CJD SUPPORT NETWORK

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Newsletter

Issue 13

March 2004

CREUTZFELDT-JAKOB DISEASE SUPPORT NETWORK NEWSLETTER

CJD Support Network family support meeting and annual general meeting

On Saturday 25 October the CJD Support Network held a family support meeting at the Quality Hotel, Westminster, London. It was a new approach for the Network to hold a family support meeting at which the annual general meeting took place.

The annual general meeting is very important for the network as it gives members the opportunity to learn what we have been doing in the past year and to enable them to have their say. It is historically very difficult to attract members to attend AGMs as it often means long journeys and a whole day's commitment at the weekend.

From members who attended we had feedback that the day was a success.

Dr Angus Kennedy, consultant neurologist at the national Prion Unit started the program speaking on CJD, diagnosis to post-mortem. Dr Kennedy was followed by James Meikle, health correspondent of *The Guardian* newspaper speaking on the public's perception of CJD. A summary of James Meikle's talk can be found in this newsletter.

There followed the eighth annual general meeting of the CJD Support Network, and the election of officers took place.

After an excellent lunch, Sir Iain Chalmers, chairman of the Prion 1 Trial Steering Committee spoke on the clinical trials for CJD. During his talk Sir Iain gave a account of his frustrations at the delays in getting all parties to agree the clinical trial protocol. He said that he was unaware, when he took up the challenge to chair the Prion 1 Trial Steering Committee, of the long standing acrimony between senior academics in Edinburgh and London, and that the Department of Health, far from confronting and containing this situation, had allowed it to fester. The delays have occurred despite the Department of Health notifying the NHS two years ago that clinical trials for CJD would be 'fast-tracked'. For further information on the clinical trials please read Lester Firkin's article in this newsletter.

Sir Ian was followed by talks from Fiona Barnett, care co-ordinator at the National CJD Surveillance Unit, and Katie Oakley, CJD incidents nurse investigator at the Health Protection Agency.

Both Fiona and Katie have written summaries of their talks and the articles can be found in this newsletter.

Summaries of some of the talks can be found on the following pages: James Meikle 10 • Fiona Barnett 11 • Katie Oakley 11

New strain of BSE disease discovered

James Meikle reported in The Guardian on the 17 February that Italian scientists have found a second form of BSE. A study of eight cows with BSE found that two had brain damage resembling that in human victims of the standard 'sporadic' form of CJD.

In addition to the holes in the brain caused by BSE – the form of disease most often found in cows – the Italian researchers found, in this sample, an accumulation of the amyloid brain plaque that is an indication of Alzheimer's disease.

However, while sporadic CJD and the new form found in cows share several characteristics, the researchers have cautioned against assuming a link between the two.

Regular readers of the CJD Support Network newsletter will have read about Prof John Collinge's research (Issue 12 May 2003, page 2) where he also linked some cases of sporadic CJD to BSE.

ON OTHER PAGES

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Email info@cjdsupport.net · Website www.cjdsupport.net · Charity number 1097173

CJD and blood transfusions

The health secretary, John Reid, gave the news to the House of Commons on the 17 December 2003 that a patient died of variant CJD after receiving a blood transfusion from a donor who was subsequently found to have developed the disease.

Mr Reid said it was possible, but not proven, that the disease had been transmitted by the blood transfusion. He told MPs that in 1996 a donor, at the time free of the signs of vCJD, gave blood to the national blood service. The blood was transfused into a patient who underwent surgery. The donor developed the disease in 1999 and died from it. The recipient died this autumn.

The health secretary said the NBC had informed the government that 15 people had received blood from donors who subsequently developed vCJD. He said the CJD Incidents Panel would be advising on a case-by-case basis, which recipients would need to be contacted as information became available, and that this group of patients would also have the opportunity for a discussion with an expert on an individual basis.

On the 15 March, the Health Secretary announced in the House that people who had received a blood transfusion since 1980 would not now be allowed to donate blood. This is a further precautionary measure against the possible risk of vCJD being transmitted by blood and blood products.

For further information on blood and CJD please read the National Blood Service article on page 00.

Recent CJD figures

The number of deaths of definite and probable cases in the UK, up to 1 March 2004, from the CJD Surveillance Unit in Edinburgh

	2002	2003	2004 *
Sporadic	72	63	5
Iatrogenic	0	5	0
Familial	4	2	0
GSS	1	2	1
vCJD	17	18	0
Total Deaths	**94**	**90**	**6**

Total deaths to date: 948*

*As at 1 March 2004

In memory

Nick Kannellides

Niko (Nick) Kannellides died of vCJD at the age of 22. His sister Anna arranged a Fun Day at work to raise awareness of CJD and raised £1,032 for the CJD Support Network.

Nina Sinnott

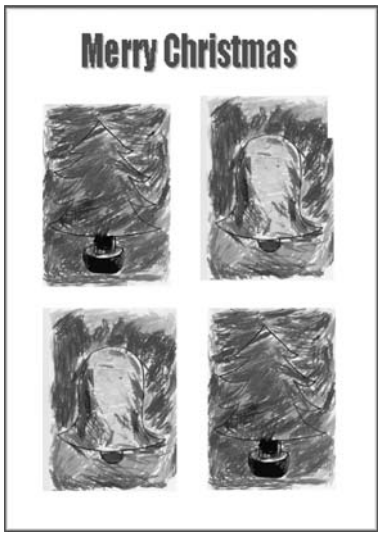
In memory of Nina Sinnott, who died of vCJD, her friend, Annaliese van den Berg arranged for her company to put a message on every email sent out during the Christmas period. The message said that the company would not be sending out Christmas Cards in 2003, but would donate a sum of £250 to the CJD Support Network.

