Emerging concerns related to CJD

AUTHOR Gillian Turner, MBE, MA, is national CJD co-ordinator, CJD Network.


Until recently, it had been hoped that the incidence of Creutzfeldt-Jakob disease was declining. However, new findings suggest that people previously thought not to be at risk may simply be incubating the disease for longer.

The disease is always fatal, and places heavy demands on families and social and health care professionals, as care needs vary and can change rapidly.

News that a second death from Creutzfeldt-Jakob disease (CJD) has been linked to blood transfusion has led to concern that the disease could affect far more people than previously thought (Peden et al, 2004). The first person to die from CJD after receiving a blood transfusion from a donor who subsequently developed variant CJD – the form acquired by eating beef infected with bovine spongiform encephalopathy (BSE) in the 1980s – was revealed last year. This led to radical changes in blood collection procedures and a ban on blood donations from people who have received a transfusion since 1980.

The second death was not caused by CJD. However, a postmortem examination found the abnormal prion protein responsible for the disease in the person’s spleen and lymph nodes, even though he had shown no symptoms of the disease. Scientists at the CJD Surveillance Unit believe it is very likely that the infection was acquired from a blood transfusion. The patient also had a different genotype (MV) than all other previous vCJD patients, which suggests the disease can affect 90 per cent of the population.

This death in a different genotype has increased concern about vCJD because it suggests that some people may incubate the disease for longer periods than those who have already become symptomatic or who have died. This means the period over which new cases may occur could be decades rather than years.

The genetics of vCJD susceptibility

To date 142 people are believed to have died from vCJD. All had shared the same versions of a gene (MM) on chromosome 20, known as the prion gene. This controls the behaviour of healthy and abnormal prion proteins found in the body. The gene can encode for two different amino acids – methion or valine, and everyone has two copies, so individuals may be MM, VV or MV genotype.

All those who have died of vCJD belonged to the MM genotype, which they share with 37 per cent of the population. The person found to be incubating the disease had the MV genotype, which is carried by 50 per cent of the population.

Before this case it was thought that only people with the MM genotype were susceptible, but now there are fears that those of MV genotype are also susceptible but incubate the disease for longer. If so, many more people may have contracted the disease from contaminated beef and in turn more cases may be acquired via contaminated blood or surgical instruments.

What is CJD?

Creutzfeldt-Jakob disease is one of a rare group of fatal diseases, which destroy neural tissue by the accumulation of abnormal prion proteins. In healthy individuals, these exist as cell-surface proteins in many cells, especially in the brain. They are soluble and can be degraded by the body, but their function is unknown. The abnormal prion protein responsible for vCJD is insoluble and resistant to breakdown, so it accumulates in neural cells, disrupting function and leading to cell death.

The disease is degenerative and is characterised by progressive damage to brain structures, causing microscopic spongiform change. Its incubation period may vary between four and 30 years. The causative agent is extremely resistant to conventional sterilisation and disinfection technologies.

CJD is one of a family of diseases known as the transmissible spongiform encephalopathies, a number of which also affect animals such as sheep and mink; five types are found in humans (Table 1). CJD has probably always been present in small numbers, but has been brought to public attention by links with BSE.

A devastating and complex disease, CJD is only partially understood but places great demands on carers, relatives and professionals. It causes dementia and a range of neurological, physical and psychological symptoms. As there is no test for CJD in living patients, it can only be confirmed at postmortem examination.

Development of services

Most carers of early cases of CJD faced the rapid deterioration and loss of their loved one with little help, as by the time support was organised it was often too late. In 1998 a CJD Support Network survey of members found there was little awareness of CJD among professionals, while health and social service response was mixed. As a result of the survey, the CJD Support Network (1998), in conjunction with directors of social services, developed good practice guidelines for social workers.

One of the initial challenges for staff in health and social services was the lack of expertise, as CJD was so rare. In many cases social workers were the first in their area to be involved with someone with CJD.

The challenge for service providers stems from the fact that CJD requires them to act and work differently. Different strains require different approaches and patients’ needs can change suddenly and dramatically.
TABLE 1. CAUSES AND INCIDENCE OF THE DIFFERENT TYPES OF CREUTZFELDT-JAKOB DISEASE

<table>
<thead>
<tr>
<th>TYPE</th>
<th>CAUSE</th>
<th>OTHER COMMENTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sporadic classical</td>
<td>Naturally occurring</td>
<td>Responsible for 85 per cent of CJD cases. Incidence is somatic mutation in PrP gene one per million people over the age of 50 mean of about four months from onset to death.</td>
</tr>
<tr>
<td>Inherited prion disease</td>
<td>Germ line PrP mutation – runs in families</td>
<td>An extremely rare disease with no significant variation in worldwide incidence. People with the disease are likely to become ill in their 40s and 50s although it has been known to start in people in their 30s or as late as their 70s. All children of those carrying the mutated gene have a 50 per cent chance of inheriting the gene.</td>
</tr>
<tr>
<td>Acquired-variant (vCJD)</td>
<td>Contamination of BSE-infected material</td>
<td>Sometimes called nvCJD. People affected have been much younger. Age range 12–74 years with a mean of 29 years. Development of the disease is much slower than CJD, with an average of about 14 months between onset and death.</td>
</tr>
<tr>
<td>classical</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acquired-iatrogenic (iCJD)</td>
<td>Contaminated surgical instruments or human graft material</td>
<td>Produced by medical treatment, for example, dura mater grafts or pituitary hormones. In 2003 it was announced that a patient had died with vCJD, thought to be a potential case of secondary transmission through a blood transfusion from a person who went on to die with vCJD.</td>
</tr>
<tr>
<td>Acquired-Kuru</td>
<td>Ritualistic cannibalism</td>
<td>Isolated to members of the Fore tribe in Papua New Guinea.</td>
</tr>
</tbody>
</table>

Following publication of CJD guidelines for social workers, the Department of Health (2000) issued guidance for health workers, while the Queen’s Institute for Nursing (2003) wrote specific nursing guidelines.

