Australian Government

Department of Health and Ageing

A VARIANT CREUTZFELDT-JAKOB DISEASE RESPONSE PLAN FOR AUSTRALIA

FEBRUARY 2010

Compiled by the

Office of Health Protection
Department of Health and Ageing

Disclaimer:

Information provided in the following document relates to risk of possible exposure to bovine products only. There are emerging theories that other animal products may also contain the bovine spongiform encephalopathy (BSE) agent, therefore when evidence of other transmission routes becomes available the information within this document will be reassessed.

A VARIANT CJD CLINICAL RESPONSE PLAN FOR AUSTRALIA

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A VARIANT CJD RESPONSE PLAN FOR AUSTRALIA

PURPOSE OF THIS DOCUMENT

This document has been prepared to guide the national health response should one or more cases of variant Creutzfeldt-Jakob disease (vCJD) occur in Australia. Depending on the circumstances surrounding the case of vCJD, this document may be used in conjunction with the policy documents and response plans for the Department of Agriculture, Fisheries and Forestry (DAFF, www.daff.gov.au), Food Standards Australia New Zealand (FSANZ, www.foodstandards.gov.au), Therapeutic Goods Administration (TGA, www.tga.gov.au) or the National Blood Authority (NBA) National Blood Supply Contingency Plan (Contamination Annex, www.nba.gov.au).

The most likely presentation of vCJD in Australia will be in persons who have resided in the United Kingdom (UK) or affected countries in Europe during the high-risk period of transmission (1980 to 1996).

> *"Because of the amount of travel undertaken by Australians to Britain each year, it is inevitable that one day soon Australia will discover its first case of variant Creutzfeldt-Jakob Disease (vCJD), the human form of mad cow disease." Professor Richard Smallwood, Australia's Chief Medical Officer, December 2001.*

As part of Australia's preparedness in addressing the potential public health, medicolegal, social, community, political, trade and international relations impact of vCJD, the Department of Health and Ageing has prepared this document as a guide for key stakeholders involved in disease surveillance and control. The document is based on a risk management approach for biological emergencies[1], that recognises that:

- such an event will occur infrequently;
- the evidence base for decision making may be limited and evolving; and
- community concern may be disproportionate to the level of physical risk.

Background

Origins of vCJD

vCJD is believed to be the human form of bovine spongiform encephalopathy (BSE, 'mad cow disease'). BSE was first reported as a new neurodegenerative disease of cattle in the UK in 1986. At that time, UK public health officials were of the view that BSE did not pose a risk to human health. As a precaution, enhanced surveillance was approved for human TSEs to ensure any risk to human health was detected. In March 1996, the UK reported 10 cases of a new clinicopathological variant of CJD in adolescents and young adults under the age of 40 years, believed to be associated with the consumption of beef products sourced from BSE-affected cattle.

vCJD is different to the common form of CJD seen in humans, referred to as classical CJD (cCJD) in this document, with distinct clinical-pathological features. cCJD occurs sporadically throughout the world at a rate of approximately 1 per million population. vCJD

[1] Rawling G, Jones R. Managing biological emergencies: a new approach. *Australian Journal of Emergency Management* 2001; 16(1):40-46.

is extremely rare, with approximately 200 cases reported worldwide from 1996 through to February 2008 (www.cjd.ed.ac.uk). In contrast to the age distribution of cCJD, which has a median age of onset of 50 years and over, the median age of onset in vCJD is 28 years (range 12-74). Whereas cCJD typically presents with rapidly progressive symptoms that are usually clearly neurological in nature, vCJD tends to present with behavioural or psychiatric symptoms and with a relatively slower progression. It may be difficult to determine that there is a neurological illness until some time has passed. vCJD is characterised clinically by a progressive neuropsychiatric disorder leading to ataxia, dementia and myoclonus (or chorea) and is rarely associated with the typical EEG appearance of cCJD. Neuropathology shows marked spongiform change and extensive florid plaques throughout the brain.

The tissue distribution of prions also differs between cCJD and vCJD, with infectivity mainly limited to the central nervous system in cCJD, but being more widespread in the lymphoreticular system in vCJD. To date, four cases of transmission of vCJD via blood have been demonstrated, whereas cCJD has not been shown to be transmissible via blood or fresh or frozen plasma products.

Classical CJD (cCJD) is diagnosed at a rate of approximately 1 case per million population per year throughout the world, or approximately 20-30 cases per year in Australia. cCJD includes sporadic, familial and health care associated (iatrogenic) cases of CJD. Procedures for diagnosis, reporting, surveillance, patient and family support and infection control already exist for cCJD and are outlined on the Department of Health and Ageing website (www.health.gov.au). Many of these procedures and networks would be utilised in the management of a case of vCJD.

All forms of CJD are Transmissible Spongiform Encephalopathies (TSEs), where the infectious agent is an abnormal, protease-resistant prion protein, relatively resistant to routine instrument processing and sterilisation methods currently used in health care establishments. TSEs are invariably fatal; currently there is no proven therapy for vCJD in clinically ill patients.