Editor's note: thank you to everyone who has generously made a donation to the Network.

Two new greetings cards available

Two new designs – a CJD Christmas card and an open card ideal for thank-you notes, birthdays etc – are now available from the CJD Support Network on 01630 673993. Five cards can be ordered for a donation of £3 plus £1 post and package

The Christmas card has been designed by Colin Marsh, who is affected by familial CJD. The open card has a picture of the CJD rose, Nina Nadine, which was cultivated in memory of Nina, who died with vCJD. It is used with kind permission of her parents, Penny and Mike Sinnott.



Fundraising

Marathon and Land's End to John O'Groats cycle ride

By Duncan Byrne

My grandmother, Pamela Byrne, had made growing old gracefully an art form. Although I knew her to be exactly fifty years older than me (we shared the same 11th April birthday), nobody would have guessed it. She had won a glamourous grandmother competition in the 70s, and had faked her birth certificate so that she could continue to work at Tesco's until her 70th birthday. Due to her youthful appearance, no one guessed that anything was amiss. When the family met up in July 2001, she was in prime fitness, yet as the summer wore on, it was clear that something was wrong as she started suffering regular falls. After weeks of doctors trying to identify the problem, we were later to discover that this was the sporadic form of CJD. The speed at which my grandmother went downhill and more importantly the knowledge that there was no cure shocked the whole family.

Shortly after my grandmother's death in March 2002, one of my aunts (her daughter) suggested that I run the London Marathon in memory of her. Having run the Marathon in 2001 and 2002, I found out in November that my application for the 2003 race had been successful. I had been previously planning a Land's End to John O'Groats cycle ride for summer 2003 after the end of my summer term (I'm a schoolteacher), so I decided to put the events together. Thinking that 30,000 people run the Marathon and that the Land's End to John O'Groats cycle ride was quite a common challenge, I came across the idea of doing the two consecutively. The Marathon was taking place the first weekend of my Easter holiday, which would give me two weeks to complete the cycling leg before term started again. More importantly, my wife and I were expecting our first baby in May – if I didn't do the cycle ride now, there was no way I'd be getting two weeks off paternal duties in the future!

Marathon day (Sunday 13th April) was very warm, and I ran 3 hours 38 minutes, which was frustratingly only a minute slower than my best. After a large lunch and a warm bath, I headed down to Land's End with my mother (also a teacher) sharing the driving duties. We arrived at our B&B in Sennen (near Land's End) at about 10 o'clock with the landlord rather incredulous that I had run the marathon that morning!

Setting off on Monday 14th April at about 9.30 from Land's End (my longest lie-in), my mother accompanied me for three days, giving encouragement and logistical support. Although I vaguely knew my route, I hadn't booked ahead, knowing that I would have to respond to how my body felt. The first day was the hardest. Although getting onto the bike in the morning was no problem, my legs suddenly remembered after lunch that they had run 26 miles the previous day, and the steep Cornish hills were agony. I was determined to reach Tavistock, but doubts whether I would succeed in my challenge had been sown. Thankfully, the next day, I felt much better on my way to Wells (Somerset) and I thereafter knew that I could do it. Averaging about 100 miles a day (more than I had anticipated), I found accommodation in B&Bs or youth hostels in Worcester, Liverpool, Shap, Wanlockhead, Crianlarich, Loch Ness and Bettyhill.

Day 10 saw me roll into John O'Groats not a moment too soon, as my left knee had refused to work that morning, and I was reduced to cycling one-legged along the North Coast of Sutherland into Caithness. After the obligatory photo opportunity, it was back to Thurso (20 miles) and the afternoon train to Inverness, and thence onto the sleeper to London.

Then came the process of collecting money. I had already collected some on the journey, from some unlikely places – such as youth hostel keepers in Wanlockhead, the Halford's assistant who sold me a new front wheel, and the café owner in Thurso who gave me the cost of my meal back to contribute to the charity. My school, Whitgift in Croydon, had mobilised staff and parents, and friends had shown incredibly generosity. The entire Byrne family collected money from their work colleagues, allowing me to raise almost £4,000 for the CJD Support Network, which certainly made the whole endeavour worthwhile.

The funny thing is that I seem to remember my trip as just a series of black-and-white images, not quite believing that I actually did it!

Update on MRC Prion Unit Prion-1 Trial

Dr Dafydd Thomas, Consultant Neurologist, St Mary's Hospital and National Hospital for Neurology

The trial is about to start initially nationally and then to be extended internationally.

