

## Report on vCJD cluster at Queniborough

The investigation into the cluster of cases of vCJD in Queniborough, Leicestershire, was published on 21 March 2001.

The cluster of variant Creutzfeldt-Jakob disease in the North Leicestershire area was first recognised in July 2000. Between August 1996 and January 1999 five people developed symptoms that were later recognised as being those of variant Creutzfeldt-Jakob disease and they have all died. They all lived in the Wreake and Soar Valley area in North Leicestershire from 1980 until 1991.

The Leicestershire Health Authority study found an association which provides a biologically plausible explanation, suggesting that four out of the five people with variant Creutzfeldt-Jakob disease may have been exposed to the BSE agent through the purchase and consumption of beef from a butcher's shop where meat could have been contaminated with brain tissue.

It concluded that the technique used in a small local abattoir to kill animals resulted in contaminated meat finding its way to butcher's slabs and contaminating other fresh meats. It ruled out a range of possible explanations for the deaths of five people in the area.

Families living in the area who used the same sources of supply of meat as those families who have died may still be anxious about

their children who may have eaten meat from the same sources.

The length of the incubation period is such that we may yet see more cases. Health and social services need to be prepared to respond rapidly if this occurs. The information gained from Queniborough may help to explain other small apparent clusters.

There are two major questions raised by the report:

- How does the fifth victim who did not eat meat from a butcher's shop fit into the report hypothesis?
- The butchery practices described in the report must have taken place throughout the country, so how do they explain the cluster of vCJD cases in one village?

Arthur Beyless, treasurer for the CJD Support Network, feels that the results of this enquiry in Queniborough will still not help him come to terms with the death of his daughter, Pamela. Pamela, who died from vCJD two and a half years ago, was one of the Queniborough cluster.

*The full text of the report is available through the Leicestershire Health Authority website: <http://www.leics-ha.org.uk/>*

## Awards for families of people with vCJD

The CJD Support Network welcomed the news that Alan Milburn, Secretary of State for Health, has announced an interim payment of £25,000 to each of the families of those who have been diagnosed with vCJD.

Clive Evers, chairman of the CJD Support Network, said, 'Although the sum of £25,000 will not bring back a loved one, we recognise that this is good news for the families who have been affected. We look forward to the wider compensation packages that will be announced in the future.'

The compensation is completely separate from the national care package reported on page 3.

*Since the government announced this compensation package for families affected by vCJD, the CJD Support Network's helpline has been inundated with calls from families who have lost family members to classical CJD, not understanding the difference between vCJD and classical (or sporadic) CJD. We have an article on page 10 clarifying the different types of CJD.*

## On other pages

2-3 News

4 Letters

5 Hospice at home

6 Infection control in dental care

8 Swallowing problems

10 Strains of CJD

12 Support Network

## Care Grants

The CJD Support Network has awarded £28,220 in care grants over the last three years to families in financial need through caring for a relative with any type of CJD. The care grants have mainly been given to help with funeral costs or extra household bills.

The care grant fund is funded totally from public donations. We would like to thank everyone who has given donations, without which we could not support families in the way we do.

Those wishing to make a donation should send their donation, made payable to the *CJD Support Network*, to Gillian Turner. Every penny of your donation will go to families.

## French families sue governments

Two French families affected by vCJD are suing Britain for 'poisoning and manslaughter'. They have blamed the British, French governments and the European Union for allowing the sale of contaminated meat. (Daily Express 27/11/2000)

## Sore throat link

A leading American scientist, Professor Stephen DeArmond, has put a theory forward that victims of vCJD may have succumbed to the disease because they ate contaminated beef when they were suffering

from a bout of tonsillitis or a simple sore throat. British researchers studying vCJD said the theory was plausible, but stressed there was no experimental data to support it. (The Times, 16 March 2001)

## Blood scare

The Times on Tuesday 30 January reported that the government was trying to trace scores of haemophiliacs after the NHS admitted treating patients with a clotting agent made from a blood donor who later learnt that he had vCJD. The Haemophiliac Society stressed that the risk was theoretical and there have been no reported cases of haemophiliacs contracting vCJD.

# In memory of my daughter

**A Scarborough mum will be putting her best foot forward during the Walk for Life in memory of her daughter who died from the human form of mad cow disease.**

Anne Wilson, from the North Side of Scarborough, will be joined by her youngest daughter to raise funds and the profile of a support group for victims and their families.

