CJD SUPPORT NETWORK

INFORMATION SHEET 6 JUNE 2001

The autopsy in patients with suspected CJD

Answers to some commonly asked questions

Why are post mortem examinations requested in patients with CJD?

Although a diagnosis of probable or possible CJD can be made during the clinical illness in individual patients, the only way to confirm the diagnosis is by performing a post mortem examination of the brain. This information is important in establishing the incidence of CJD in the UK. Performing a post mortem examination also allows the opportunity to study these devastating diseases – how the brain is damaged, the changes occurring in other parts of the body and scientific studies on the nature of the agent causing these diseases.

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

A diagnosis of CJD can sometimes be confirmed by examining only a small portion of brain tissue –– this can also be done in life by performing a brain biopsy. However, this can give a number of potentially misleading results. 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.

How can I give consent for a post mortem examination?

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.

Why are some post mortems on patients of 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. However, variant CJD and iatrogenic CJD (for example, in people who received human growth hormone) are considered by many coroners not to be naturally occurring diseases. In such instances, a coroner may express an interest in the case and instruct an autopsy examination. Some coroners will also take an interest in cases of presumed sporadic CJD so that the diagnosis can be confirmed. It is up to the individual coroner to decide on the cases in which they will become involved and for which an inquest will be held.

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

Most large hospitals with regional neurology centres can perform post mortems on patients with suspected CJD, but smaller hospitals will not have adequate facilities and the body will be transported to the nearest suitable mortuary. 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. All extra costs relating to a post mortem, including transport of the body to a suitable mortuary, will be paid for by the CJD Surveillance Unit.

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, which is removed and retained for diagnosis in the usual way. The remainder of the body is not examined, and 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 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. 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 is there often a delay in confirming the diagnosis?

In order to confirm the diagnosis of CJD the brain has to be examined carefully. Histological slides of brain tissue to be examined under a microscope are prepared. This usually takes around eight weeks after the post mortem has been performed, and in difficult or complicated cases the timescale will be increased. The diagnosis of CJD often requires special staining techniques and biochemical studies on frozen brain tissue. Many attempts have been made to speed up these processes. However, in some cases this has been unsuccessful and the preparation of the microscopic slides has been ruined, further delaying the diagnosis.

What are the benefits of agreeing to a post mortem examination on a relative who has died with suspected CJD?

The most important benefit is that an accurate diagnosis will be obtained and that the family will know the true nature of the disease which has affected their relative.

In many suspected cases of CJD the diagnosis will be confirmed, but in some other patients an alternative diagnosis will be made – for example, Alzheimer's disease or stroke. Examination of the brain is the only way to make a definite diagnosis of variant CJD. Also, in this condition, it has been particularly important to examine other tissues in the body (for example, the tonsils and lymph nodes) to detect the abnormal prion protein. Permission to conduct research on autopsy tissues has provided valuable information on the relationship between variant CJD and BSE, and will help us and others develop new methods of diagnosis and, we hope, a treatment for these fatal disorders.

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