Diagnosis of vCJD

A sub-committee of the Communicable Disease Network Australia (CDNA) has recently reviewed the Australian case definition for vCJD (Attachment A). This case definition can be found on the CDNA website (www.health.gov.au/CDNA) and the website of the National Notifiable Diseases Surveillance System (www.health.gov.au).

There is currently no minimally invasive test available to detect vCJD infection before the onset of symptoms. There is a pre-symptomatic period during which disease transmission is presumed to be possible. Definitive diagnosis of vCJD is by neuropathological examination of brain tissue following biopsy or autopsy. However, brain biopsy is not recommended by the World Health Organization (WHO) as a routine procedure to confirm the clinical suspicion of vCJD. Although a definite diagnosis of vCJD cannot be achieved using clinical criteria alone, vCJD should be considered in the differential diagnosis of a patient presenting with the clinical criteria listed in Appendix A.

There are 3 diagnostic tests which provide support for a diagnosis of vCJD (www.cjd.ed.ac.uk, accessed 24.10.07):

- tonsil biopsy;
- the presence of protein 14-3-3 in cerebrospinal fluid (CSF); and
- imaging techniques such as magnetic resonance imaging (MRI).

In contrast to cCJD, electroencephalography (EEG) may be normal in the earlier stages of vCJD. Throughout the illness it tends to become non-specifically abnormal. The typical periodic discharges of sporadic CJD have not been seen in any case of vCJD.

As vCJD diagnosis cannot be confirmed without pathological examination, clinicians are encouraged to request permission for post mortem examination of brain tissue from patients or next-of-kin in all cases of suspect vCJD. Confirmed diagnosis is absolutely essential for public health surveillance and to guide investigations and look-back, particularly if there is a risk of nosocomial transmission (section A9.4).

Australia's strategy to remain BSE and vCJD free

To date, no cases of BSE have been detected and as far as we know, Australia remains free of indigenous animal TSEs. Australia's actions to minimise the risk of BSE and vCJD are detailed on the DAFF website (www.daff.gov.au) and on the Department of Health and Ageing website (www.health.gov.au). A ban on the importation of all stockfeed of animal origin into Australia, except fish meal and those sourced from New Zealand, has been in place since 1966. Risk assessments approved by the World Organisation for Animal Health (OIE) have shown that there is a negligible risk that BSE could be established in the Australian cattle population as a result of importation of cattle. The small number of cattle previously imported from BSE affected countries have been traced and those remaining alive have been placed under official quarantine and are prohibited from entering the human food or animal feed chains. The OIE has classified Australia as a 'negligible BSE risk' country (May 2007).

Food Standards Australia New Zealand (FSANZ, formerly the Australian New Zealand Food Authority - ANZFA), have undertaken extensive reviews of food products under their control to identify the country of origin of constituents of bovine origin.

The Australian Red Cross Blood Service has put into place, among other safeguards, donor deferral procedures to protect the safety of the Australian blood supply. The Therapeutic Goods Administration has undertaken a rigorous examination of the risk of medical and complementary medicines to ensure their safety. Blood and fresh or frozen plasma products evaluated by the TGA have been subject to risk assessments and where appropriate have implemented prion inactivation or removal steps in their manufacturing process.

Due to the relative resistance of the prion protein to inactivation and its tendency to adhere to metallic surfaces, there is a potential for transmission of vCJD via surgical instruments that have been used subsequently on patients who go on to develop vCJD. The draft infection control guideline for vCJD is under development by a sub-committee of the Communicable Disease Network Australia (CDNA) and will be updated within this document once finalised, and as required when new scientific evidence comes to light.

Government agencies are also maintaining a watching brief of international developments that may impact on Australia's regulatory initiatives, including the risk of BSE in other ruminants (e.g. sheep, goats and deer), the emergence of different strains of TSEs and the

development of new detection methods for the BSE agent in foods containing beef and beef products.

Potential risk factors for vCJD

There are several potential risk factors for acquisition of vCJD:

- residence in or travel to the UK in the relevant years (1980 to 1996) or another BSE endemic country where infected meat may have been consumed;
- consumption of imported foods containing beef and beef products from countries with endemic BSE;
- exposure to high risk surgical or dental procedures that may implicate contaminated instruments that have previously been used on a patient who had risk factors for vCJD;
- use of medications produced using high risk bovine ingredients (such as brain or spinal cord) sourced from a BSE endemic country, including medications purchased illegally or over the internet; and
- receipt of blood, fresh or frozen plasma products, tissue or organ donation, from a donor who had risk factors for vCJD.