Miss Wilson's 30-year-old daughter, Anita Bradshaw, died last year after being diagnosed with new variant Creutzfeldt-Jacob disease (CJD).

Her family could only watch helplessly as her condition deteriorated before their eyes. She started losing her memory and eventually the ability to talk, walk and eat. 'We were always holding out that maybe some treatment would come to light to save her, but it wasn't meant to be,' said Miss Wilson, who works at Scarborough Indoor Pool.

When Anita, from Accrington, first became ill doctors thought she was suffering from postnatal depression as it was soon after the birth of her second child. However, she failed to respond to treatment, her condition worsened and she was eventually diagnosed with variant CJD at the Royal Preston Hospital in December 1999. She died seven months later. Tests after her death showed she had a gene consistent with several other identified cases of CJD which could have made her more susceptible to the disease.

Miss Wilson is now busy raising funds for the CJD Support Network which has helped the family through a traumatic year. She took part in last year's Walk for Life when she collected about £230 for the organisation and will repeat her efforts during this year's event on Sunday 1 April with her ten-year-old daughter Maria.

'I was very emotional during the walk last year with tears in my eyes and concern because of the state



**Anne Wilson, ready to take part in the Walk for Life in aid of the Network**

Anita was in at the time, yet still hoping they would find a cure for her,' she said. 'I don't know how I will feel this year. I continue with life and keep busy, but things trigger me off out of the blue.

'It doesn't really matter how much we raise, it's a case of making people aware this organisation exists. When it first hit us we didn't know anything about a support group and had to search them out, but they were a big help.'

*Reprinted with kind permission from the Scarborough Evening News*

## Surgical risks

The Daily Mail reported on the hospital sterilisation proposals announced by the DoH on 4 January as a precaution against the spread of vCJD. It stated that the government's Spongiform Encephalopathy Advisory Committee (SEAC) had recommended disposable, single use instruments for operations on tonsils. It was also stated that the proposals will include restrictions on instruments used in brain and eye surgery and appendectomies.

### Grant for steralisation

The Government announced that £200m would go to modernise NHS decontamination and sterilisation facilities and on the introduction of single-use instruments for tonsil surgery to minimise the threat from vCJD.

### Dental survey

CJD Support Network and the Glasgow Dental School are working together to survey families with a family history of CJD about refusal of access to dentistry services. [See letter on page 4 and article on page 6].

### Tonsil operations cancelled

The Daily Mail (22 Jan 2001) reported that 5,000 tonsil operations had been cancelled in a CJD alert. Health officials had told hospitals to switch to disposable instruments as surgical equipment might be contaminated with vCJD. The guidance on using disposal instruments was issued 'because the particles associated with CJD have been found in the tonsils and adenoids of some people who suffer from this disease.'

Due to overwhelming demand, hospitals were unable to acquire disposable instruments as there was only one firm in the UK which made the instruments.

# £1 million national care package for CJD

Judy Anderson, Department of Health

**On 26 October 2000, following receipt of the BSE Inquiry Report, the government announced a package of measures for people suffering from CJD and their families, as well as the families of people who have already died from the disease.**

Forming a key part of this package was the establishment of a £1 million national care fund for all people with CJD. The care fund, which has been set up to meet the additional needs of people with CJD and their families, is intended to ensure a speedy response to diagnosis and improvements in the quality of care. Whilst it is not intended to replace existing services, it can be used to plug any gaps in local service provision and ensure that we can meet the rapidly changing needs of patients.

Voluntary organisations, including representatives from the CJD Support Network and family members, have played and will continue to play an important role in helping the Department of Health to ensure that the care fund is effective and meets the needs of patients.

The care fund is co-ordinated through the national CJD Surveillance Unit in Edinburgh. Gordon McLean, the care co-ordinator from the Unit is being asked to look at each patient's package of care following his attendance at a case conference arranged by the key worker. Together they will agree the care

package and make a decision on whether additional money is needed from the care fund to solve either health or social care problems.

The Department of Health is developing guidance for the NHS and social services. This will set out the processes for accessing the care fund, what the fund can be used for and who should be contacted for further information and advice. This guidance will build upon the Guidance for Healthcare Workers issued in August 2000.

A special advice team is also being set up by the Department. This team will consist of knowledgeable and experienced individuals from around the country, with first hand experience of caring for people with CJD, who can be called upon for advice and guidance when required.

