

## Outbreak Investigation Express

### Amoebiasis Cluster Infection in a Mental Retardation Institute, Tainan City

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#### Abstract

In March 2011, a cluster infection of amebic dysentery has occurred in one mental retardation institute located in Tainan City. Initial physical examination for the 39 residents and 20 employees found one resident without diarrhea was positive for *Entamoeba histolytica* microscopically. He was then confirmed as a case of amebic dysentery by Taiwan Centers for Disease Control (TCDC) using polymerase chain reaction (PCR) test. To clarify the infection scale, an enzyme-linked immunosorbent assay (ELISA) and PCR test were performed for all members in the institute. Results showed 12 other residents were infected, but no hospital staff was involved. In total, 13 amoebiasis cases in the institute were confirmed with 22% attack rate. Patients in the

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institute for mentally handicapped have difficulty in dealing with their own personal hygiene and explaining their health problems, so it is a challenge for health management and infection control in the institute. Good living environment and sanitation, surveillance on common fever and diarrhea, annual physical examination, medical clearance of the mentally ill person prior to admission, and pre-employment health check-up are effective measurements to prevent the attack of gastrointestinal infection.

**Key word:** Amebic dysentery

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## Original Article

# Management and review of the first probable variant Creutzfeldt-Jakob disease in Taiwan

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### Abstract

The first probable variant Creutzfeldt-Jakob disease (vCJD) case was reported by Taiwan's Department of Health in 2010 and provoked extensive public discussion. This article elaborates current strategies and future prospects for the prevention and control of Creutzfeldt-Jakob Disease (CJD) by reviewing the management of recent probable vCJD case, history of CJD prevention in Taiwan, case reporting system, investigation procedures, disease prevention measures, case publishing standard and autopsy examination scenarios. The reporting and investigation procedures for CJD are very much alike in Taiwan and in other developed countries. This patient is the first probable vCJD case and was under investigation by our neurological experts. Based on the clinical presentation and related academic journal references, they determined this patient as a probably vCJD case. It is necessary to establish a collaborative

consultation platform with England, for example, WHO Reference Laboratories and related international experts. Accordingly, we can improve the investigation efficacy through the interchange of experiences.

### Introduction

Creutzfeldt-Jacob disease (CJD) is classified as a transmissible spongiform encephalopathy [1-3] and is a rare, progressive neurodegenerative disease. It is caused by an infectious protein called "Scrapie Prion Protein" (PrP<sup>Sc</sup>). In human, this protein can cause CJD and kuru, a disease among the Fore tribe of Papua New Guinea via cannibalism. It is also responsible for diseases in animals, like Bovine Spongiform Encephalopathies (BSE) in cattle. This protein, PrP<sup>Sc</sup>, which accumulates in cerebral tissue, destroys nerve cells and results in a spongy appearance of brain. CJD was first discovered in 1920s and the annual incidence was 0.5-1 case per million persons worldwide. There are four types of CJD: 1. Sporadic CJD; 2. Genetic CJD; 3. Iatrogenic CJD; 4. New variant CJD. The first three types are generally considered as traditional CJD, which are related to inheritance, gene mutation and medical behavior. Variant CJD (vCJD), or New Variant CJD (nvCJD), is highly related to BSE, also known as "mad cow disease" [4]. The transmission route of sporadic CJD is still unclear, but prion protein (PrP<sup>Sc</sup>) is considered the explanation. Otherwise, vCJD is contracted by human consuming products from cattle infected with BSE. CJD may also be transmitted by blood transmission or other invasive medical procedures, such as organ transplantation.

The goal of prevention and control for CJD is to implement disease surveillance and medical infection control [2, 8] and to prevent the risks of iatrogenic infection. Taiwan's Department of Health officially announced the first probable vCJD case in December 18, 2010. This article demonstrates the management and review of this case.

### **Investigation procedure of reported CJD in Taiwan**

A reporting and monitoring system of CJD has been established by former Bureau of Quarantine and Taiwan Neurological Society since 1996, and all suspected CJD cases from 1980 to 1996 were reviewed retrospectively. Until 1999, the task was succeeded by Centers for Disease Control. Taiwan Neurological Society was authorized to assemble a CJD task force in charge of investigation for reported cases and education of health care personnel. The task force consists of professors of neurology in medical centers and experts in related fields. The result of investigation is highly reliable. CJD (including CJD and vCJD) has been listed in category 4 communicable disease since 2007. The systems of case reporting, surveillance, investigation and infection control as well as reporting guidelines have been well-established [5]. According to the Law of Communicable Disease, physicians should report any CJD-suspected cases to public health authorities and provide patient's medical records within 30 days. These cases include patients who clinically have rapidly progressive dementia of unknown etiology accompanied by motor dysfunction, family

member infected with CJD, or abnormal electroencephalogram of periodic peak waves. This task force would hold meetings every month regularly to investigate every reported case.

### **CJD occurrence and prevention measures in Taiwan**

Until December 18, 2010, there were 436 reported CJD cases. 184 cases were excluded after investigation. 246 cases were determined as likely or probable CJD (including sporadic and genetic CJD). Five cases were confirmed as CJD and one as probable vCJD case. This probable vCJD patient has been living in England, a BSE high risk area, for 8 years. The exposure history of this patient was explicit, thus this case was classified as an imported CJD case.

