

A large graphic consisting of a thick yellow outer ring and a light blue inner circle, which serves as a background for the main title text.

**Fact Sheet:**  
**Autopsy in patients**  
**with suspected CJD**  
answers to some  
commonly asked questions



## About the CJD Support Network

The CJD Support Network is the leading care and support charity for all forms of CJD. The CJD Support Network:

- Provides practical and emotional support to individuals, families and professionals concerned with all forms of CJD
- Provides emotional support to people who have been told that they are at a 'higher risk' of CJD through blood or surgical instruments
- Links families with similar experiences of all forms of CJD
- Offers financial support for families in need
- Provides accurate, unbiased and up-to-date information and advice about all forms of CJD
- Provides a national helpline on all forms of CJD
- Promotes research and the dissemination of research findings
- Promotes good quality care for people with all forms of CJD
- Encourages the development of a public policy response for all forms of CJD
- Provides support, education and training to professionals concerned with CJD

For more information about the activities of the CJD Support Network, contact:

Admin and general enquiries - [admin@cjdsupport.co.uk](mailto:admin@cjdsupport.co.uk) or +44 (0)7494 211476

Support - [support@cjdsupport.co.uk](mailto:support@cjdsupport.co.uk) or 0800 774 7317

Website - [www.cjdsupport.co.uk](http://www.cjdsupport.co.uk)

Post - PO Box 3936, Chester, CH1 9NG

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### **Fact sheet: Autopsy in patients with suspected CJD**

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## ***Why are post mortem examinations requested in patients with CJD?***

In most cases, a confident diagnosis of CJD can be made in life, on the basis of the clinical picture, the exclusion of other possible diagnoses and the results of some helpful diagnostic tests. There can be varying degrees of confidence, depending on the individual circumstances. In many cases, a highly probable diagnosis can be made, whereas, in others, it may be regarded as possible. At present, the only way to make an absolutely definite diagnosis is via pathological examination of brain tissue. This can be done in life, via a brain biopsy, but this is done only rarely, in certain specific situations, and, usually, it is done at a post-mortem (autopsy) examination.

There are four main reasons for performing an autopsy in relation to CJD:

- Significant uncertainty about the diagnosis in life (because of unusual clinical features and/or unhelpful diagnostic test results). This may be uncertainty about the diagnosis of CJD in general, or about the type of CJD (e.g. sporadic or variant).
- A wish, by the relatives and/or clinicians to be absolutely certain about the diagnosis, even if it was made with confidence in life.
- An autopsy is directed by a Coroner or, in Scotland, a Procurator Fiscal.
- To contribute to research into CJD.

## ***Is it necessary to examine the whole body at autopsy, or can it be limited to the brain alone?***

An autopsy can be limited to the brain alone, to include only certain other body tissues, or to look at the whole body. The type of autopsy that is diagnostically ideal will depend on the individual clinical situation, but the wishes of the family are taken into account in the consent process.

## ***Is it necessary to remove the entire brain during post mortem examination?***

It is internationally accepted that the best way of confirming a diagnosis of CJD is to retain the brain at the time of post mortem for full examination in the laboratory. During the post mortem, the brain is carefully removed (using an approach from the back of the head) and most of the tissue is fixed in formaldehyde. Some of the tissue may be frozen in order to perform biochemical studies to identify the abnormal prion protein in the brain.

However, a diagnosis of CJD can be confirmed by examining a small portion of tissue from one part of the brain (as is done in a biopsy), but there is a possibility that disease could be missed, and, if brain sampling is done at autopsy, it would be usual to take small samples from several different brain areas.

The way in which the brain is examined at autopsy, needs to reflect the specific clinical circumstances and the reasons for undertaking the autopsy, however, the wishes of the family are taken into account in the consent process.

## ***How can I give consent for a post mortem examination?***

Consent is usually given through the clinician looking after the patient, but a specialist unit (such as the National Prion Clinic or the National CJD Research and Surveillance Unit) may be involved. A new consent

form for post mortem examinations was established in 2000 on the basis of guidelines from the Royal College of Pathologists. Using this form, relatives are asked for permission to perform an autopsy on a patient with suspected CJD and then asked for specific permission to retain the brain for diagnosis. Additional permission is also requested to use the brain tissue for research and teaching purposes so that more can be found out about these diseases. Relatives will be asked if they would like to have the brain tissue returned for subsequent burial or cremation. If genetic testing is to be performed on autopsy tissue, this requires specific consent.

### ***Why are some post mortems in patients with CJD performed under the instructions of the coroner (Procurator Fiscal in Scotland)?***

The commonest forms of CJD (sporadic and familial CJD and GSS) are naturally occurring diseases which are found in countries across the entire world. As such, there is no general legal requirement for autopsy. However, variant CJD, (related to BSE) and iatrogenic CJD (for example, in people who received certain types of human growth hormone), are considered by many Coroners/Procurator Fiscals not to be naturally occurring diseases and, as such, they may express an interest in the case and instruct an autopsy examination. Coroners/Procurator Fiscals sometimes also take an interest in cases of presumed sporadic CJD so that the diagnosis can be confirmed. It is up to the individual Coroner/Procurator Fiscal to decide on the cases in which they will become involved and for which an inquest will be held. If a Coroner/Procurator Fiscal directs that an autopsy is to be performed, it is a legal direction and no family consent is required

### ***Will agreeing to a post mortem hold up the funeral or delay obtaining a death certificate?***

Only certain large hospitals with regional neurology centres can perform post mortems on patients with suspected CJD as many hospitals either do not have adequate facilities or will not agree to carry out the procedure. Often the body must be transported to the nearest suitable mortuary. Usually, this can be arranged quickly, and most post mortems on patients with suspected CJD are performed within two days of death. Funeral arrangements are therefore not usually delayed and if there are any special circumstances, for example a funeral service in another part of the country, these can be accommodated. If there are extra costs relating to a requested post mortem, including transport of the body to a suitable mortuary, ways of meeting these need to be discussed on an individual basis, with local clinical services, and any involved research institutes.

