
<thead>
<tr>
<th>Bagging</th>
<th>Viewing in funeral parlour</th>
<th>Embalming</th>
<th>Hygienic preparation in funeral parlour</th>
</tr>
</thead>
<tbody>
<tr>
<td>Must</td>
<td>Not allowed:不可以</td>
<td>Not allowed:不可以</td>
<td>Not allowed:不可以</td>
</tr>
<tr>
<td>必須</td>
<td>不可以</td>
<td>不可以</td>
<td>不可以</td>
</tr>
</tbody>
</table>

### Category 3

- In handling dead bodies, Standard Precautions are required.
- In addition, the following precautions are also required:

<table>
<thead>
<tr>
<th>Bagging</th>
<th>Viewing in funeral parlour</th>
<th>Embalming</th>
<th>Hygienic preparation in funeral parlour</th>
</tr>
</thead>
<tbody>
<tr>
<td>Must</td>
<td>Not allowed:不可以</td>
<td>Not allowed:不可以</td>
<td>Not allowed:不可以</td>
</tr>
<tr>
<td>必須</td>
<td>不可以</td>
<td>不可以</td>
<td>不可以</td>
</tr>
</tbody>
</table>
Carer Stress

1. Feeling overwhelmed
2. Lack or mixed information
3. Frustration

- Caring difficulties as rapid deterioration of activities of daily living
- Myoclonus: usually not cause pain to patient but cause significant distress to family members
- End of life issue
- Goal: provide comfort and the highest quality of life possible to patient
References

• Prion Disease, Eric Eggenberger, Neurology Clin 25 (2007) 833-842
• Blue books of Neurology: the dementia, John H. Growdon, Martin N. Rossor
• Uptodate.com