Once a suspected CJD case is reported, nosocomial infection preventive measures should be initiated based on "Guidelines for Infectious Disease" and "Guidelines for Infection Control and Case Management of CJD and Other Human Transmissible Spongiform Encephalopathies"[5]. Besides, Taiwan CDC would inform Taiwan Blood Service Foundation and Bureau of Medical Affairs, Department of Health, by classified mail about CJD task force investigation results. Therefore, patient's records of organ donation, organ transplantation and blood donation will be tracked to prevent iatrogenic infection [2, 7-10]. If patient's blood donation record is found in blood center, the blood recipient should be monitored for health condition and restricted from blood donation. Consequently, iatrogenic CJD via blood transfusion can be avoided [9-10]. Meanwhile,

the hospital where CJD patient is diagnosed should implement nosocomial infection preventive measures immediately to prevent CJD transmission. For those who passed away during the investigation or are already determined as confirmed, probable or likely CJD, their corpses should be cremated at 1,000 °C for 30 minutes to eliminate the infectious ability of PrP<sup>Sc</sup> in case of entering the environment and food chain [11].

Department of Health amended “The Health Standard for Blood Donation” in 2000 [12]. Accordingly, people who has received blood transfusion or spent more than 3 months in England between 1980 and 1996, and those who has traveled or stayed in Europe more than 5 years after 1980, should postpone blood donation. Furthermore, CJD patient or people who has received human pituitary-derived growth hormone injection, human pituitary gonadotropins injection, bovine-derived insulin injection, a dura mater graft, or whose family member has been diagnosed CJD, should be indefinitely restricted from donating blood.

### **Reporting and investigation of the first probable vCJD case in Taiwan**

This patient was born in 1974 and went to England from 1989 to 1997 for studying. He has gradually developed symptoms of memory disorder since the second half of 2008 and was reported as a suspected CJD case in March 27, 2009 by a medical center in Taipei City. CJD investigation task force held a meeting in March 28 and determined this case as a probable CJD case according to his clinical symptoms, MRI images and electroencephalogram results. Further tonsil

biopsy study is suggested, but his family refused. This patient passed away in May 2010. With insufficient pathologic information, investigation team concluded this case as a probable sporadic CJD. However, this patient’s electroencephalogram did not show the characteristic waveform. He also has alcohol drinking problem, which may affect the clinical symptoms. Furthermore, there is no prior experience of vCJD in Taiwan. The leader of CJD task force advised this patient’s attending physician to submit an article about this case to an academic journal to obtain more opinions from other neurological experts outside the country. The article was submitted in May 2010 and then was accepted for publication in ‘Psychiatry and Clinical Neurosciences’ [13]. The CJD task force convened another meeting to re-investigate this case in December 18, 2010. This case was then determined as a probable vCJD case and was reported to WHO [14].

### **Disease preventive measures for the first probable vCJD in Taiwan**

There is no difference in disease preventive measures between vCJD and sporadic CJD. This case was reported in 2009 and public health authorities carried out the procedures immediately according to “Guidelines for Infectious Disease” and “Guidelines for Infection Control and Case Reporting of CJD and Other Transmissible Spongiform Encephalopathies.” Such measures included nosocomial infection control, medical records reviewing, patient’s blood donation record inspection, and, if any, blood recipient’s health condition monitoring and blood donation record inspection. This

probable vCJD patient had donated his blood once 10 years before his symptoms occurred. Two people received his blood. One of which died of postoperative sepsis in the next year after receiving blood transfusion. The other one is still alive without any neurologic symptoms for over 10 years. Meanwhile, because the infectious ability of PrP<sup>Sc</sup> can be destroyed at 1,000°C, the corpse of this probable vCJD patient was cremated without contaminating the environment. This patient has stayed in England, a BSE high risk area, for 8 years. Thus, it is considered as an imported CJD case. All disease preventive measures have eventually been completed and the patient's body was cremated. There is no worry about CJD transmission.

#### **Case publishing standard for CJD in Taiwan**

The purposes of CDC publishing infectious disease information are to provide the public a warning message, to launch related disease prevention measures, and to prevent disease transmission. The information can be published on the website, in statistical data system, in journals, in annual reports or by media. A press release or a press conference will be made when the information is related to public benefits and requiring public cooperation, or when there is incorrect disease information needs to be corrected. The investigation results of reported CJD-suspected cases would be released on Taiwan CDC website regularly [2-3]. This case was first listed as probable CJD case on the website in 2009, but then was revised as a probable vCJD case after the re-investigation in December 18, 2010. The

result was immediately updated on Taiwan CDC website.