A death certificate with a provisional diagnosis of CJD can be issued immediately – before the results of the post mortem examination are known. The doctor completing the death certificate will indicate that a post mortem is to be performed and that the results of this investigation will be available in due course. Details of the final autopsy findings are sent to the registrar of deaths and the post mortem findings will be incorporated into the final certificate.

### ***What happens during an autopsy on a patient with CJD?***

Post mortem examinations on patients with suspected CJD are performed in a similar way to general autopsies. However, the pathologists and mortuary technicians involved need to take special precautions to prevent any chance of accidental transmission of the disease to them during the procedure. As a minimum requirement, the brain is carefully removed from the head and retained for further examination

and diagnosis. In many cases the other body organs are also examined and tissue samples are taken in order to establish the immediate cause of death (for example, bronchopneumonia) and so that additional studies on the disease can be carried out.

### ***Can I request that a limited post mortem is performed on my relative?***

It is entirely possible for permission for a limited post mortem to be performed in cases of suspected CJD. Under these circumstances, the autopsy is confined to the examination of the brain, or samples taken from the brain, as detailed above. After the diagnosis is confirmed, the brain tissue can be returned to the undertaker for burial or cremation if required.

### ***Are there any risks to relatives in viewing the body of a patient who has died with CJD?***

CJD is a transmissible disease but only under very specific circumstances and it is not highly infectious. Precautions are required for the transport of bodies through the use of a body bag to protect against accidental seepage of body fluids following death. However, the body bags can be opened to allow relatives to view the body, even after an autopsy has been performed, without risk to those relatives. The Department of Health has recently issued guidance to undertakers on this important matter. There is no need for special burial requirements for patients with suspected CJD.

### ***Why does the autopsy diagnosis take so long?***

In order to confirm the diagnosis of CJD the brain has to be examined carefully. Histological slides of brain tissue are prepared for examination under a microscope. The diagnosis of CJD often requires special staining techniques and biochemical studies on frozen brain tissue. This usually takes around eight weeks after the postmortem has been performed, and in difficult or complicated cases the timescale will be longer.

### ***What are the benefits of agreeing to a postmortem examination on a relative who has died with suspected CJD?***

- The most important benefit is that an absolutely definite diagnosis, of CJD, and its type, will be obtained; the family will know for certain the true nature of the disease which has affected their relative. However, as mentioned above, with modern investigations, a highly confident diagnosis of CJD can often be made in life and both clinicians and families may be content with this high degree of certainty. Attitudes to autopsy vary considerably and families who, for personal reasons, do not wish for an autopsy should not feel that one is necessary in cases where the diagnosis is highly probable. If there was significant diagnostic uncertainty in life, then families and clinicians may value having a definitive autopsy diagnosis.
- There are important public health concerns, and policies, connected with CJD, especially vCJD. Autopsy examination of cases where there is diagnostic uncertainty is obviously important for having accurate information as to the numbers of CJD cases and whether they are changing over time.
- As mentioned above, autopsy allows researchers to study these diseases and to obtain tissues vital for certain aspects of research. Confirmation of diagnosis is also important in establishing the accuracy of new diagnostic tests.



## Further information and contacts

Further information about CJD may be found on the CJD Support Network website at [www.cjdsupport.net](http://www.cjdsupport.net) our fact sheets are also available by post on request to the Network.

Support and information may be obtained from the organisations below:

### CJD Support Network

Admin and general enquiries - [admin@cjdsupport.co.uk](mailto:admin@cjdsupport.co.uk)  
Support - [support@cjdsupport.co.uk](mailto:support@cjdsupport.co.uk) or 0800 774 7317  
Website - [www.cjdsupport.co.uk](http://www.cjdsupport.co.uk)  
Post - PO Box 3936, Chester, CH1 9NG  
Registered charity no. 1097173

### National CJD Research and Surveillance Unit

Western General Hospital, Crewe Road, Edinburgh EH4 2XU  
Website - [www.cjd.ed.ac.uk](http://www.cjd.ed.ac.uk)  
0131 537 1980 telephone number for general enquiries and  
[loth.securecjd@nhslothian.scot.nhs.uk](mailto:loth.securecjd@nhslothian.scot.nhs.uk) for Care Team enquiries/patient families

### National Prion Clinic

National Prion Clinic, Institute of Prion Diseases, Courtauld Building, 33 Cleveland Street, London, W1W 7FF.  
Helpline for National Prion Clinic - 020 7679 5142 / 020 7679 5036  
[uclh.prion.help@nhs.net](mailto:uclh.prion.help@nhs.net)