*Guidance for Health Care Workers is available on the Internet at [www.doh.gov.uk/cjd/cjdguidance.htm](http://www.doh.gov.uk/cjd/cjdguidance.htm). Further information on the national care fund can be obtained by phoning Helen Wiggins at the Department of Health on 0113 2545301.*

## Can you help?

Dear editor

I was one of the ladies who received gonadotrophine treatment for infertility at the Woman's Hospital in Birmingham in the 1970s. When we were first told that we were potentially at risk of contracting CJD, we attended group support meetings at the hospital. However, these stopped and I have lost contact with other ladies in the group. Although I have made enquiries at the hospital no one can give me any information about the other recipients of the treatment.

The worrying thing is the fact that we have had to live with the fear that we may get CJD and we have had to carry this fear for years. The worry just takes over one's life and no one seems to care. I just feel that I have been thrown on one side.

Through your newsletter could I ask any other lady who has had this treatment and attended the group in Birmingham to contact me through your office.

*Jean*

## Dental problems

Dear editor

I feel that you should be made aware of the recent success that I have had with regard to the provision of dental equipment and treatment for myself.

In March 2000, I had a routine check-up with my local Dentist at a practice where I had been a patient for over twenty years. I had been given an information sheet to present to the Dentist from my local Health Authority Infection Control Unit. This notified anyone who came into contact with me, of the required medical 'control procedures' for patients, such as myself, with inherited Prion

disease. However, after I gave this information to the Dentist, he refused to proceed any further with me and placed all of his instruments into bags for disposal. (I should add that when I was first made aware that I was a 'carrier' of the disease, I notified the Head of Practice – who at the time was President of the British Dental Association – and he had no concerns whatsoever about treating me.)

For all future treatment, I was to be referred to Guy's Hospital in London and was told by the Public Dental Health Officer to go to the ME section of my local hospital if I had toothache in the meantime!

In order to make sense of this situation, I was advised to contact my local MP and after doing so, he agreed to find me a local dentist who would treat me in isolation. I had already contacted the local authority infection control nurse who had produced the information

sheet and she proved very helpful and supportive.

In all, it took nearly eight months to resolve my case, during which time my husband and I suffered considerable anguish. I am now very pleased to say that I have been seen by my new dentist who is very pleasant and that the necessary equipment has been provided for my sole use. I am also pleased with the accessibility of the practice, which is important for me as I am disabled.

I do hope that others in a similar position to myself will take comfort from this success and that they too may be treated locally before too long. The Dental Health Officer hopes to produce an article for the dental profession showing how my case has been resolved and I have given my full support to this.

*Sarah Ridgwell (Dunnett)*

*See our article about infection control in dentare care on page 6.*

## Diagnosis

A difficult diagnosis

### Richard Shepherd

In March 1999 Ricky, who was suffering from violent mood swings, was first seen by his GP. Approximately two months later he was admitted to a psychiatric ward but was released after only a week. His family were being told that he was simply 'attention seeking'.

After bringing him home, his mother asked the neurological surgeon if Ricky could be tested for CJD. She was concerned that he had previously worked in an abattoir and cut his arm there.

However, she was told that this was not simply a matter of a blood test and heard no more about it.

Ricky was cared for at home by his family and girlfriend until 23 December 1999 when he was admitted into the Royal Preston Hospital. At that point he could no longer stand or walk without help. He underwent tests and in January 2000 the family was told with 90 per cent certainty that Ricky had vCJD.

Ricky returned home in early February and was looked after by his family, girlfriend and an excellent nursing team until his death on 12 May 2000.

# Hospice at home

Amanda Hannigan, the Hospice at Home Team, Yorkshire

**We learn by our experiences, and during the past two years my nursing team and I have had three experiences nursing patients with CJD.**

I co-ordinate a Hospice at Home team in the Yorkshire region, a nursing service which provides additional support, comfort and advice, bringing the skills and practical care associated with 'hospice' into the home. Although our team was set up to provide care for cancer patients, we recognise that CJD patients need the full range of palliative care services available in the community.

**Alex**, a gentleman in his 60s, had, after weeks of investigation, received a diagnosis of CJD. He quickly lost independence, needing equipment and carers several times per day, respite care and support for his family and eventually 24-hour home care.

A small team of staff was involved as Alex had some visual impairment, hallucinations and a fear of strangers, so familiarity helped. The family was also very concerned about confidentiality and the numbers of people involved with care.

Alex became immobile, confused, unable to communicate, eat or drink and doubly incontinent. Three weeks after diagnosis, he died at home.