Quinacrine

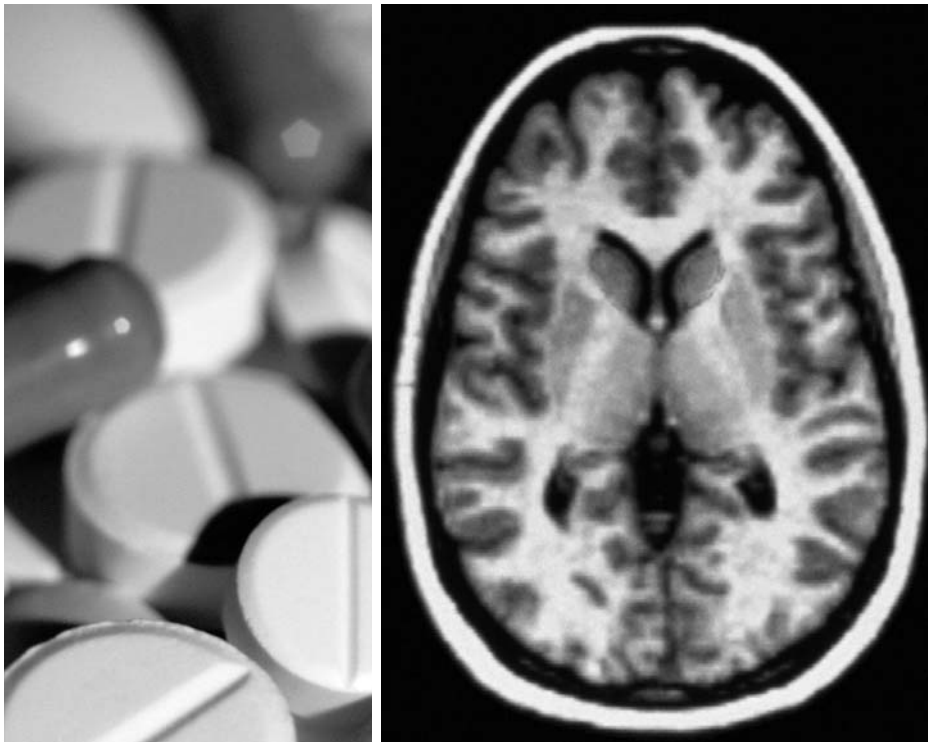
The first drug to be used in the MRC Prion trial is quinacrine, given orally. The pilot study, after local ethical approval, has taken place over the last two years centred on the National Prion Clinic at St Mary's. Almost twenty people have received quinacrine. It is usually well tolerated and there have been no serious side effects. One patient had to stop because of loose motions, another because of a skin rash. Virtually everybody has a slight yellow colouration of the skin which is a direct result of quinacrine and not an allergic effect – it is not jaundice.

The trial proper is about to start. Last month, it received provisional MREC approval (ie multiregional ethical approval) for it to be used nationally.

All patients will be offered quinacrine. Some will start immediately after their initial assessment, others can choose to delay starting treatment for several weeks. Using careful, frequent assessments, we should be able to detect a beneficial effect of quinacrine, if there is one. Clearly, the answer will be found more quickly the more patients are referred and included.

Pentosan

The MRC Prion-1 team has been asked by the Department of Health to monitor any patients who are given pentosan. This will be done confidentially. Patients having pentosan and their relatives will appreciate the need for collecting this important individual information, not only for their own benefit, but for others with CJD, now and in the future.



Quinacrine can be taken orally. Pentosan, because it cannot cross from the blood to the brain, has to be injected directly into the ventricles (fluid-filled cavities) within the brain

Treatment with Pentosan is very experimental. There is too little information on it at the moment to justify a proper clinical trial – more animal work is required. It has not been 'passed' by the Committee of Safety of Medicines or by the CJD Therapy Advisory Group. It does not cross the barrier between the blood and the brain so it cannot be given by mouth or by intravenous injection, it has to be given directly into the chambers of the brain (the ventricles). Tubes have to be placed into the ventricle on the left and the right side of the brain. This is done at a microsurgical operation. Tubes are connected to a further tube that is passed under the skin of the scalp and the neck to a reservoir in the chest wall or in the abdomen. Through this reservoir, the pentosan injections are given. No-one knows what the correct dose is or how frequently it needs to be given. Despite these uncertainties, and based largely on some anecdotal information, patients and relatives are sometimes desperate enough to request pentosan treatment. At the moment, in England and Wales, a court order is required to allow this to proceed. An injunction has been placed on the media to protect patients from the press. In Scotland, no court order is required and there is no media injunction in place.

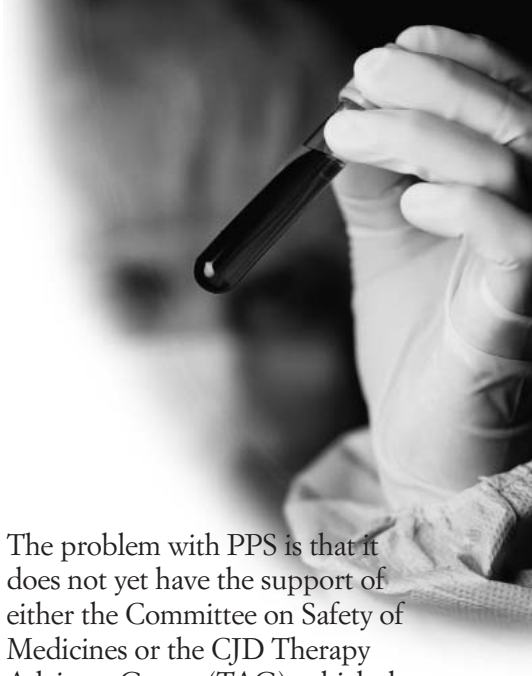
Monitoring this experimental pentosan treatment properly is clearly essential so that information can be co-ordinated as quickly as possible to allow more satisfactory advice to be given to patients and relatives.

