

Communicable Diseases

WATCH



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FEATURE IN FOCUS

Update of Creutzfeldt-Jakob disease in Hong Kong

Reported by Ms Sheree CHONG, Scientific Officer, Communicable Disease Surveillance and Intelligence Office, Surveillance and Epidemiology Branch, CHP.

Creutzfeldt-Jakob disease (CJD) is a rapidly progressive, invariably fatal neurodegenerative disease. It belongs to a family of human and animal diseases known as the transmissible spongiform encephalopathies (TSEs) or prion disease. CJD is the most common form of TSE in human and there are other TSEs found in specific kinds of animals. For example, bovine spongiform encephalopathy (BSE) is found in cows and is often referred to as “mad cow” disease, and scrapie affects sheep and goats.

According to the aetiology, CJD is classified into four forms¹⁻³:

- ◆ *Sporadic CJD* - It is the most common type of CJD and accounts for at least 85% of cases. The causes of the disease remain unknown;
- ◆ *Familial CJD* - It is associated with inherited mutations of the prion protein gene and makes up 5 to 15% of CJD cases;
- ◆ *Iatrogenic CJD* - It is caused by accidental transmission via the use of contaminated surgical equipment or as a result of corneal or meningeal transplants or the administration of human-derived pituitary growth hormones. This form of CJD accounts for less than 5% of cases; and
- ◆ *Variant CJD (vCJD)* – It was first reported in the United Kingdom (UK) in March 1996. The occurrence of the disease is strongly linked to the consumption of food of bovine origin contaminated with the BSE agent. There were also four cases of vCJD infection identified in the UK that were associated with blood transfusion.

CJD affects about one person in every million people per year worldwide². The onset of symptoms typically occurs above age 60 and the vast majority of CJD patients die within one year of illness onset. The incubation period of CJD is long, usually in terms of years and can be as long as 30 years. Most cases of the sporadic, familial and iatrogenic forms are seen in older people and have a relatively shorter duration of illness. These CJD cases are characterised by personality changes and progressive dementia.

On the other hand, vCJD tends to affect younger patients and tends to present with depressive symptoms or a schizophrenia-like psychosis. The duration of illness is usually longer with a median of 14 months as opposed to 4.5 months in the traditional forms of CJD¹. Patients of all four forms will then develop more and more neurological signs including unsteadiness, involuntary movements and difficulty in walking. There is no curative treatment for any forms of CJD. Management of CJD remains supportive and is aimed at alleviating symptoms and stopping the progression of the disease.

Diagnosis of CJD is based on the fulfilment of a set of clinical and epidemiological criterias as well as findings of neurological studies. The Centre for Health Protection (CHP) of the Department of Health (DH) has adopted the case definitions for the classification of human TSEs published by the World Health Organization (WHO)⁴. A CJD case is classified as a “definite”, “probable”, or “possible” case based on the clinical, laboratory and electroencephalogram criteria. These criteria can be found in the Communicable Disease Surveillance Case Definition of CHP/DH available at https://cdis.chp.gov.hk/CDIS_CENO_ONLINE/disease.html. Confirmatory diagnosis requires neuropathological and/or immunodiagnostic testing of brain tissue obtained either at biopsy or autopsy.

CJD is a rare disease in Hong Kong. It has been made notifiable since July 14, 2008. All medical practitioners are required to report any suspected and confirmed cases to CHP/DH. From July 2008 to August 2017, a total of 64 cases of CJD were recorded (Figure 1). The annual number of cases ranged from one to nine during the period from 2008 to 2016.

This article summarises the epidemiology of the CJD cases recorded in the past five years. The annual number of cases from 2012 to 2016 ranged from five to nine cases. This year, there were seven cases recorded so far (as of August 31). All of these 46 cases were sporadic form; among them, one was definite, 41 were probable and four were possible according to WHO classification. They were 17 (37.0%) males and 29 (63.0%) females. Their ages at disease onset ranged from 46 to 90 years with a median age of 70 years. Except for a Filipino lady, all other cases were of Chinese ethnicity. Among these 46 cases, 42 passed away, and CJD was the underlying cause of death for 40 of them. Two cases died of other disease conditions and four remained alive (one case reported in 2016 and three cases reported in 2017). The survival duration after disease onset of the 40 fatal cases ranged from 73 days to 846 days (median: 160 days), while most (29 cases, 72.5%) of them died within one year from disease onset.

