

Types of CJD

There are a number of different types of CJD.

1. **Sporadic (or classical) CJD** was first described in the 1920s. It is rare, but accounts for most cases of CJD (about 85%) and its cause is not known. It occurs mostly in people aged 50 to 75.
2. **Genetic (or familial) CJD** Some cases of CJD are linked to a genetic abnormality and are known as familial.
3. **Iatrogenic CJD** is caused by accidental transmission through medical or surgical procedures. Iatrogenic CJD has occurred (a) in some people treated with growth hormone from human pituitary glands, (b) in recipients of grafts of dura mater (a membrane lining the skull) and (c) corneal graft recipients (uses tissue from the front of the eye). A few cases have also been documented in the past involving neurosurgical devices previously used on someone with the disease.
4. **Variante CJD (vCJD)** is caused by exposure to Bovine Spongiform Encephalopathy (BSE) and typically occurs in younger people, many of whom are thought to have eaten contaminated beef in the 1980s. At post-mortem examination the brain of vCJD patients has distinctive appearance which distinguishes it from other types of CJD.

How many people have CJD?

Very few. Only about 1 case of sporadic CJD occurs per year per million people. Genetic CJD and iatrogenic CJD are also very rare. Based on figures produced by the National CJD Research and Surveillance Unit in Edinburgh, we believe that 177 patients have been diagnosed in the UK with either definite or probable vCJD since 1990, all of whom have died.

How is it diagnosed and is there a cure?

The diagnosis is usually made on clinical signs, symptoms and specialist tests, but confirmation of the diagnosis can be made only by microscopic examination of brain tissue, usually obtained at post-mortem. Currently, there are no treatments that have been shown to slow, halt or reverse the disease.

If you wish to discuss any concerns, contact details are:
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INFORMATION FOR PATIENTS
ABOUT
CREUTZFELDT-JAKOB
DISEASE (CJD) AND MEDICAL
AND SURGICAL PROCEDURES

If you require this information in an alternative language or format (such as Braille, audiotape or large print), please contact the Microbiology Department on 01423 555663

Introduction

This information leaflet has been produced to answer some commonly asked questions about CJD (Creutzfeldt-Jakob disease), and to explain why we may ask patients about CJD before some medical and surgical procedures, such as operations or endoscopy.

What is CJD?

CJD is sometimes known as the human form of BSE (bovine spongiform encephalopathy) or mad cow disease, and is a severe and fatal brain disease. CJD is not thought to be caused by bacteria or viruses, but by abnormal proteins called prions.

Why are we checking if you are in an “at risk group”?

Most operations do not involve organs or tissues which are affected by CJD. As a precaution however, the Department of Health has recommended we check to see if anyone has ever contacted you to say that you have been put at risk of catching CJD, the human form of mad cow disease. This might have been the Blood Transfusion Service, a Public Health Doctor or your own General Practitioner. If you have been contacted, or if you haven't been but are having a procedure involving tissues which may be affected by CJD, we will need to ask some more detailed questions. All this is to ensure that the equipment used is either disposable, protected by a disposable sheath, or cleaned,

disinfected and sterilized by the appropriate procedures recommended by current Department of Health guidance. This is because the agents (prions) causing CJD are resistant to normal decontamination measures for instruments used in medical and surgical procedures.

Who is at risk?

Certain individuals are considered as having a slightly higher risk than normal of developing CJD

- Those with a definite family history
- Patients who have had a previous corneal graft, dura mater graft or injections of human growth hormone
- Patients who have had brain or spinal cord surgery before 1992
- Patients receiving certain plasma products in the past (these patients have been identified and notified)

Anyone with one or more of the above risk factors is considered to be in the “at risk group”.

How do we check?

We ask all patients having operations or endoscopy:

Has anyone ever contacted you to say that you have been put at risk of catching CJD, the human form of mad cow disease?

If you answer yes to this question, or if you are having a procedure involving a tissue which may be affected by CJD, we ask the following questions:

1. Do you or anyone in your family have CJD, the human form of mad cow disease, or anything similar?
2. Did you ever receive growth hormone or other hormone treatment, even as a child (not insulin in diabetes)?
3. Have you had brain surgery or spine (back bone) surgery; if so was it since 1992?
4. Since 1980 have you received transfusions of more than 50 units of blood or blood products, or transfusions on more than 20 occasions?

What help will I get if I am in an “at risk group”?

We will get a specialist to contact your General Practitioner to discuss what it means to be in an “at risk group.” This is because you may have many questions which cannot be answered in this brief leaflet. However in most cases it only means that you need to inform a doctor or dentist before you have a medical, surgical or dental procedure performed.

Further useful information on CJD can be found at the website of the National Creutzfeldt-Jakob Disease Surveillance Unit, based in Edinburgh:

www.cjd.ed.ac.uk

This website includes links to care and support groups