I have now taken over from Dr Angus Kennedy as Consultant Neurologist to the MRC Prion-1 Trial and to the National Prion Clinic. Our thanks should go to Dr Kennedy for all the hard work he has put in over the last two years.

Dr Dafydd Thomas

Searching for a therapy

Lester Firkins



We are all acutely aware of the devastating effect that news of any form of CJD can bring to a sufferer, their family and friends. What compounds this pain is to learn that, at present, there is no known cure and therefore the outlook is terribly bleak. This short article is written to bring you up to date with what is going on as regards research into possible therapies – and in particular formal drug trials in the UK.

Scientists all over the world have their own theories on the nature of prion diseases and how they could be arrested. The two therapies most in the news at the moment are quinacrine and pentosan polysulphate (PPS).

Unfortunately, because of the current estimates of 'relatively' low numbers of victims, it is not a sufficiently large enough patient group to attract huge investments from drug companies into research. That said, the Medical Research Council Prion Unit is running a research programme trying to identify new potential therapies.

In May 2002 the Medical Research Council (MRC) organized an excellent consumer workshop to share with families the challenges of undertaking trials where there are (fortunately) so few sufferers – and also, most importantly to gain their views on the way forward.

Based on the opinions of the families and friends it was agreed to design a 'protocol' on which future clinical trials could be based. In essence a 'protocol' is a plan and set of rules by which a research team can work with patients (with any of the forms of CJD) and the medical profession, about how data can be collected, compared and analyzed.

We are very close to having such a protocol (known as Prion-1) signed off by the chair of the Trial Steering Committee. Progress in making arrangements for Prion-1 owes a great deal to the hard work of a number of people over many months – particularly by Sarah Walker of the MRC Clinical Trials Unit, Angus Kennedy of the National Prion Unit, and Richard Knight of the National CJD Surveillance Unit.

The research plans have now received ethics clearance and every victim and family who is affected by CJD will be offered the opportunity of participating in the Prion-1 trial. The protocol will be publicly available on www.mrc.ac.uk and thus contributing to the 'learning' that is achieved from any particular drug treatment. Not everyone will want to take a particular drug, and there will be no pressure to do so, but such patients can contribute to the research by allowing their progress to be monitored, so that comparisons can be made with those people who are taking a drug.

With any trial, the major challenge is to compare what happens to the progress of the disease among people taking drugs with otherwise similar people not taking them. In addition to ensuring that the comparison groups are as similar as possible, it is necessary to make sure that we are not misled by the play of chance, and this means that it is necessary to study a sufficiently large number of patients. Accordingly, to get reliable results, as many people as possible need to participate in the study (whether or not they take a drug). At the present time the drug selected for study is quinacrine, but the 'protocol' has been designed in such a way that if it was felt that another drug may offer a better therapy then, subject to proper checks etc, it could be offered to patients instead. PPS is much in the news at the moment and is 'under consideration'.

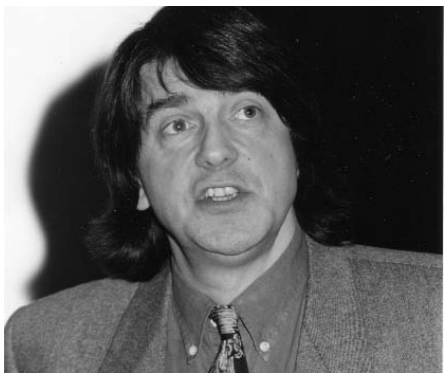
The problem with PPS is that it does not yet have the support of either the Committee on Safety of Medicines or the CJD Therapy Advisory Group (TAG), which the Department of Health established to act as the 'Radar' for new treatments. Whilst the TAG understands that some patients and families may wish to try the PPS treatment, it believes that there is currently insufficient data available on the safety of the drug (and in particular the way in which in needs to be administered direct to the brain) for them to support it. However against a backdrop of no known cure at the moment patients and families will be far more likely to 'take risks' than the establishment.

The Prion-1Trial Steering Committee wants to ensure that the experience of those treated with PPS is not lost, and it has urged the Department of Health to support use of the Prion-1 testing infrastructure for patients receiving PPS.

As a result of the MRC consumer day, everyone is acutely aware that the needs and desires of patients, their families and friends must always come first. As will be clear, proper testing of potential treatments for CJD is not only urgent but poses some very special challenges.

Lester is a member of the Therapy Advisory Group and assistant chairman of the Prion-1 Steering Committee. A former chairman of the Human BSE Foundation, Lester lost his son Ellis to vCJD in March 2001.

Intraventricular pentosan polysuphate (PPS) in prion diseases



A summary of the present position – December 2003

Dr Richard Knight, consultant neurologist, National CJD Surveillance Unit

PPS (pentosan polysuphate) is derived from beechwood and has anti-thrombotic and anti-inflammatory properties. It has been used in routine clinical practice for many years in the treatment of thrombotic disorders and interstitial cystitis.