Transmission risks

Although vCJD is believed to be contracted through the consumption of products infected with the BSE disease agent, there is evidence of person-to-person transmission of vCJD via non-leucocyte-depleted red cell concentrates in four cases in the UK. All cases involved the transfusion of blood products from apparently healthy donors who subsequently developed vCJD. cCJD has been shown to transmit via corneal transplants, dura mater grafts, human-derived pituitary hormone and contaminated surgical instruments.[2]

There is evidence that there is a more widespread distribution of the abnormal prion protein in tissues from vCJD patients than from patients with cCJD[2]. In patients with clinical vCJD, abnormal prion protein has been detected in various lymphoid tissues, including tonsils, spleen, gastrointestinal lymph tissue (appendix and rectum), lymph nodes, thymus and adrenal glands, as well as the central nervous system. Anterior and posterior eye tissues and olfactory epithelium are also considered to be infective. Abnormal prion protein has been detected in some patient tissues before the development of clinical symptoms, suggesting that patient tissues are infective during the disease incubation period prior to the onset of symptoms or diagnosis. This is clearly supported by the blood product transmissions.

The Australian National CJD Registry

The Department of Health and Ageing funds the Australian National CJD Registry (ANCJDR), based in the Department of Pathology at the University of Melbourne. The ANCJDR should be consulted as soon as vCJD is considered in the differential diagnosis. The ANCJDR can provide expert advice on sampling for diagnostic testing, clinical diagnosis, patient management, risk factor determination and infection control. The ANCJDR can assist clinicians and the state and territory health authority to arrange an autopsy or for the samples to be collected to confirm the diagnosis. The contact details for the Registry are

[2] WHO guidelines on tissue infectivity distribution in transmissible spongiform encephalopathies. World Health Organisation 2006.

listed in Appendix D.

The ANCJDR undertakes epidemiological surveillance of human TSEs, provides diagnostic services (including the 14-3-3 test and pathological examination of tonsil and brain tissue), monitors the CJD literature and provides expert technical advice. Referral of suspected cases of human TSE to the CJD Registry is either through clinical surveillance (passive and biannual active clinical surveillance principally by neurologists and neuropathologists), death certificate, National Death Index and hospital separation audits or referral by families of affected individuals and other sources. Some cases of cCJD have been ascertained retrospectively through death certificates or through the National Death Index.

The ANCJDR assists in determining the level of risk of surgical and invasive diagnostic procedures performed on patients at higher risk of all types of CJD and advises on infection control-related incidents and diagnosing all types of CJD.

vCJD Response Plan

Potential presentation scenarios for vCJD arriving in Australia

There are several possible scenarios for presentation of the first case of vCJD in Australia.

Scenario 1- Acquisition in a BSE endemic country;

Scenario 2- Acquisition in Australia but with identifiable risk factors for having acquired vCJD (e.g. imported food, medical procedures);

Scenario 3- The suspect vCJD patient who dies or is lost to follow-up before the epidemiological investigation can determine risk factors or where the patient acquired vCJD; or

Scenario 4- The patient with confirmed vCJD for whom no recognised risk factor is identified after a comprehensive epidemiological investigation.

The presentation scenario will effect the extent of the required health response, and the effect on the Australian beef industry, blood and plasma product supply and/or medicines industry. Any notification of probable or definite vCJD in Australia will require an epidemiological investigation through interviews with patients and their carers to determine the likely source of infection and potential risk factors. The extent of the response to a suspect case of vCJD in relation to these scenarios is discussed below.

How will a case of vCJD come to our attention?

Suspected cases of vCJD are likely to be identified by:

- neurologists;
- neuropathologists;
- psychiatrists;
- other clinicians;
- medical microbiologists who receive CSF with a request for 14-3-3 testing;
- patients- or family-referral; or
- retrospective audits of death certificates, the National Death Index or hospital separations.

Key stakeholders

Appendix B is a matrix describing the likely diagnostic pathway and public health response for a case of suspected vCJD in Australia. A flow chart for the response to vCJD is provided in Appendix C. The key stakeholders following a case of vCJD are listed below:

- the index case, their family or carers and their primary health care provider;
- diagnostic networks of neurologists, neuropathologists, psychiatrists;
- hospitals and care facilities in the public and private sectors;
- The Australian National CJD Registry (ANCJDR);
- the Chief Health Officer (CHO)/ Director of Public Health in the affected jurisdiction, and later all CHOs/Directors of Public Health through the Australian Health Protection Committee (AHPC);
- the Commonwealth Chief Medical Officer (CMO) and the Office of Health Protection in the Department of Health and Ageing (DoHA);
- the Australian Red Cross Blood Service (ARCBS) and the National Blood Authority (NBA);
- The Communicable Diseases Network Australia (CDNA) and the broader public health sector;
- the Chief Veterinary Officer, and subsequently state and territory departments responsible for agriculture and the Australian beef industry;
- WHO International Health Regulation (IHR) Focal Point (based in the National Incident Room of the Australian Government Department of Health and Ageing Office of Health Protection);
- regulatory bodies and other health protection agencies (including the TGA);
- other government departments including their media units;
- counselling and patient support services;
- lawyers, civil organisations regarding confidentiality and liability issues;
- vulnerable groups within the community, including individuals at increased risk of classical CJD (cCJD);
- the Australian and international media; and
- the broader Australian and international community.