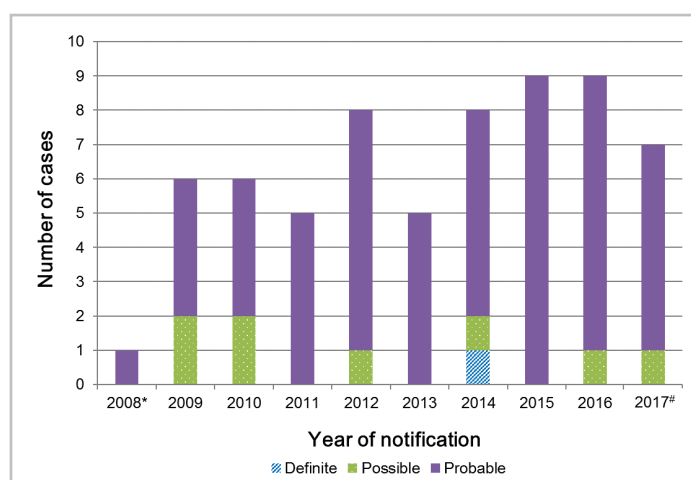


Figure 1 - Number of CJD cases in Hong Kong, 2008 (*from July 2008 after CJD was made notifiable) to 2017 ([#]as of August 31, 2017).

For the 46 cases recorded from 2012 to August 31, 2017, majority of cases (40, 90.0%) did not have history of travel to the UK. There was no family history of CJD for all the cases. They also had no known history of blood product transfusion or donation. Of the 27 cases who reported to have history of medical procedures performed, none of them was neurosurgery, corneal transplantation or injection of growth hormone.

The definite sporadic case in 2014 was a 64-year-old Chinese man, who presented with rapid decrease in cognitive function since December 2013. His brain biopsies showed spongiform encephalopathy, which was consistent with CJD, and prion protein staining showed diffuse granular staining and vacuolar staining. He had no known family history of CJD and there were no risk factors for iatrogenic CJD. All surgical equipment for the brain biopsies had gone through autoclaving cycle at 134 degree Celsius and was quarantined for re-use exclusively on this patient. After the death of this patient in January 2015, all the equipment was destroyed.

To prevent the disease from spreading, tissue or organ transplant from any CJD patients or re-use of potentially contaminated surgical instruments should be avoided. Based on the understanding that vCJD is linked to the consumption of bovine products contaminated with the BSE agent, WHO recommends that countries should not permit tissues that are likely to contain the BSE agent to enter the food chain.

References

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Review of invasive *Haemophilus influenzae* type b infection

Reported by Ms Fanny WS HO, Scientific Officer, Vaccine Preventable Disease Office, Surveillance and Epidemiology Branch, CHP.

Invasive *Haemophilus influenzae* type b (Hib) infection is caused by the bacterium *Haemophilus influenzae* type b, mostly affecting children under five years of age, though infection may occasionally occur in older age groups. The disease is usually spread by contact with nose or throat secretion of an infected person. *Haemophilus influenzae* type b can cause a range of serious infections, depending on the organ that it affects. The most common clinical manifestations caused by invasive Hib infection include meningitis, pneumonia, bacteraemia, epiglottitis, septic arthritis, cellulitis and osteomyelitis.

Invasive Hib infection is uncommon in Hong Kong as compared to western countries, and incidence has remained low since the disease became notifiable in 2008. Between January 1, 2012 and September 24, 2017, 13 cases of invasive Hib infection were reported to the Centre for Health Protection (CHP) of the Department of Health. The annual notifications varied from zero to six cases, corresponding to an incidence rate of 0 to 0.08 cases per 100 000 population per year (Figure 1). More males (eight cases, 62%) than females were affected. All cases were sporadic infections, eleven (85%) of which acquired the infection locally while two were imported from Mainland China and the Philippines.