Background rationale for treatment with PPS

The present background rationale for any treatment with PPS rests essentially on experimental laboratory work, in vitro (laboratory work with chemicals or cells) and in vivo (animal) work.

The in vitro experimental results can be summarised as indicating that PPS has effects on prion protein production, replication and associated cell toxicity. However, there are certainly other chemical compounds which have shown in vitro promise that have then failed to show any clear cut clinical effect when used in the treatment of human disease. In addition, while the prion protein is central to diseases like CJD, the precise mechanism by which neurones become damaged and die is uncertain in prion diseases. Also, currently, there is no scientific rationale to suggest that drugs like PPS will cause or aid any recovery to previously damaged neurones.

The in vivo experimental results can be summarised as showing that PPS has a prophylactic effect in various animal models. In other words, if PPS is given to experimental animals at a time relatively close to the point of experimental infection, then there may be an increase in the incubation period of disease (ie a prolongation of the time between inoculation of infectivity and the appearance of clinical disease). In some instances, animals appear to be completely protected from the development of disease.

The clinical relevance of these findings is, of course, uncertain. Firstly, it is difficult to know how to extrapolate from laboratory animals (typically mice or hamsters) to humans, especially as there are no data concerning the experimental treatment of potentially more related animals, such as non-human primates. Certainly, the reported in vivo experimental effects of PPS, as with most drugs, are highly dependent on the dose and route of administration. Secondly, these experiments usually involve species adapted strains of prion disease (often rodent-adapted scrapie) and it is uncertain whether these results can be extrapolated to all prion strains in naturally occurring situations. Thirdly, the demonstration of a prophylactic value may not immediately relate to the usual human situation where patients present with established disease. While some researchers have suggested that PPS has benefits in established disease, there is no specific experimental evidence of this. In the case of humans with variant CJD (and other forms of acquired CJD), by the time they become clinically ill, they have already gone through the incubation period. In sporadic CJD, the concept of 'incubation period' does not simply apply, but there is likely to be some period of clinically silent disease development before the onset of actual symptoms. The duration of this incubation period for variant CJD is uncertain but the minimum incubation period may be 5 years and it would not be surprising if the average incubation period were around 10 years. It is important to realize that the current treatment of humans with CJD is a treatment being given after the actual onset of clinical disease. If a treatment is efficacious in progressive neurological disease, then it is very reasonable to believe that the sooner it is given, the better the positive results. Therefore, it is important that patients seeking treatment with PPS be given it as early as possible. However, the diagnosis must be as certain as possible when considering a treatment like intraventricular PPS administration and this may take a while, especially as current diagnosis necessarily involves observing the clinical progress over a period of time. Clinicians do see individuals who present in a manner similar to that of CJD and yet turn out to have other illnesses (sometimes with specific treatments of their own). The development of better, earlier diagnostic tests would be helpful in this respect, but, at present, clinicians must work with the available methods of diagnosis. The use of treatments like PPS in the incubation period of human CJD (prior to any symptoms) is not presently feasible; such an approach would require definitive demonstration of safety and efficacy in humans and a validated pre-symptomatic test (that does not, currently, exist).

Whatever evidence is available suggests that PPS may have some effects on the disease mechanisms of CJD, but there is not evidence that it reverses (or even halts) neuronal death and the associated neurological dysfunction.

A recent report from Doh-ura and colleagues from Japan concerns the intraventricular administration of PPS. PPS does not cross the blood brain

barrier. Their preliminary results (in animal experiments) suggest that this is a possible means of delivering PPS to the central nervous system using standard neurosurgical techniques. This is a preliminary report, relating to experimental animal work, involving a hamster-adapted scrapie strain in experimental mice expressing hamster PrP and again essentially studying incubation period. Therefore, although this work represents something new (in relation to mode of administration), it does not provide a firm scientific rationale for the routine clinical treatment of humans with established disease. Doh-ura's work is not yet published (although it is reported as having been accepted for publication by a scientific journal) and therefore only limited data are available.

There is UK DH-funded research being undertaken by Farquhar et al into PPS in animal prion models. This includes studying BSE (301V) in mice, as well as mice and hamster scrapie models. However, there are only very limited data published at this point.

It is important to understand that the precise pathogenesis of prion diseases (including variant CJD) is not completely understood. It is not necessarily the case that actions directly involving the prion protein are the specific or only reason for efficacy of drugs like PPS and other modes of drug action might be clinically useful. From a strictly clinical point of view, the mechanism of action of drugs is a rather secondary theoretical matter in relation to the important practical one of whether these drugs actually work. As indicated above, PPS does indeed have some kind of efficacy in experimental animal model settings. On present understanding, any process which slows or even halts the progress of CJD is extremely unlikely to lead to any recovery of previously injured or dead neurones. There is, therefore, limited scientific rationale for the use of PPS in prion diseases but no good scientific rationale for its being employed as a routine treatment in clinically established human prion disease. Of course, the absence of evidence is not the same as the evidence of absence.

However, there have been specific individual circumstances in which clinicians, affected individuals and their families have wished to consider the treatment, particularly given the inevitable progression and fatal outcome of prion illnesses. Indeed, there are several UK individuals who have received the treatment in these circumstances over the last year or so.

Is there any other presently available treatment with proven therapeutic benefit?