Clinicians

A patient is considered to have ‘suspect’ vCJD as soon as vCJD is considered as part of the differential diagnosis. The roles and responsibilities of clinicians can be summarised as follows:

- contact the ANCJDR as soon as vCJD is considered as part of the differential diagnosis to obtain guidance on sampling and testing;
- notify the State or Territory health Department as soon as vCJD is suspected;
- collect patient history as advised by the ANCJDR to allow the State or Territory Health Department to identify potential risk factors for acquisition of vCJD and also for potential

risk factors for transmission of vCJD; and
- explain the importance of and request post mortem testing for vCJD from the patient and/ or next-of-kin.

Because vCJD could present with neuropsychiatric symptoms in a younger patient, there is a need to raise the level of awareness of vCJD among all health care providers. There is a fact sheet for health care providers on the Department of Health and Ageing website (www.health.gov.au), called *Australia's Response to the first case of vCJD – A guide for doctors and health care workers*.

Health care providers should be aware of the existence of the ANCJDR (ancjdr.path.unimelb.edu.au) and arrange for a comprehensive neurological assessment and diagnostic testing by the ANCJDR. Health care providers should be aware of the need to organise a 'care coordinator' for the patient (the patient's general practitioner or nurse) and of the availability of the CJD Support Group Network for families and carers (www.cjdsupport.org.au).

Collecting adequate patient history allows for a more accurate assessment of the risks to healthcare workers and to other patients in the healthcare setting. It is essential to collect as much information as possible about the patient's history and risks of exposure to TSEs. For example, has the patient lived in the UK, has the patient undergone any surgical procedures such as eye surgery or gastrointestinal procedures such as colonoscopy, has the patient donated blood, plasma or other tissues? Such information is critical when attempting to trace potential sources of infection both forward and back.

Definitive diagnosis aids prevention of spread of vCJD through blood, fresh and frozen plasma products, and organ donation and identification of surgical instruments used on that patient that need to be destroyed. Clinicians should discuss autopsy with the patient's next-of-kin to obtain permission for definitive testing for vCJD.

Notification process for vCJD

In 2003, CDNA added all types of CJD to the list of nationally notifiable diseases. Suspected cases of vCJD must be reported to state/territory health authorities. The state/ territory health authority notifies the National Notifiable Disease Surveillance System. The Australian National CJD Registry (ANCJDR) at the University of Melbourne should be notified of all suspected cases of vCJD. The ANCJDR provides laboratory testing for vCJD (examination of tonsil tissue and the 14-3-3 test) and advice. Definitive diagnosis of a probable clinical case of vCJD is not possible until a brain biopsy or autopsy has been performed and examined pathologically by the ANCJDR.

State and Territory Health Authorities

The public health response to probable or definite cases of vCJD is a state and territory responsibility. However, close lines of communication will be established between the CHO/Public Health authorities in the affected jurisdiction and the office of the CMO. Any diagnosis of vCJD in Australia will be of international significance, and may impact on broader sections of the Australian community, so it will be imperative to ensure a nationally consistent approach to the release of information and an effective national response.

State and territory health authorities are responsible for the surveillance and control of all communicable diseases that affect the Australian population, including vCJD. All jurisdictions have agreed to provide appropriate mortuary facilities and trained staff (or transport of bodies to and from such facilities) to conduct autopsy on suspect CJD patients.

All forms of CJD, including vCJD, have been agreed as nationally notifiable diseases and as such, any possible cases of vCJD would be reported to the Department of Health and Ageing as part of the National Notifiable Disease Surveillance Scheme, depending on individual State/Territory requirements. The State/Territory CHO, the CMO, the Office of Health Protection (including the WHO IHR Focal Point), and the ANCJDR, should also be informed. The State or Territory, in consultation with the CMO is then responsible for informing:

- National CJD Incident Panel;
- CDNA and AHPC;
- ARCBS, NBA and TGA; and
- other organisations, as appropriate.

Information provided to such groups should be handled with appropriate sensitivity and discretion, noting the potential public anxiety should information not be managed optimally. A list of key contacts is provided in Appendix D.

Agreement should be reached between the CHO/Director of Public Health and CMO on:

- The timing of notification to other CHOs/Directors of Public Health and communicable disease networks, including CDNA and the Chief Veterinary Officer;

- The level of diagnostic certainty required before the case is notified to disease control networks;

- Timing of notification to the Australian and international communities; and

- Consistency and timing of media comments.

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The CMO and Office of Health Protection should be notified of all cases being investigated as soon as vCJD is suspected.

The CMO will assist the coordination of a national response to a suspected or confirmed case of vCJD in collaboration with the CHO in the affected jurisdiction. The specific roles and responsibilities of the CMO will include:

- ministerial liaison;
- notification of key Australian Government, national and international stakeholders (including the WHO IHR focal point); and
- development of agreed and consistent media messages.

Should the media become aware of a case under investigation before the diagnostic and

epidemiological investigations are completed, it is important for key stakeholders to have agreed on a national notification and communication strategy. Unsubstantiated reports of vCJD may have serious repercussions for the affected individual, their family, carers and their community. Public confidence in health protection agencies, the beef and cattle industry and Australia's international relations could also be compromised by unsubstantiated reports of vCJD.

