



INFORMATION FOR CLINICIANS REGARDING POTENTIAL POST MORTEM EXAMINATION OF CASES SUSPECTED OF CREUTZFELDT-JAKOB DISEASE IN NSW

The Royal Prince Alfred Hospital (RPAH) Department of Neuropathology acts as the NSW referral centre for all suspected cases of Creutzfeldt-Jakob disease (CJD). The following guide has been prepared to assist clinicians in the handling of these cases.

1. For clinically suspected CJD cases it is suggested that a pre-mortem CSF 14-3-3 test be performed. The specimen should be sent to:

Australian National CJD Registry (ANCJDR)

Dr Christiane Stehmann

The Florey Institute, University of Melbourne

Parkville VIC 3052

Phone: (03) 8344 1949 or (03) 8344 6530, Fax: (03) 9349 5105

2. When death is impending, to ensure the case is suitable for the CJD autopsy protocol, clinicians must telephone the RPAH Department of Neuropathology at (02) 9351 0741 or (02) 9351 0943 (if not answered, follow the instructions on the message). Appropriate consent and other documentation will be provided which must be completed (see below). Please indicate if there is any suspicion of New Variant CJD.
3. After death, the clinician needs to complete the normal 'Medical Certificate of Cause of Death' form as well as a Cremation Certificate.
4. Relatives should be advised that the body cannot be viewed after autopsy as there is no restitution of the body due to the risk of needle-stick injury to the technician and for infection control reasons. As a result, a viewing prior to the transfer of the deceased, either on the hospital ward or at the funeral home, is recommended. If the family wish to dress the deceased in a particular outfit, this should occur before the transfer for the autopsy and no valuables should be left on the body as these cannot be retrieved after the autopsy.

5. **A summary of the clinical history**, the **'Medical Certificate of Cause of Death'** and **Cremation certificate** (or photocopies) must accompany the body to Forensic Medicine Sydney based at Lidcombe. A Life Extinct certificate is not acceptable. Please include details of the name(s) of clinician(s) and their contact telephone and fax numbers, as well as details of the family's nominated funeral director.

6. A **'Non-Coronial Post Mortem Consent & Authorisation for ?CJD Brain Examination'** form for permission to carry out an autopsy, with retention of the brain, must be completed by both the **clinician** and the most **senior available next-of-kin** as per a normal hospital autopsy. It is preferable that the next-of-kin also tick the box indicating 'for medical, scientific & teaching'. If the patient died in a hospital, the consent form must also be signed by that hospital's **'Designated Officer'**.

Next-of-kin hierarchy is limited to (in order): (a) spouse, (b) adult offspring, (c) parent then (d) sibling. There must be no objection from someone of equal or higher ranking. There is no option to delegate to an individual of a lower ranking, unless the higher ranked individual has no capacity to give consent – e.g. has dementia, is under 18 years of age, or is un-contactable.

Please note that Guardianship and Power of Attorney arrangements cease upon death and do not affect the requirement for senior available next-of-kin consent.

7. Summary of documents that must accompany the case and be sent to the RPAH Department of Neuropathology by fax at (02) 9114 4020 or emailed to neuropathology.lab@sydney.edu.au

- Medical Certificate of Cause of Death;
- **'Non-Coronial Post Mortem Consent & Authorisation for ?CJD Brain Examination'** signed by the **senior available next-of-kin** and local **Designated Officer** (if patient died in a hospital) including details of the nominated **funeral director**;
- Cremation certificate, and
- Clinical summary or copy of relevant medical records.

8. After all paperwork is received by the RPAH Department of Neuropathology, they will liaise with Forensic Medicine Sydney based at Lidcombe to obtain local designated officer signoff and permission to transfer the body. They will then organise body transfer through Statewide Transfers.

9. **Autopsy procedures.** Mortuary staff will wear isolation suits with independent air-supply to prevent direct contact with the brain tissue. The scalp is cut over the vertex and is retracted from the skull. A bone saw is used to remove the skull cap, and the brain separated from the body by cutting through the dura, blood vessels, nerves and spinal cord. The brain is removed. The (un-reconstructed) body is sealed into a body-bag labelled “infectious”. The body is returned to cold storage to await pick-up by the family-appointed funeral directors.

10. **Brain preparation procedures.** After removal of the brain, a portion of the frontal lobe is removed and placed in a (labelled) plastic bag and container in the freezer room. The rest of the brain is fixed in 10% neutral buffered formalin. The formalin-fixed and frozen brains are sent to the RPAH Department of Neuropathology for neuropathological diagnosis. For confirmed case of CJD, the frozen specimen is sent to the ANCJDR in Melbourne for glycotyping, Western blotting and, if familial CJD is suspected, for genotyping of the *PRNP* gene.

11. **Neuropathological diagnosis.** After at least three weeks in formalin, the neuropathological examination takes place, where tissue blocks are taken for staining and microscopic analysis. If CJD is confirmed, a neuropathology report is generated which generally takes approximately 6-8 months to complete. If CJD is excluded, additional testing will subsequently take place to attempt to identify any neuropathology that may account for the symptoms. This extends the time period of completion of the neuropathology report.

Upon completion of the final neuropathological report, the report will be faxed to the referring physician / nominated general practitioner. For this reason, it is essential that the referring physician supply contact details and fax numbers with the accompanying documentation.

Confirmed cases of CJD are also notified by the RPAH Department of Neuropathology to the NSW Department of Health, where the name, gender, date-of-birth, date-of-death and address of the deceased are supplied for epidemiological studies.