There have been other compounds of recent interest (eg quinacrine, flupirtine), but no definitive reports of benefit. Quinacrine is being assessed in a clinical trial at the National Prion Clinic in London.

Currently there is no proven therapy for CJD in clinically ill humans.

The blood brain barrier problem

Essentially, if one is to treat established clinical disease, the central nervous system, ie the brain, will be involved. Therefore, it is logical that any treatment that is to be effective must reach the brain. PPS does not readily cross the blood brain barrier. Therefore, intraventricular administration has been proposed and, in fact, used in order to deliver the drug directly to the central nervous system, avoiding the blood brain barrier. This sort of approach has been taken in other clinical situations, with other drugs, in different diseases. PPS unfortunately does not cross the blood-brain barrier after oral administration and intraventricular administration delivers PPS to the brain. Spinal intrathecal administration has been considered (but it has not been tried, as far as is known). It is thought, amongst other considerations, that it is not likely to give sufficient concentrations at brain level.

The possible risks and benefits of intraventricular PPS

Risks of neurosurgery

There are clearly risks involved in any neurosurgical operation and any anaesthesia. The surgery involves opening the skull by means of drilling a small hole and inserting an appropriate thin tube device into the ventricles of the brain. This must be associated with risk including cerebrovascular trauma and potential infection. However, this kind of neurosurgical procedure is used (for a variety of reasons) on a regular and routine basis in all neurosurgical units. The risks are very small and can be discussed with the appropriate neurosurgeon if such treatment is being considered. Long-term intraventricular access (as is required in this context) may be associated with some problems that, again, can be discussed with an experienced neurosurgeon prior to treatment.

Risks of PPS

PPS does have anticoagulant properties and there is therefore some potential for haemorrhage in the brain, either related to the drug administration itself or to the drug administration associated with the surgical trauma. There is also experimental evidence to suggest other toxicity. Any side effects of PPS (perhaps particularly any associated haemorrhage) will be partly dependent on the dose administered and on the route of administration. The relevant route for consideration here is the intraventricular one.

In Doh-ura's experimental work with intraventricular PPS, there were significant side effects. However, these were dose- and species-dependent. With lower doses of PPS (110-230 g/kg/day), there were no adverse effects. With higher doses (eg 345 and 460 g/kg/day), there were adverse effects in dogs but not in mice. In some dogs, at these higher doses, there were seizures and some of the affected dogs died. Some deaths were associated with intracerebral haematomata, but not all. Again, it is important to note that this work is not yet published and so detailed data are not available. However, the three basic facts are:

1 Adverse effects are dose-dependent.

2 Adverse effects appear to species-dependent.

3 Fatality is not always related to haemorrhage.

In the DH-funded work of Farquhar and colleagues, there are preliminary data concerning toxicity. In some mice scrapie experiments, deaths resulted from intraperitoneal and intravenous PPS administration, some related to haemorrhage but others apparently not. Simultaneous intracerebral (not intraventricular) inoculation of scrapie and PPS resulted in fits and death, by unspecified mechanism. The PPS dosages used were reportedly much higher than those used in the Doh-ura rodent experiments and there has been no experimentation using the intraventricular route. There are relatively few data in the public domain from this group at present.

It is reasonable to postulate that these toxic effects relate to high doses of PPS being administered either directly to the central nervous system or gaining access to the central nervous system because of a breached blood brain barrier at the time of intracerebral inoculation of scrapie.

It is difficult to provide a definitive assessment of these results. However, the simplest interpretation is that there is dose-related toxicity in relation to PPS and the central nervous system. In the relatively lower doses used by Doh-ura and colleagues, there was no evidence of such toxicity.

It is difficult to know whether such toxicity is species-specific in any way. However, the adverse effects associated with higher dosages in Doh-ura's work were observed only in dogs and not in mice and rats. This certainly could argue for species variability and therefore one would have to have additional caution in extrapolating any indications of safe and toxic doses to human beings.

Intraventricular PPS treatment has been undertaken in several individuals with prion illnesses (it is not possible to state the number treated with certainty). There are few details in the public domain, but no significant side effects of PPS have been reported to date, despite some 14 months of treatment in at least one case. There are no reports in the public domain of complications relating to the surgery or to the long-term placement of intraventricular catheters (this does necessarily not mean there have been none). The precise dosage schedule used in these patients has not been published, but, in at least some cases, it is understood to correspond to the lowest dose used in the Doh-ura studies (with suitable corrections from animal to man).

It is possible that this treatment will slow (or even halt) progression of disease but there is no guarantee

Benefits 1

It is possible that this treatment will slow (or even halt) progression of disease but there is no guarantee and no present means of objectively quantifying the degree of possibility. Any such effect, IF it were to occur, would probably be temporary. It is not possible to give any indication of any time limit on such an effect. The possibility that any such effect could continue throughout the duration of treatment cannot be absolutely excluded but it seems unlikely. Clearly, the later in the disease process that such a treatment were to be undertaken, the less likely any benefit would be seen. Also, in late stages of disease, slowing of progression might be difficult to objectively assess. There are, currently, no validated laboratory or imaging assessments that could be used to give unequivocal evidence of efficacy; one would have to rely on more subjective assessments including everyday functional ability and serial neurological impairment examinations, perhaps with the adjunct of neuropsychological assessments. However, research is being undertaken to see if certain tests could be useful in monitoring disease progression.