Extent of the response to vCJD

The State or Territory Health Department will be responsible for coordination of the response to a case of vCJD. The major considerations include not only where the infection may have been acquired, but any potential for transmission to other Australians. The State or Territory Health Department will work closely with CDNA, the Australian Government Department of Health and Ageing, the CMO and AHPC. The national response to a case of vCJD will be informed by the epidemiological investigation and may differ with different presentation scenarios. Once an initial investigation has been made, all stakeholders such as the TGA, the ARCBS, Department of Agriculture, Fisheries and Forestry (DAFF), FSANZ, the Department of Foreign Affairs and Trade (DFAT) and New Zealand (under the Joint Food Standards Treaty) would be notified through the office of the CMO, regardless of whether they subsequently play a role in case management or risk mitigation. Technical advice may be sought from the National Health and Medical Research Council (NHMRC) TSE Advisory Committee (TSEAC- formerly the Special Expert Committee on TSE- SECTSE) and the CJD Incidents Panel.

The public health response will vary on a case by case basis depending on how much clinical and epidemiological information is available. The need for post-mortem examination to confirm possible diagnosis is key and will require sensitive handling.

If the patient has consumed beef or beef products from a BSE endemic country, such as in Scenarios 1 and 2 above, this would be the most likely route of infection. It will be important to establish the safety of the Australian beef industry and the Australian food supply. Investigation of such a case may trigger re-examination of imported products thought to contain bovine material and a review of the list of countries currently included in Australia's certification process (import control). The medications that the patient has received (including those obtained illegally or over the internet) would also be investigated.

If the patient has received a blood transfusion, fresh or frozen plasma or organ or tissue donation, or has had a medical or dental procedure, a look-back will be required to establish a potential route of infection. If the patient has donated blood, plasma, organs or tissues or had a medical or dental procedure that exposed high infectivity tissues a look-back will be required to establish potential risk to other Australians. The Department of Health and Ageing has established a National CJD Incident Panel to provide advice on the management of infection control breaches related to both cCJD and vCJD. The States and the Territories have also implemented similar incident control initiatives.

Investigations

In the event that a patient with suspect vCJD has previously had a medical or dental procedure that contacted high infectivity tissues, any reusable instruments may be contaminated with the vCJD agent. The vCJD agent is highly resistant to standard instrument reprocessing. Patients who had procedures involving the same surgical instruments after the affected patient may have been exposed to vCJD. Health care establishments have a

responsibility to contact local public health authorities or the State/Territory CHO directly about any incident related to possible vCJD exposure.

The health care establishment, in consultation with the State or Territory health authority, is responsible for tracing individuals suspected of exposure to vCJD. This procedure may include seeking advice from state/ territory and National CJD incident panels. Health care establishments should develop a contingency plan that can be activated in the event an exposure is suspected. The plan should allow for tracing of potentially exposed individuals and assessment of their potential exposure to risk, and consideration of ethical and legal issues and counselling requirements, known as a 'lookback' investigation. The contingency plan should also ensure that any 'lookback' investigation is initiated only after the level of risk is fully assessed and the need for such an investigation is confirmed.

In determining the need for a 'lookback' study, consideration should be given to the benefit of informing individuals of a hypothetical risk of vCJD and their 'right to know' against the real risk of psychological injury and their right 'not to know' about the (probably remote) risk of developing a disease that has no treatment or cure. Every effort must be made by health authorities to protect the confidentiality of the individuals concerned and to avoid publicity and media involvement in the 'lookback' unless it is strictly necessary to locate those affected.

Any medical practitioner or organisation proposing to initiate a 'lookback' investigation for vCJD should first advise their local regional health authority of the circumstances and proposed 'lookback' method, and obtain appropriate advice.

Media response to a case of vCJD in Australia

One of the most important elements of a public health response will be the communication strategy to ensure that accurate information is provided to the media, the community and other domestic and international stakeholders, as premature release of inaccurate information could generate unnecessary community concern. Established networks and plans would be activated through the National Incident Room and the Primary Industries National Communication Network. It is important that the media are presented with up to date and factual information in order to minimise speculation and public concern. The Department of Health and Ageing website will have up to date information and media releases.

For media inquiries, please contact:
The Director
Media Unit
Information and Communications Division
Australian Government Department of Health and Ageing

Phone: (02) 6289 5027
Fax: (02) 6289 4044
Mobile: 0412 132 585

Conclusion

Australia continues to be BSE-free. National and international risk assessments have consistently found that Australian cattle, beef and beef products do not pose a BSE risk. The World Organisation for Animal Health (www.oie.int) has classified Australia as 'negligible BSE risk', the category of least risk.

The following principles underpin a coordinated national approach in the event of a case of vCJD diagnosed in Australia.

- Preparedness in the event of a rare biological emergency.
- Coordination of policy and operational arms at a state/territory and national level, including agreement on roles and responsibilities and who needs to know. This will require considered operational plans based on the best evidence currently available and a process of consultation between states/territories, the Australian Government, the ANCJDR and TSEAC.
- Regular communication between key policy and operational stakeholders. These lines of communication should be established now and have the ability to deal with interactions with the media and media speculation.