**Rachael** was a bright, attractive young girl in her 20s. The community care teams used the same approach together with advice from the National CJD Surveillance Unit. Rachael's parents had made some decisions before Rachael's condition prompted

intervention, such as discussing artificial feeding and hydration, and had decided against this option.

On a good day Rachel was able to go out and experience normal family life. On a bad day the family stayed in together or took advantage of our respite help – not going out but catching up sleep or work. Rachael too was able to remain at home.

**Gordon** was a gentleman in his 60s living with his wife and son. He was also able to enjoy family days out and trips to familiar places whilst well. As Gordon deteriorated, night time care was needed to enable the family to care throughout the day. The family needed support and advice which was provided by the district nursing teams on a daily basis. At night, his wife shared a cup of tea and a chat with the Hospice at Home staff. As with the other two patients, Gordon was able to remain at home.

## Common factors

We found many elements common to all three patients:

- Rapid deterioration
- Incontinence
- Shakiness/jerks
- Visual disturbances
- Hallucinations
- Swallowing problems
- The need for mobility aids, moving/handling aids, pressure relief appliances to be anticipated
- Family concerns about confidentiality
- Large numbers of professionals involved
- Need for a small team of 2-3 staff.

## What we have learnt

- The disease can have a **rapid progression**. Sometimes a patient can lose abilities and independence on a daily basis and needs must be anticipated to cope with this, eg the pre-ordering of mobility equipment.
- A small team of staff can **ensure continuity**, and help with visual disturbance and fear of strangers experienced by some patients. It can also reassure families about confidentiality.
- **Physiotherapy and gentle massage helps** with jerking ataxia and limb contractures and immobility.
- **Cup of tea care** – a lot of listening to patients, carers and families about their worries and anxieties. To lead them gently and prepare them for the changes to come. To give reassurance about confidentiality and privacy.
- To ensure **communication with all agencies**, especially out of hours services, which may be called upon in an emergency.
- **Education** – to help allay people's fears, and misunderstanding of the disease amongst professionals and the general public.

Our shared experiences have enabled our team to expand our knowledge and confidence in caring for patients with CJD and their families and carers.

# Infection control and the dentist



The dentist is a part of the multidisciplinary team involved in the care of patients with CJD. Oral health is an essential factor for maintaining quality of life. It is important that a dental examination is carried out as early as possible after an appropriate diagnosis has been made. This will enable prompt management of conditions such as extensive dental decay before it becomes too difficult to obtain full co-operation from the patient. This is also an ideal time for the dentist to meet the patient and discuss future oral care with them and their family.

There have been many fears expressed over the possibility of transmitting infectious prion protein during dental treatment. All dentists adhere to the principle of universal precautions, which means that all patients are treated to the same high standard of infection control. These minimum requirements are outlined in the British Dental Association advice booklet A12.

This means that, for most purposes, patients with CJD should be treated no differently from other dental patients. However extra precautions are necessary for dealing with contaminated dental instruments. This is particularly related to the fact that normal sterilisation methods are ineffective at destroying prion proteins from these instruments. To address these issues SEAC and ACDP have suggested that more stringent procedures be used for instruments used in the mouth. However, these procedures are not readily available in general dental practice (or dental hospitals). In order to overcome this problem we are recommending that dentists use disposable instruments on patients with, or at risk from, CJD. However, these procedures are costly for family dentists to follow so you may find that some practitioners will refer some patients to an NHS hospital or clinic for non-emergency treatment. ALL dental practitioners should see and treat dental emergencies for ALL patients.

# Dental care of patients with CJD

AJ Smith, Lecturer/Hon Spr Microbiology, Infection Research Group, Glasgow Dental Hospital and School, Glasgow.  
MY Martin, Senior Lecturer/Hon Consultant Microbiology, Dept of Clinical Dental Sciences, University of Liverpool

Looking to the future, treatment of patients with or at risk from prion diseases, in a normal family practice is likely to become the norm, once arrangements have been agreed to reimburse dentists for the cost of additional single-use instruments and similar precautions. Disposable mirrors and probes are already on the market and disposable handpieces have even been developed in recent years, in response to concerns about the spread of blood borne viruses such as Hepatitis B virus and HIV. Currently we are working with a major dental supplier to adapt these instruments and include them into a kit that can be used for the treatment of patients with CJD. It is important to remember that these instruments in no way impair the dental treatment of patients with CJD and the type of dentistry being performed should be such as to provide maximum comfort from minimum intervention.