Benefits 2

On the basis of current understanding, there is no realistic possibility of actual improvement in the sense of reversal of previously established neurological deficit. Again, this cannot be absolutely excluded but it seems highly improbable, especially in relatively late stages of disease.

Benefits 3

To date, at least several human individuals with prion disease have been treated with intraventricular PPS. There are also families who have considered the treatment and decided that they do not wish for it. There are no hard data currently in the public domain. However, one individual (with vCJD) is said to have not shown any clear evidence of deterioration over a period of at least 14 months. The person concerned was at a late stage of illness at the time of treatment, with very significant neurological impairment and it could be difficult to assess any signs of deterioration or minor improvement in this sort of situation. This period of apparent clinical stability could be taken as evidence that this treatment has indeed had a beneficial effect in this one individual. However, some individuals with prion diseases go through 'plateau' periods and, to some extent, survival in the later stages of illness depends on the level and quality of general nursing care provided. The present duration of apparent clinical stability must at least suggest the possibility of some treatment efficacy. In addition, there have been suggestions of minor clinical improvements. However, at present, it cannot be stated that this one treated individual provides definite evidence of efficacy of intraventricular PPS. No information is currently available on the other treated individuals.

The best possible outcome from intraventricular PPS

On the basis of the available evidence, the best possible outcome that could be expected after treatment with intraventricular PPS is that there may be some temporary slowing or halting of the disease progression. However, there is little likelihood of significant clinical improvement. Nor is there a likelihood of permanent halting of disease progression. Of course, to some extent, this might depend on the duration of intraventricular PPS administration. It is not clear on what basis one would decide on the duration of treatment.

Naturally, a treatment which stabilises an individual's condition

could conceivably lead to that individual being in a state of potential suffering for a longer period of time. It might be proposed that any slowing of progression or halting of progression might allow an individual to survive longer and therefore receive more beneficial treatment if it were to become available in the future. However, this would be a speculative view and, while treatments for CJD are being researched, there is no realistic expectation of a complete cure in the immediate future.

There are clearly important issues of consent and issues about the full understanding of those involved as to the potential benefits and risks

Additional comments

Any conclusion concerning these above considerations, in the context of an individual person, would necessarily involve a number of very difficult or personal judgements about quality of life and the degree of suffering experienced by an individual in a disease like CJD. Any such judgements are bound to be subjective and reflect both general belief systems and personal evaluations of the individual patient. There are clearly important issues of consent and issues about the full understanding of those involved as to the potential benefits and risks. Clearly, there are few hard data on which to make clear decisions. At this point, the human treatment data do not allow for any specific comments can be made concerning problems or benefits, aside from the facts that there are no reported major serious complications and that there has been no obvious clinical deterioration over some months of treatment in at least one case.

Any decision about a given patient would have to be taken in an entirely individual way, based on a detailed assessment of both the patient and the concerned relatives. The overriding principle should be, what is in the best interests of the individual patient? Bearing in mind that CJD is inevitably and invariably a progressive and fatal disease.

There is, of course, an argument that such treatment should be evaluated in the context of a properly organized clinical trial. There are no current plans to set up such a trial and, in relation to this specific issue, the comments of two relevant professional bodies (given below) should be noted.

Advice from relevant professional bodies

The DH have statements on intraventricular PPS on their website: www.doh.gov.uk/cjd/pentosan_revised.htm. This includes statements of advice from the CJD Therapy Advisory Group and the CSM.

The advice from the CJD Therapy Advisory Group can be summarised as follows:

- There are insufficient clinical data to support the claim that PPS is effective during clinical disease.
- There are insufficient safety data on which to base a rational treatment regimen in humans.
- Further animal model experimental work is warranted.
- Nevertheless, at the dosage used in at least one individual, there have been no definite harmful effects attributable to the drug.
- All patients with prion disease should undergo appropriate monitoring during disease progression, in a way that allows collection of data on the natural history of disease and on any treatments that might be given.

The advice from the CSM is similar and states further that:

- There is no evidence in support of its use as a treatment in late stage disease.
- In the light of the limited information on PPS treatment of clinically established vCJD it is impossible to assess the risk/benefit relationship of PPS in these indications.
- There was insufficient information to reach any conclusions about the efficacy of treatment in [the single case known about at the time of the statement].
- They also recommended that further study of PPS should be undertaken in a clinical trial setting.

Neither of these statements precludes the possibility of an individual clinician deciding to treat an individual patient; such decisions remain absolutely individual ones. However, clinicians would wish to consider any such decisions in the light of all the available information and advice.

The case of the first individual who received this treatment was referred to the High Court and there was a ruling in favour of this particular individual being allowed to receive such treatment, as being in that individual's overall best interests. However, this was an individual ruling relating to a particular individual. It is understood that other cases have been referred to court, but again on an individual basis.

There are important issues of consent regarding such 'experimental' treatment, both with regard to the age of some affected individuals and also with regard to competence (where disease affects the brain).