People suffering from CJD need 24-hour care. They have great difficulty in swallowing and are likely to choke. In the early stages they need help with standing and walking, and once in bed permanently are likely to be doubly incontinent and need help with drinking and feeding. Patients need lots of pillows to cushion them from involuntary jerks, which can cause bruising and cuts. The physical effects of CJD are compounded by the fear and anxiety patients experience and they need lots of support and reassurance. It is also extremely difficult for families to watch their loved one deteriorating, knowing there is no cure for the disease.

**Care package**

Because of the challenging symptoms of CJD, each case requires a multidisciplinary, coordinated approach. A three-year case coordination project by the CJD Support Network, along with a report by Douglas et al (1999) and the findings of the BSE Inquiry (Phillips et al, 2000) enabled the Doh to develop a unique care approach by funding care coordinators to be based at the National CJD Surveillance Unit and by establishing the national care package for CJD.

The CJD Care Fund was also set aside by the DoH to cover shortfalls in care provision. The CJD care package works in conjunction with local health and social services, while the care fund minimises delays in the provision of care and equipment.

When neurologists suspect a case of CJD they inform the National CJD Surveillance Unit. A research doctor and nurse visit the patient and complete a risk factor questionnaire. If they confirm a possible or definite case of CJD a care coordinator will contact the family to offer to help local clinicians organise a case conference with a view to organising an appropriate care package.

**Infection control**

The key infection control challenge with CJD is the remarkable resilience of the abnormal prion protein. Exposure of infectious material to steam heat at 134°C may not completely inactivate it. However, prior treatment of instruments with appropriate detergents will reduce the amount of adherent tissue. These findings emphasise the importance of thorough cleaning of equipment before sterilisation.

Current evidence suggests that normal social or routine clinical contact does not present a risk to health care workers, families or others. While there is uncertainty about the level of risk from blood, universal (standard) infection control precautions should minimise risks.

**REFERENCES**


Queen’s Institute for Nursing (2003) CJD Information and Nursing Care Guidelines. Market Drayton: CJDSN.

This article has been double-blind peer-reviewed.

For related articles on this subject and links to relevant websites see www.nursingtimes.net
The DoH (2004) published updated guidance on safe working with transmissible spongiform encephalopathies (TSEs). These categorise patients into confirmed, suspected and at-risk groups and recognise different levels of infectivity in different tissues.

It is important to liaise closely with the members of the infection control team, as not all instruments will have to be disposed of following the treatment of patients.

The CJD Incidents Panel was originally set up as a subcommittee of the Spongiform Encephalopathy Committee and the Advisory Committee on Dangerous Pathogens to advise on incidents involving potential transmission of CJD between patients through clinical interventions. This may require tracking the surgical instruments used and taking them out of circulation, until such time as either the CJD Incident Panel recommends no action be taken or the instruments are destroyed.

Blood
Even before probable links with blood transfusion were found, it was believed that there might be a risk of transmitting CJD through blood, and the National Blood Service put in place measures to protect the national blood bank. In 1999 a system of leucodepletion of all blood components was implemented, while the NBS also recalled blood components, plasma derivatives or tissue obtained from any individual who later developed CJD. Plasma is now imported from the US to manufacture plasma derivatives.

Clinical trials
There is still much to learn about CJD. While it is always fatal, in the last few years potential prophylactic drugs have been identified and a clinical trial has been designed by the Medical Research Council Prion Unit.

Quinacrine, once used to treat malaria, is now being given to some patients in the hope that it will bind to the distorted prion and slow down the degeneration. Pentosan polysulphate (PPS) is another possible treatment, which is injected directly into the brain. Although it is not yet the subject of an official clinical trial, some patients have chosen PPS therapy with the support of the High Court.

Conclusion
Although there is still much to learn about CJD, our knowledge is developing rapidly. There have been some improvements in diagnostic tests, such as tonsil biopsies and magnetic resonance imaging (MRI) scanning, but more work is required in this difficult area, especially concerning the ethics of performing tests for incurable and fatal diseases.

It is hoped that treatments will soon become available that will at least slow the progression of the disease. In the interim it is vital that as soon as a case of CJD is diagnosed a case conference is convened with a multidisciplinary team to plan the patient’s long-term care (Barnett, 2004). This must be facilitated by a care coordinator to oversee the provision of care and support from many health care workers.

USEFUL CONTACTS

- CJD Support Network  www.cjdsupport.net  Helpline 01630 673973
- Department of Health  www.doh.gov.uk/cjd/index.htm
- Human BSE Foundation  www.hbsef.org  Helpline 0191 3894157
- National CJD Surveillance Unit  www.cjd.ed.ac.uk
- National Prion Unit  www.st-marys.org.uk/specialist/prion/index.prion.htm