## Further reading

Advisory Committee on Dangerous Pathogens, Spongiform Encephalopathy Advisory Committee (HMSO 1998). Transmissible spongiform encephalopathy agents: safe working and the prevention of infection.

Health Service Circular 1999/178 Variant Creutzfeldt-Jakob Disease (vCJD) Minimising the risk of transmission

Health Service Circular 1999/179 Controls assurance in infection control: Decontamination of medical devices

Creutzfeldt-Jakob Disease: Guidance for healthcare workers 2000 (<http://www.doh.gov.uk/cjd/cjdguidance.htm>)

BOA advice sheet A12 Infection control in dentistry BDA Education and Science Department (ed-sci@bda-dentistry.org.uk), tel 020 7573 4541

## Key points

- **Oral health is a key component of quality of life**
- **Dentists are part of the health care team and should be involved as early as possible.**
- **All dentists provide a high standard of universal precautions of infection control, which (apart from disposable instruments) pose no extra risk to other patients.**
- **Disposable dental instruments must be used on patients confirmed, suspected or at risk from CJD.**

## A reluctant dentist

Kathleen Goldsworthy wrote to Gill Turner regarding her dentist's refusal to carry out any treatment.

Kathleen herself does not suffer from CJD but her father died from the Sporadic strain in February last year. As a precaution, she was told at the time that she should not donate her blood or organs and that any surgical instruments used to treat her should be disposed of afterwards.

Kathleen asked if a blood sample could be taken from her father and tested for Genetic CJD. The test came back negative.

At a regular check-up appointment with her dentist, Kathleen decided to explain all of the above. Her dentist refused to carry out any treatment, aside from the check-up, or to allow her to see the hygienist. She was told that she would have to contact the hospital.



# Swallowing problems in prion disease

**Food and eating is central to living in terms of pleasure and socialising as well as survival, therefore eating and swallowing problems have a major impact both on the person directly affected and those caring for them. It is possible to compensate for many difficulties, and support and guidance from a speech and language therapist can make the difference.**

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Clare Morris, CANDID, Dementia Research Group, National Hospital for Neurology and Neurosurgery, London

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The ability to swallow efficiently and safely is controlled by the brain. Brain diseases such as prion disease may cause difficulties with swallowing during the course of the disease. If eating or swallowing is a problem for the person you care for, it is important to ask your GP for a referral to a speech and language therapist. They will be able to make an assessment of the problem and advise you on ways to make the process safe and more enjoyable. Other ways to access services are by contacting your local hospital or the Royal College of Speech and Language Therapists for information on services locally. Other professionals, such as a dietician, occupational therapist, physiotherapist, or district nurse, may need to be involved.

## The Cough

A cough is the body's response to 'foreign bodies' entering the airway or windpipe. It is our way of protecting our lungs from getting clogged up and interfering with breathing. Coughing is under neurological control and can therefore be affected by damage to

the brain. It is important to understand that if someone can cough when you ask them to, it doesn't necessarily mean they will cough to clear their windpipe. Similarly, if someone is unable to cough on request, it may be that they will have an adequate 'protective' cough.

## Aspiration

Aspiration is when liquids or food go down the wrong way and are not removed by coughing. It is not always obvious when someone is aspirating – it can happen 'silently'. While coughing can be alarming, it is important to remember that it is doing 'its job' in protecting the airway and lungs.

## Common problems

I have listed on the right some common problems and, on the opposite page, strategies used to facilitate swallowing. It is recommended, however, that the advice of a speech and language therapist is sought, as the strategies recommended will vary according to the way in which the swallowing mechanism is affected and the person themselves.

## Common problems with swallowing

- Difficulty chewing and/or difficulty moving food to the back of the mouth
- Spitting lumps of food out
- Eating very fast or putting too much into the mouth
- Eating insufficient amounts or refusing food and/or drink
- Talking with food or drink in the mouth and forgetting to swallow causing coughing
- Coughing/choking on food and /or liquids
- Complaints of food not going down or getting stuck in their throat
- a 'wet' or 'gurgly' voice after swallowing
- Difficulty swallowing tablets
- Dribbling
- Chronic chestiness or recurring chest infections
- Being unaware of food when it arrives in the mouth
- Failing to do anything with food in the mouth, just holding it there



# Ways to promote safer eating

Ways to promote safe eating fall into three categories:

## 1 Altering the way food is eaten

- Sitting upright, keeping the chin down. If you put your head back to drink, you are opening up the airway more – ie if the swallow reflex is slow, it is easier for food and drink to go down the wrong way.
- Take small sips of drink, perhaps from a teaspoon. Avoid the use of drinking vessels that encourage the head to tip back (eg feeder beakers)
- Take small mouthfuls of food.
- Alternate food and drink to help clear the mouth of food: this should be discussed with a speech and language therapist.
- Try encouraging the swallowing of each mouthful twice to clear any food or drink that may remain in the mouth or in the throat after the first swallow.
- If the person has not swallowed what is in their mouth, sometimes it helps to present an empty teaspoon rather than more food. This can encourage the second swallow mentioned above.
- Frequent swallows to counteract dribbling.