Appendices

Appendix A - Case definition for vCJD
Appendix B - Matrix for the investigation and response to vCJD
Appendix C – Flow chart for response to vCJD
Appendix D – Key contacts
Appendix E - List of acronyms

Appendix A - Case definition for vCJD

NOTE: a sub-committee of the Communicable Disease Network Australia (CDNA) has recently reviewed the Australian case definition for vCJD. This can be found on the CDNA website (www.health.gov.au/CDNA) and the website of the National Notifiable Diseases Surveillance System, (www.health.gov.au/internet/wcms/publishing.nsf/content/cda_surveil-nndss-dislist.htm).

Reporting

Confirmed and probable cases should be notified. (NB: a "confirmed" case is equivalent to the ANCJDR classification of "definite")

(a) Confirmed case

A confirmed case requires laboratory definitive evidence AND clinical evidence

Neuropathological confirmation of vCJD

Progressive neuropsychiatric disorder

(b) Probable case

A probable case requires clinical definitive evidence
OR
Clinical suggestive evidence AND laboratory suggestive evidence.

1. Progressive neuropsychiatric disorder AND duration of illness greater than six months AND routine investigations do not suggest an alternative diagnosis AND no history of potential iatrogenic exposure AND no evidence of a familial form of TSE

AND

2. Four of the following symptoms:
   a. Early psychiatric symptoms
   b. Persistent painful sensory symptoms
   c. Ataxia
   d. Myoclonus or chorea or dystonia
   e. Dementia

AND

3. Bilateral pulvinar high signals on magnetic resonance imaging (MRI) scans

AND

4. Electroencephalogram (EEG) which does not exhibit the typical appearance of classic CJD

1. Progressive neuropsychiatric disorder AND duration of illness greater than six months AND routine investigations do not suggest an alternative diagnosis AND no history of potential iatrogenic exposure AND no evidence of a familial form of TSE

1. A PrP^{SC} positive tonsil biopsy

Current case definition used by the WHO
http://www.who.int/zoonoses/diseases/Creutzfeldt.pdf

Appendix B - Matrix for the investigation and response to vCJD

Surveillance network activities and dependencies for suspected vCJD				
Action	***By whom***	***What***	***When***	***Critical success factors***
Suspected case of vCJD	- Neurologists - Neuropathologists - Psychiatrists - Clinicians - Self- or family-referral OR - Retrospective audits of death certificates, the National Death Index or hospital separations by the ANCJDR.	Clinical presentation or autopsy results consistent with vCJD. E.g. a younger person with rapidly progressive behavioural disorder or incoordination	Patient presents.	- Appropriate referral from primary health care systems. - Need to raise awareness among primary health care providers and possibly psychiatrists & paediatricians.
Involvement of ANCJDR	- Neurologists - Neuropathologists - Psychiatrists - Clinicians - Self- or family-referral - Audit by ANJCDR	Involvement of, or detection by, the ANCJDR.	As soon as vCJD is considered in the differential diagnosis	- Appropriate referral from primary health care system & community awareness. - vCJD considered in the differential diagnosis by clinicians. - Active notification or detection through stimulated or passive surveillance.