#### **Autopsy examination should based on case scenario**

Referring to Article 50, paragraph 2 of Infectious Disease Control Law, whenever the cause of the infectious disease is unable to be identified or the infection is unable to be controlled without the autopsy result, public health authorities have the right to enforce autopsy which patient's family cannot defy. The notion of this law is to empower the public health authorities to proceed with autopsy for proper infection control and public benefits protection, but not to unscrupulously execute the autopsy. The public health authorities should base on the case scenario, carefully and objectively make a professional, reasonable decision for the necessity of autopsy. Autopsy should not be performed if the result is irrelevant to disease diagnosis or infection control. Furthermore, instead of penalty on family who declined the autopsy, public health authorities will support the funeral expenses for those who cooperate with the autopsy examination according to Article 50, paragraph 4 of Infectious Disease Control Law. CJD is proved that it cannot be transmitted between humans by direct contact, and there were no beef products imported to Taiwan from England. In addition, all necessary preventive measures have been fulfilled after the case was considered as a CJD probable case. Without risks on disease transmission or jeopardizing public benefits, consequently, autopsy of this case was not performed in respect of family's wish.

## Discussion

The definition of Creutzfeldt-Jakob disease used in Taiwan is based on definition made by WHO and European Union (EU) [15]. CJD task force investigates every reported case according to clinical symptoms, MRI images, electroencephalogram, 14-3-3 protein in cerebrospinal fluid (CSF) and pathological examination of the brain. The definite diagnosis of CJD requires pathological examination of the brain. Otherwise it can only be categorized as a likely or probable case. The clinical symptoms of CJD and vCJD are similar. This reported patient did not have characteristic electroencephalogram waveforms for CJD and also had a long-term alcohol drinking problem. Thus, without pathological examination of brain tissue, it was difficult to differentiate between CJD and vCJD in this case. The experience of managing vCJD is generally insufficient worldwide. It is quite common that the prior result of the CJD investigation needed to be revised after the autopsy examination. According to the data of Japan's Ministry of Health, Labour, and Welfare, the first vCJD case in Japan was reported in December 2001 because of suspected symptoms, and then died in September 2004. It was firstly considered as a sporadic CJD in September 2004, but then corrected as a vCJD case after the autopsy result came out on February 3, 2005. It took 3 years from symptoms occurred to disease confirmed as a vCJD case in Japan [16-17]. However, it took 1 year and 9 months to verify the first imported probable vCJD in Taiwan. CJD investigation team advised to submit a case report to academic journals for more

opinions from international neurological experts, so that we can establish a reference for other disputable cases in the future. Although the CJD task force in Taiwan has had sufficient experience on sporadic CJD, it is necessary to establish a collaborative consultation platform with England, or other WHO Reference Laboratories and related international experts. Accordingly, we can improve the investigation efficacy and reliability for further controversial vCJD cases from other international experts' opinions.

Taiwan CDC started to amend the previous 2003 edition of "Guidelines for Infection Control and Case Management of CJD and Other Human Transmissible Spongiform Encephalopathies" since CJD was classified as category 4 communicable disease in 2007. Moreover, Taiwan Neurological Society has provided a series of medical education courses for neurologic specialists, stomatologists and long-term care facility staffs in 2008 and 2009, to enhance medical staffs' knowledge and awareness of CJD. The number of reported CJD case was significantly increased after it is classified as communicable disease, which indicates increase in CJD surveillance sensitivity. Training of long-term care facility staffs also benefits recognition and caring of CJD patients. Thus, local public health bureau would give the long-term care facilities which had sent staffs for CJD training the priority to CJD patients care. CJD is a rare disease, with annual incidence of 0.5-1 per million persons [18]. Most CJD cases were not related to consumption of beef products. Therefore, public health authorities should not only

provide continuing education for medical staffs, but also for general public, about the difference between CJD and vCJD, and the confusion between human CJD and bovine spongiform encephalopathy (BSE).

Public health authorities should regularly renew the information about CJD on website and with a press release. As for vCJD-suspected case or other highly controversial case, public health authorities should associated with agricultural policy and food safety offices and actively convey accurate messages to the media and general public to diminish people's anxiety about CJD situation in Taiwan. Besides, public health authorities should immediately clarify any incorrect information to avoid public panic or social stigma on CJD.

In Taiwan, CJD patients and some high risk group of people are restricted from blood donation according to "The Health Standard for Blood Donation". Before donating blood, blood donors should carefully read and fill out a blood donor registration form, and then have an interviewed with the medical staffs of blood donation center, which includes the donor's past and present medical history as well as previous travel and residency history. The purpose of interview is to protect blood recipient's safety. Hence, the Department of Health is now amending related law about penalties on blood donors who have concealed conditions that are unsuitable for blood donation.

vCJD is highly related to consumption of beef products from cattle infected with BSE, while the cattle contract BSE by eating PrP<sup>Sc</sup> contaminated forage. Because this is correlated with the food chain, both

agricultural policy office and food safety office should take the responsibility of this issue. In other respects, infection control authorities and medical facilities are responsible for vCJD occurrence, disease surveillance, medical treatment and infection control. So far there is no effective medical treatment for CJD, which means this disease is incurable. As a result, management of livestock breeding, veterinary monitoring and food safety are very important for vCJD prevention. Public health authorities and agricultural policy office should collaborate closely, such as exchanging information about human CJD and cattle BSE, to protect public health.

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