## Swallowing

For a fuller description of the swallowing mechanism see Morris, C and Murray, M (1997) 'Swallowing Problems – How to Help' in the *Journal of Dementia Care* Vol 5 No 2 Hawker Publications, London. Alternatively you can download it from the candid website ([www.candid.ion.ucl.ac.uk](http://www.candid.ion.ucl.ac.uk))

- Check mouth after finishing eating to ensure no food or fluid remains.
- You may have to sit with the person to remind them to use these strategies, and you may find that mealtimes take much longer.

## 2 Making changes to diet

- Special diets (soft or puree), merely avoiding certain foods, or preparing them differently can make a big difference. Foods that may present difficulty for someone with a swallowing problem include:
  - a) mixed textures such as food in a lot of fluid like minestrone soup, or cornflakes and milk
  - b) stringy textures such as bacon, cabbage, runner beans
  - c) floppy textures such as lettuce, cucumber
  - d) small, hard textures such as peanuts, peas, sweet corn and broad beansCooking food longer so it becomes softer, mashing food with the back of a fork or liquidising it in a blender can all help.
- Thickening fluids to yoghurt or sometimes porridge consistencies may help as they are easier to control. There are a number of thickening agents available through your GP or from a dietician. It is important to speak to a speech and language therapist about this. It can also be helpful to have guidance in using thickeners as the fluids may become lumpy which looks unappealing and may be offputting!
- The use of nutritional supplements if necessary.
- Crushing tablets or using a syrup form may be easier for someone with a swallowing problem but seek advice from your GP as some tablets need to be taken whole.

## 3 Using special equipment

Specially designed cups which allow drinking with the chin down; cutlery, plates and non slip mats are available. An occupational therapist can advise you on this.

Sometimes making these changes is not sufficient to ensure an adequate dietary intake. Feeding directly into the stomach is an alternative. This is called a gastrostomy, and it can be used in conjunction with eating small amounts orally, or can be used alone. The decision as to whether to introduce alternative feeding is a very personal one, and will involve patient, family and clinical staff.

## Further information

Information about prion disease, dementia, and speech and language therapy services is available from:

Prion Unit  
Department of Neurology  
St Mary's Hospital  
Praed Street  
London W2 1NY  
Tel: 020 7886 6883  
Fax: 020 7886 1422  
Email [k.prout@ic.ac.uk](mailto:k.prout@ic.ac.uk)

CANDID (Counselling and Diagnosis in Dementia),  
The National Hospital for Neurology and Neurosurgery,  
8-11 Queen Square,  
London WC1N 3BG.  
Tel: 020 7829 8772  
Fax: 020 7209 0182  
Email:  
[c.morris@candid.ion.ucl.ac.uk](mailto:c.morris@candid.ion.ucl.ac.uk)

The Royal College of Speech and Language Therapists,  
7 Bath Place,  
Rivington Street,  
London EC2A 3DR  
Tel: 020 7613 3855  
Fax: 020 7613 3854

# Strains of CJD

There is often confusion between the different types of CJD – particularly between the newest form, variant CJD, which is linked to BSE in cattle, and the most common form, sporadic (or 'classical') CJD. Dr Richard Knight, consultant neurologist at the National CJD Surveillance Unit, explains.

Creutzfeldt-Jakob disease is an illness which exists in different forms. These forms are essentially distinguished by the cause of the illness in each case. However, in addition, the symptoms and course of the illness tend to be relatively different in the different forms. Also, the neuropathological features (what one sees under the microscope) tend to be different in the different forms.

The four forms of disease are:

- Genetic CJD
- Iatrogenic CJD
- Sporadic CJD
- Variant CJD

## Genetic CJD

Genetic CJD is a very rare illness. In this form, CJD is caused by an inherited abnormal gene. The illness is therefore not 'caught' in any way and there is no causal relationship between genetic CJD and BSE. In most cases, the illness is known within the family because of the family history. Occasionally, genetic cases are seen in which no previous family history is identified. The definitive test in relation to genetic CJD is a blood test. This enables analysis of the gene to see whether there is any genetic abnormality. The UK has a population of around 58 million and there are only a few deaths due to genetic CJD in a year.