Cases of invasive Hib infection occurred in all ages from 25 days up to 92 years (median: 21 years). Six of these cases (46%) were children, five of whom were less than two years of age. All the children were previously healthy, except a 25-day-old infant who was prematurely born at 32 weeks' gestation and cared in the neonatal intensive care unit since birth. As for the seven adult cases (aged 21 to 92 years), six of them had one or more pre-existing medical conditions, including diabetes mellitus, hypertension, hyperlipidemia, and recurrent pyogenic cholangitis, etc.

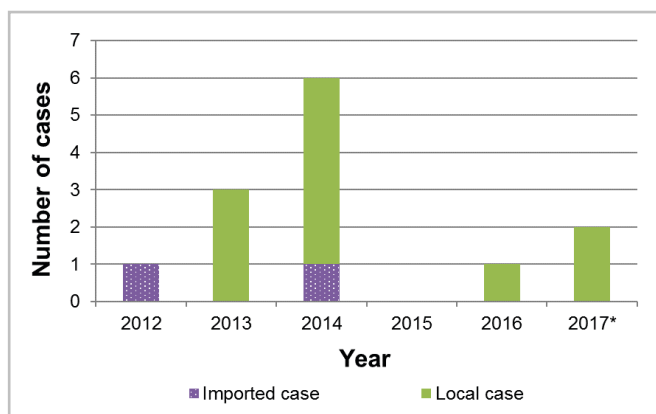


Figure 1 - Notifications of invasive Hib infection in Hong Kong, 2012-2017 (*as of September 24, 2017).

Sepsis due to Hib was the most common presentation, accounting for 69% (9/13 cases) of the cases overall. Meningitis was reported in seven (54%) cases, pneumonia in four (31%) and suppurative pericarditis with pericardial effusion and empyema in one case (4%). The diagnoses of eight (61%) cases were confirmed by blood culture, three (23%) by positive culture or detection of Hib antigen in cerebrospinal fluid, one by positive pleural fluid culture (8%), and one by culture from both blood and tracheal aspirate (8%).

All of the cases were hospitalised for treatment, six of whom required intensive care. There were two (15%) fatalities recorded. One occurred in 2013 involving a 32-year-old man with pre-existing medical conditions and was living in a residential care home for the disabled. He presented with shortness of breath and died of sepsis one day after admission. The second death was recorded in 2016 involving an 85-year-old man with underlying illnesses, who presented with pneumonia and sepsis. Blood culture of both fatal cases showed the presence of *H. influenzae* serotype b. Ten patients recovered and were discharged after 11 to 62 days of hospitalisation. The remaining patient who is still hospitalised is a 25-day-old neonate born prematurely at 32 weeks of gestation, who presented with sepsis and meningitis and required prolonged ventilator support.

In Hong Kong, the Hib vaccines are available in either monovalent form, or in combination with other vaccine components. In general, a two or three-dose primary series is given at the age of two to six months, followed by a booster dose at 12 to 18 months. Members of the public may consult their family doctors for Hib vaccination for personal protection of their children. In addition to active immunisation, good personal and environmental hygiene are essential in preventing the spread of disease. Clinicians are also encouraged to continue reporting of any suspected Hib cases to CHP. For more details on invasive Hib infection, please visit the CHP website: <http://www.chp.gov.hk/en/content/9/24/8870.html>.

NEWS IN BRIEF

A case of neonatal listeriosis

On September 11, 2017, the Centre for Health Protection (CHP) recorded a case of listeriosis affecting a 2-day-old baby girl. She was born on September 9 by emergency Caesarean section in a private hospital at 34 weeks of gestation for reduced fetal movement. She developed respiratory distress and hypotension at birth and was intubated and required inotropic support. She was transferred to a public hospital for intensive care after birth. Her blood culture collected on September 9 and 10 both grew *Listeria monocytogenes*. She was treated with antibiotics. Her condition gradually improved and was transferred to special care baby unit since September 22. She was in stable condition. Investigation revealed that the baby's mother had uneventful antenatal course and had diarrhoea on the day of delivery. Cultures of her high vaginal swab and breast milk were negative but blood and stool specimens were not collected for culture then. The mother denied consumption of high risk food item during incubation period. Other household contacts remained asymptomatic.