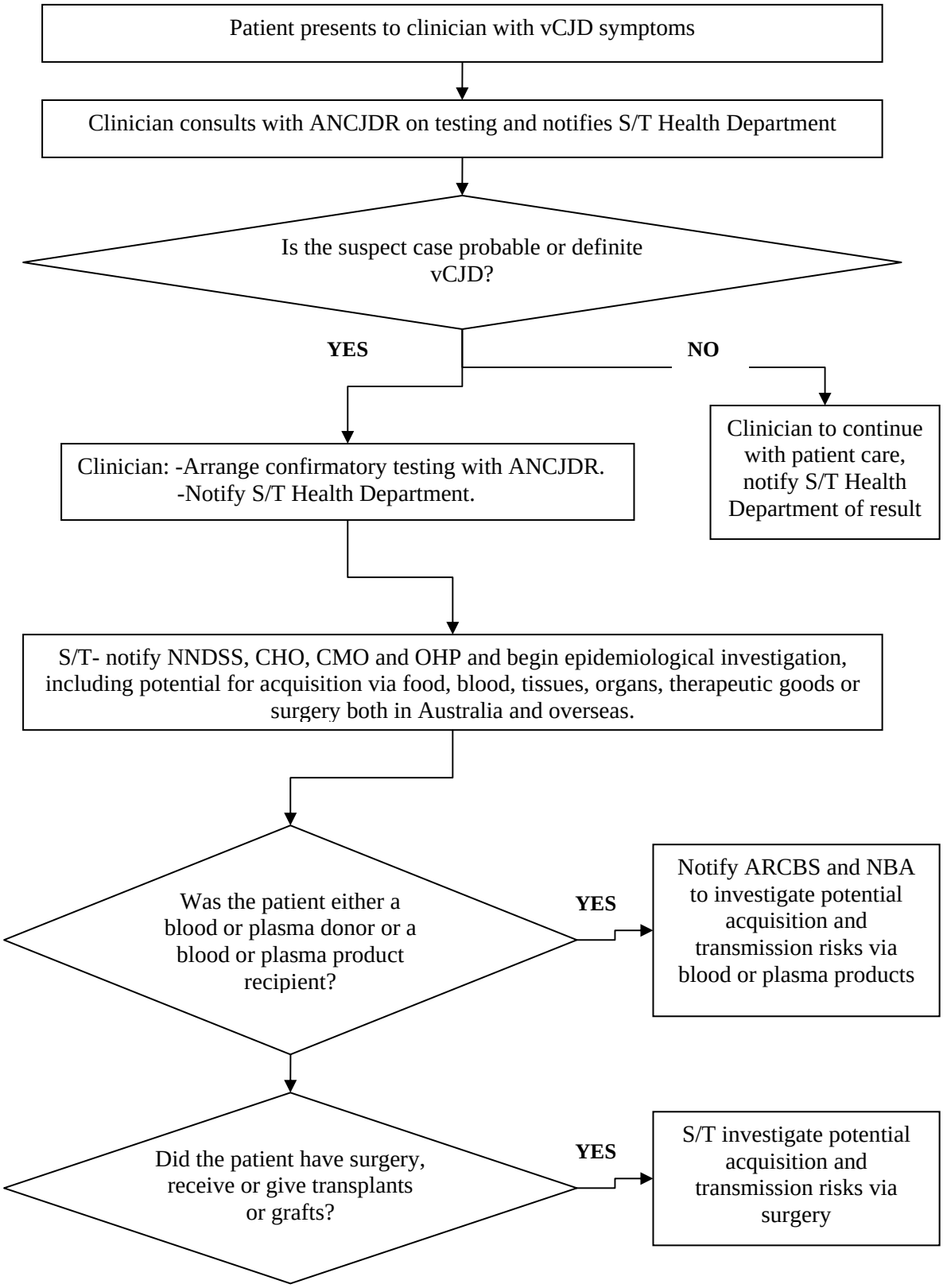
Surveillance network activities and dependencies for suspected vCJD				
Action	***By whom***	***What***	***When***	***Critical success factors***
Notification of a suspected case to the State/Territory CHO and to the CMO	Via a local public health unit OR ANCJDR or clinicians directly	- Notification of a suspected case and the expected time to a confirmed diagnosis or rejection as a case of vCJD. - Initiation of case investigation protocol. - Notify CDNA.	Should occur as soon as possible after clinician referral and ideally within 24 hours.	- Agreed referral protocols.
Initial epidemiological investigation	Jurisdictional epidemiologists in consultation with CJD Incidents Panel	Detailed case investigation to exclude other forms of CJD while awaiting definitive clinicopathological findings	Immediately after notification of a suspected case of vCJD.	Availability of credible exposure history
Confirm the diagnosis	ANCJDR in collaboration with clinicians.	- Diagnosis confirmed on the basis of clinical and pathological findings. - Confirmation in collaboration with international experts.	When ante-and/or post-mortem specimens are available.	- Availability of specimens. - Predictive value of tests in the Australian population.
Variant CJD Response Plan activated.	CMO	- Lead a national response team in accordance with perceived risk - Information link to relevant Australian Government Ministers and departments (e.g. DAFF). - Australian Government/State/Territory coordination & links to experts. - Activate communication strategy.	As soon as possible after initial epidemiological investigation identifies risk AND/OR When the case becomes a probable case of vCJD.	Strength of evidence supporting diagnosis of vCJD AND/OR Media interaction or response to public speculation.

Surveillance network activities and dependencies for suspected vCJD				
Action	***By whom***	***What***	***When***	***Critical success factors***
Detailed epidemiological investigation undertaken.	Team involved will depend on the key findings of the initial epidemiological investigation. Team members may include: - State/Territory CHO and communicable disease epidemiologists - Hospital infection control staff - ANCJDR - National & State/Territory CJD Incidents Panels - CDNA - Department of Health & Ageing - TGA - DAFF - FSANZ - ARCBS/Blood & Organ Donation Agencies - NBA - TSEAC	- Active (patient/ family member recall) or passive (community announcement) lookback investigations in the case of infection control breach(es) within health care settings. - May involve a review of risk management procedures already in place to protect human and animal health, including review of restrictions on imported beef products. - May require notification to NZ under treaty obligations and cross-Tasman approach	As soon as possible after initial epidemiological investigation identifies risk AND/OR When the case becomes a probable case of vCJD.	Availability of epidemiological information that can inform the nature and size of the required response, and the agencies that should be involved.