## Iatrogenic CJD

Iatrogenic CJD is also very rare. This is CJD which has been accidentally transmitted during the course of medical or surgical procedures. The most important example of this in the UK relates to CJD transmitted via human growth hormone treatment in childhood.

There are only a few deaths per year due to iatrogenic CJD in the UK. The diagnosis is usually clear from the history of a relevant medical or surgical treatment in the past.

## Sporadic CJD

Sporadic CJD is numerically the most common form of CJD. It is not confined to the UK and, indeed, has been found in every country in the world where it has been looked for. In general, it affects about one person per million of the population. There are therefore some 50 to 60 deaths per year due to sporadic CJD in the UK. Similar figures are seen in other countries in the European Union and other countries such as Australia, Canada and USA. The cause of sporadic CJD remains uncertain. However, the most favoured current theory suggests that the normal prion protein in the brain undergoes a spontaneous change to the abnormal form, thereby resulting in disease. If this theory is correct (and it has not been proven at this point) then the disease arises simply as a chance event inside the brain. On this basis it would not be 'caught' in any way.

## Variant CJD

Variant CJD was first reported in 1996. At this point in time the CJD Surveillance Unit has not seen any cases of variant CJD with symptoms that began before 1994. Aside from two cases in France and one case in the Republic of Ireland, variant CJD has been confined to the UK. The current view on variant CJD is that it has resulted from transmission of infection from BSE in cattle to humans via infectivity in food.

## Differences between sporadic and variant CJD

We have had many enquiries about the relationship between sporadic CJD and variant CJD. Sporadic CJD is of unknown cause. However, detailed investigation over many years has failed to provide any evidence to suggest that it is related to diet. It was first described in 1921 and therefore predates the BSE epidemic by many years. Sporadic CJD is found in countries throughout the world regardless of the presence of BSE. There is therefore no evidence to suggest that sporadic CJD is in any way the result of BSE. On the other hand, the timing of the appearance of variant CJD and its geographical distribution in the world strongly suggested a connection with BSE in cattle. Laboratory scientific work has shown that the protein agent involved in sporadic CJD has quite different behavioural properties from that seen in the protein agent from variant CJD. In addition, the behaviour of the variant CJD agent is very like that of BSE. There is therefore scientific laboratory evidence to support the view that variant CJD and BSE are related, whereas sporadic CJD is not causally related either to BSE or variant CJD.

## Clinical differences

Sporadic CJD and variant CJD have certain clinical and pathological differences.

The age of onset is generally different in variant and sporadic CJD. Variant CJD has tended to affect younger individuals with an average age of onset of around 27.

Sporadic CJD has tended to affect middle-aged and elderly individuals. However, this difference is not absolute. There are those with variant CJD with a relatively older onset (including one recent case aged 74). Sporadic CJD may also affect very young individuals on occasions, including those in their teens and twenties. There is therefore a small overlap in the age group affected by variant CJD and sporadic CJD. The age of an individual is not an absolute guide to the type of CJD.

### Duration of illness

The duration of illness is generally different in variant CJD and sporadic CJD. Many cases of variant CJD have durations of a year or more. The duration of sporadic CJD is typically a few months and, in a few cases, a few weeks. However, there is again no absolute distinction. There are

cases of variant CJD where people have died after an illness of only a few months and there are occasional cases of sporadic CJD with durations of one or two years or even longer. Therefore, the duration of illness is not an absolute guide to the form of CJD.

### Symptoms

The symptoms of sporadic and variant CJD tend to be different. In particular, sporadic CJD tends to present with a clearly neurological illness that follows a very rapidly progressive course. In variant CJD, the initial presentation is often with psychiatric or behavioral symptoms and it may not be clear that the individual has neurological illness until several months after the onset. An experienced neurologist can generally distinguish the clinical patterns of sporadic and variant CJD. However, there is some overlap in the symptoms of the two

forms, and, on occasions, it may be difficult to be certain as to the classification of the type of CJD if based on the clinical symptoms alone.

Some investigations which are undertaken in CJD may be of great help. In particular, the EEG and the MRI scan may be useful. The EEG shows a typical pattern in the majority of cases of sporadic CJD. The typical abnormality has never been seen in variant CJD. The cerebral MRI shows a typical abnormality in the majority of cases of variant CJD which has not been seen in sporadic CJD.