A sporadic case of *Streptococcus suis* infection

On September 11, 2017, CHP recorded a case of *Streptococcus suis* infection affecting a 49-year-old man with underlying illnesses. He presented with fever and shortness of breath on September 7 and was admitted to a public hospital on September 8. His blood sample collected grew *Streptococcus suis*. He was treated with antibiotics and his condition was stable. He worked as a butcher in wet markets and had handled raw pork during the incubation period. He recalled laceration injury over his left ring finger two weeks before illness onset. His home contacts and colleagues were asymptomatic.

A probable case of sporadic Creutzfeldt-Jakob disease

On September 20, 2017, CHP recorded a probable case of sporadic Creutzfeldt-Jakob disease (CJD) affecting a 66-year-old woman with underlying medical illness. She presented with progressive decline in cognitive function and delusion in May 2017. She was admitted to a public hospital on August 15 for confusion and was found to have akinetic mutism, cerebellar disturbance, myoclonus and extrapyramidal signs. Finding of electroencephalography was suggestive of CJD. Her condition was stable. She had no known family history of CJD. No risk factors for either iatrogenic or variant CJD were identified.

Field Epidemiology Training Programme (FETP) training course 2017

The Hong Kong FETP of CHP organised a five-day training course for public health professionals on the topic of “Surveillance” during September 11 to 15, 2017. The objective of the course was to equip participants with knowledge of the principles of surveillance, how to set up and evaluate surveillance systems, and skills to analyse surveillance data using relevant IT tools. The training course included presentations by the facilitators, practical exercises, case studies and hands-on practices on data analysis using different softwares. A total of 10 public health professionals attended the course and it was well received by the participants.

CA-MRSA cases in August 2017

In August 2017, CHP recorded a total of 87 cases of community-associated methicillin resistant *Staphylococcus aureus* (CA-MRSA) infection, affecting 50 males and 37 females with ages ranging from eight months to 76 years (median: 33 years). Among them, there were 64 Chinese, 4 Caucasian, 4 Indian, 4 Pakistani, 3 Filipinos, 1 Korean, 1 Nepalese, 1 Singaporean, 1 Vietnamese and 4 of unknown ethnicity.

Eighty-five cases presented with uncomplicated skin and soft tissue infections while the remaining two cases had severe CA-MRSA infections. The first severe case affected a 63-year-old man with underlying illnesses. He presented with fever, epigastric pain, diarrhoea and vomiting on July 29 and was admitted to a private hospital on the same day. Both chest X-ray and computed tomography showed right lower zone consolidation and pleural effusion. He was diagnosed to have pneumonia with septic shock and was transferred to intensive care unit of a public hospital on July 29 for further management. His sputum specimen collected on July 29 was cultured positive for CA-MRSA. His condition improved after antibiotic treatment and he was discharged on August 21. The second severe case affected a 52-year-old man with good past health. He presented with fever, cough with sputum, runny nose and sore throat on July 20. He attended the outpatient department of a private hospital for persistent symptoms on August 3. His chest X-ray showed right middle and lower zone consolidation. He was diagnosed with pneumonia and was admitted for management. His sputum specimen collected on August 3 was cultured positive for CA-MRSA. He was treated with antibiotics and was discharged on August 8.

Among the 87 cases, one was a nurse working in a public hospital. Investigation did not reveal any epidemiologically linked cases. Besides, two household clusters, with each affecting two persons, were identified in August 2017.

Scarlet fever update (August 1, 2017 – August 31, 2017)

Scarlet fever activity in August decreased as compared with that in July. CHP recorded 56 cases of scarlet fever in August as compared with 177 cases in July. The cases recorded in August included 28 males and 28 females aged between three and 20 years (median: five years). There was one institutional cluster occurring in a kindergarten, affecting two children. No fatal cases were reported in August.