Surveillance network activities and dependencies for suspected vCJD				
Action	***By whom***	***What***	***When***	***Critical success factors***
Risk communication	CMO in collaboration with jurisdictional CHO and other relevant agencies depending on the facts of the case	- Detailed communication strategy developed in collaboration with Department of Health & Ageing media unit and DAFF. - Notification to the World Health Organization focal point under the International Health Regulations (significant public health event). - Notification to other CJD Registries and the international community as appropriate.	- Management of media interactions at any stage of the investigation. - Controlled media release as soon as a probable case of vCJD is close to final confirmation. - Notify the World Health Organization when diagnosis confirmed if there are no media/network leaks in the mean time.	- Timing and nature of media releases will depend on the scenario encountered and whether there is an ongoing risk to the Australian community. - Timing of international notification dependent on confidentiality being maintained by national & international partners involved in diagnosis & case investigation.
Patient support and family services.	State/Territory and Australian Government. CJD Support Group Network	Examination of the availability, efficiency, effectiveness and acceptability of support services and need for financial relief by family/carers/hospitals etc.	As soon as diagnosis is suspected or confirmed	State/Territory commitment to financial relief.
Independent epidemiological review	An expert group of independent investigators.	Re-interview the family to confirm the patient's history and re-examine all the clinicopathological and epidemiological evidence.	An independent review is recommended in Scenario 4 ie. the patient with confirmed vCJD in whom no recognised risk factor is identified after a comprehensive epidemiological investigation.	Availability of panel AND Cooperation by the family/carers of the index case.

Surveillance network activities and dependencies for suspected vCJD				
Action	***By whom***	***What***	***When***	***Critical success factors***
Debriefing and iterative review of the vCJD response plan.	Teams at local, jurisdictional and national levels.	- Identify strengths and weaknesses of response plans, including coordination. - Economic evaluation. - Applied research arising out the investigation as appropriate.	As required.	Agency/partner participation.

Appendix C – Flow chart for response to vCJD



Appendix D– Key Contacts

Australian National CJD Registry **(ancjdr.path.unimelb.edu.au)**
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The University of Melbourne
Parkville VIC 3010
Ph: (03) 8344 1949 Fax: (03) 9349 5105
Email: ANCJD-REG@unimelb.edu.au

WHO IHR Focal Point
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Office of the Chief Medical Officer
Office of Health Protection
Department of Health and Ageing (www.health.gov.au)
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health.ops@health.gov.au

For media inquiries, please contact:
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Department of Health and Ageing
Ph: (02) 6289 5027 Fax: (02) 6289 4044
Mobile: 0412 132 585

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(www.health.act.gov.au)
GPO Box 825
Canberra City ACT 2601
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NSW Health Department
(www.health.nsw.gov.au)
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North Sydney NSW 2059
Ph: (02) 9391-9000
Email:
NSWhealth@dog.health.nsw.gov.au

QLD Health Department
(www.health.qld.gov.au)
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Brisbane QLD 4000
Ph: (07) 3234-0111

NT Health Department
(www.health.nt.gov.au)
PO Box 40596
Casuarina NT 0811
Ph: (08) 8999-2400

WA Health Department (CDCD)
(www.health.wa.gov.au)
PO Box 8172
Perth Business Centre
Perth WA 6849
Ph: (08) 9388 4852
Email: episurv-cdcdwa@health.wa.gov.au

SA Health Department (CDCB)
(www.health.sa.gov.au)
PO Box 6
Rundle Mall
Adelaide SA 5000
Ph: (08) 8226-7177

VIC Health Department
(www.health.vic.gov.au)
50 Lonsdale Street
Melbourne VIC 3000
Ph: 1300 650 172

TAS Health Department
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GLENHAVEN NSW 2156
National Toll Free 1800 052466

s.solvyns@cjdsupport.org.au
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National Blood Authority
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nationalbloodauthority@nba.gov.au
6211 8325 or 0413 486 433
Australian Red Cross Blood Service
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MELBOURNE VIC 3001
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M: 0420 961 489
E: news@arcbs.redcross.org.au
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Appendix D - List of acronyms

AHPC	Australian Health Protection Committee
ANCJDR	Australian National CJD Registry
ARCBS	Australian Red Cross Blood Service
BSE	Bovine spongiform encephalopathy
cCJD	Classical CJD.
CDNA	Communicable Diseases Network Australia
CHO	Chief Health Officer
CJD	Creutzfeldt-Jakob disease
CMO	Commonwealth Chief Medical Officer
CSF	Cerebrospinal fluid
DAFF	Department of Agriculture, Fisheries and Forestry
DFAT	Department of Foreign Affairs and Trade
DoHA	Department of Health and Ageing
EC	European Commission
EEG	Electroencephalogram
FSANZ	Food Standards Australia New Zealand
IHR	International Health Regulations
NBA	National Blood Authority
NHMRC	National Health and Medical Research Council
OIE	World Organisation for Animal Health
SECTSE	Special Expert Committee on Transmissible Spongiform Encephalopathies (now TSEAC)
TGA	Therapeutic Goods Administration
TSE	Transmissible spongiform encephalopathy
TSEAC	Transmissible Spongiform Encephalopathy Advisory Committee (formerly SECTSE)
UK	United Kingdom
vCJD	Variant CJD