### Under the microscope

The neuropathological features of variant and sporadic CJD are different.

In determining whether an individual has CJD or not, the only

CONTINUED ON BACK PAGE

# Monthly Creutzfeldt-Jakob disease statistics

2 April 2001

These figures show the number of suspect cases referred to the CJD Surveillance Unit in Edinburgh, and the number of deaths of definite and probable cases in the UK, up to 2 April 2001

Year	Referrals	Sporadic	Iatrogenic	Familial	GSS	vCJD probable still alive	vCJD deaths awaiting pm. results	vCJD confirmed*	Total
1990	[53]	28	5	0	0	-	-	-	33
1991	75	32	1	3	0	-	-	-	36
1992	96	44	2	5	1	-	-	-	52
1993	78	38	4	2	2	-	-	-	46
1994	116	51	1	4	3	-	-	-	59
1995	87	35	4	2	3	-	3	-	47
1996	134	40	4	2	4	-	10	-	60
1997	161	59	6	4	1	-	10	-	80
1998	154	63	3	4	1	-	18	-	89
1999	169	61	6	2	0	-	-	15	84
2000	177	44	1	2	0	-	1	27	75
2001+	30	4	1	0	0	7	4	2	18

+ To 2 April 2001.

Total number of definite and probable cases of vCJD=97

\*including 9 probable deaths from vCJD without neuropathological confirmation.

absolute test at present is that of neuropathology. Therefore, if an individual has not had neuropathology undertaken on either a brain biopsy in life or at post mortem, then one cannot be absolutely sure of the diagnosis. In addition, the neuropathological features of sporadic CJD and variant CJD are quite distinct and this would represent the main definitive method of distinguishing between these two forms of CJD.

### 'Probable' cause

There are individuals who do not undergo brain biopsy in life and do not have an autopsy. These individuals may be diagnosed on the basis of 'probable sporadic CJD' or 'probable variant CJD'. Although this does not represent an absolutely definitive diagnosis, if an individual is considered as having 'probable' CJD, then it is very likely indeed that this is what they had. Probable sporadic CJD carries a certainty of around 95 per cent or more. To date, all the individuals who have been diagnosed with probable variant CJD in life, who have subsequently had an autopsy have been found to have had variant CJD.

# Who's who in the CJD Support Network



**Clive Evers**  
Chairman of the Network and director of information and education at the Alzheimer's Society.



**John Williams**  
John's daughter Alison died of vCJD. John is a retired local government engineer.



**Gillian Turner**  
National CJD case co-ordinator. Gillian acts as a link between network members, carers and professionals, co-ordinating care and answering enquiries on the helpline from a wide range of people.



**John Gilbert**  
John's brother in law died with classical CJD. John is a company director.



**Bill Mitchell**  
Vice chairman of the Network and a trustee of the Alzheimer's Society.



**Arthur Beyless**  
Treasurer of the Network. Lost his daughter Pamela to vCJD.

### CO-OPTED MEMBERS

**Kathryn Prout:** clinical nurse specialist at the Prion Unit, St Mary's Hospital London.



**Patricia Nolan**  
Pat's mother died with classical CJD. She is a senior social worker in Liverpool.

**Dr Richard Knight:** consultant neurologist at the CJD Surveillance Unit.



**Maria Byrne**  
Maria is a mother with three young children whose husband Graham had GSS.

**Sarah Shadbolt:** Sarah's husband died of classical CJD.

**Roger Tomkins:** Roger's daughter Clare died of vCJD.

## CJD Support Network membership application

The CJD Support Network is part of the Alzheimer's Society. Becoming a member of the Society adds to our strength and enables you to take a full part in the decision-making process and the work of the Network.

- I would like to become a member and receive the Alzheimer's Society's monthly newsletter.  
 I would like to become a member but **not** receive the Alzheimer's Society's monthly newsletter.

There is no fixed subscription, but please give generously to help our work.

£8  £12  £25  £50  Other

Please make cheques payable to **Alzheimer's Society**

However, if you are a carer and would appreciate free membership, please tick the box

Name  Title

Address

Postcode  Telephone

I am caring for someone with CJD:  at home  in residential care

I am:  a concerned relative/friend  former carer  professional  interested

CJDSN

**CJD**  
**SUPPORT**  
**NETWORK**

The views expressed in this Newsletter are personal and not necessarily those of the CJD Support Network.

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