The Department of Health has sought to identify certain selected hospitals where intraventricular PPS treatment might be instituted, if there is a strong desire for it by a family and agreement by their relevant clinician, so as to centralise any experience with this treatment. Such centres will develop protocols for the referral of patients and the process of arranging such treatment (including the various potential legal issues, such as consent) should thereby become simpler. However, there are no current plans to establish a formal scientific trial of intraventricular PPS and the treatment is one that is being given essentially on a speculative basis. The present provision of IVPPS remains an individual decision between patient, family and their immediately responsible clinician, after full understanding and discussion of the facts as detailed above.

Summaries of three talks given at the CJD Family Support Meeting and annual general meeting held in London, in October last year.

See the cover for a general report of the proceedings.

The public's perception of CJD

James Meikle, health correspondent at the Guardian

James Meikle put media coverage of CJD in the context of the debate over risk and 'responsible' news reporting. He cited a report from the King's Fund health think-tank implying the media were not balanced in choice of news and feature material.

He quoted figures collated by researchers which calculated how many deaths were needed for particular news articles or broadcasts to appear. On BBC outlets, there were around 8570 deaths by smoking per story, 4700 deaths for each alcohol story, 2500 deaths per obesity-related story and 0.33 deaths for each vCJD story. In newspapers, there were 4440 deaths for each smoking story, 2540 per obesity story, 846 per alcohol story and 1.5 per vCJD story. And vCJD was regarded as 'a scare story' rather than a proven health risk. 'In other words, I should be writing a little less about you and a bit more about other things. Quite apart from the fall-out for public health policy from the vCJD disaster, particularly in areas such as blood supply and decontamination of medical instruments, I disagree.'

James Meikle explained that the established risk of today, such as smoking's link to cancer and heart disease, was the less established risk of 50 years ago. Anyway, despite this so-called over-coverage, the public still did not know very much about CJD in general. There was often media confusion, discussing variant CJD as if it were the same as sporadic, for instance journalists, let alone the public needed further education. Most thought the BSE and vCJD crisis was over, unaware of incubation periods or how public health officials were still keeping their fingers crossed. Mr Meikle discussed pressure from within newspapers to personalise health stories to make them accessible to readers. Cancer charities and drug companies often provided 'case studies' of volunteer patients for press announcements. But most people knew someone who had or had had cancer, even if they had not had the disease themselves, and could use this experience judging any media news about, say, an improvement in treatment. With the one in a million incidence of CJD, perhaps it was different. It would be beyond most people's experience. 'Going public' was very much up to individual families. In some cases, with their permission, he had named them. In others, he had advised families not to be identified because of the media attention that might result.

He also discussed the difficulties of reporting CJD incidents, marrying journalists' desire for a 'scoop' with proper consideration of the consequences on families. When writing about possible contamination of surgical instruments at Middlesborough General Hospital in 2002, he had hoped to give the hospital time to inform patients who might have been at potential risk before the news was in print, but there was a communications failure between Department of Health officials and the hospital. An official inquiry exonerated the hospital but blamed people who had talked to him for causing what was said to be 'unnecessary suffering' to families. He argued that it had been a matter of public interest.

Visit Guardian Unlimited – www.guardian.co.uk

The Health Protection Agency

Katie Oakley, CJD incidents nurse investigator at the Health Protection Agency

Katie outlined the role of the CDSC (Communicable Disease Surveillance Centre) CJD Team based at Colindale in North London. CDSC is now part of the Health Protection Agency. The Health Protection Agency (www.hpa.org.uk) is a new national organisation for England and Wales established on 1st April 2003.

The CJD team is headed by Dr Nicky Connor, consultant epidemiologist and medical secretary to the CJD Incidents Panel.

The CJD team:

- Provides the secretariat to the CJD Incidents Panel
- Supports local public health teams investigating and managing incidents
- Is establishing a national anonymous tonsil archive for studies of detectable abnormal prion protein
- Coordinates the investigation of geographically associated cases of vCJD.

Katie then focussed on the work of the CJD Incidents Panel (www.doh.gov.uk/cjd/incidentspanel.htm) and took questions.

CJD Care Team

Fiona Barnett, care co-ordinator at the National CJD Surveillance Unit

The CJD Care Team was set up following the BSE Inquiry (2000) and Douglas et al (1999) which highlighted deficiencies in the care of patients with CJD. Gordon McLean was employed as a national care co-ordinator in February 2000 and other team members followed in September 2001.

The CJD Care Fund is an additional sum of money set-aside by the Department of Health and designed to cover shortfalls in care provision across the UK. It is administered and accessed by the two national care co-ordinators. The CJD Care Package does not replace local health and social services responsibility to provide and fund appropriate services but works in conjunction with these agencies. The purpose of the CJD Care Fund is to assist in minimising delays in the provision of care and equipment thus allowing a person with CJD to remain at home if so desired.

The activities of the CJD Care Team include:

- Ongoing support to the patient and family
- Visits if required 4-6 weekly
- Attendance at case conferences
- Teaching sessions and communication with local professionals
- Post-bereavement support
- Service development
- Dealing with general enquires about CJD
- Administration of the National Care Fund

References

The BSE Inquiry (2000) *Volume 1: Findings and Conclusions.* House of Commons paper 1999-00 887-1 The Stationary Office London

Douglas MJ, Campbell H and Will RG (1999) *Patients with new variant Creutzfeldt-Jakob Disease and their families: care and information needs* http://www.cjd.ed.ac.uk/carers.html

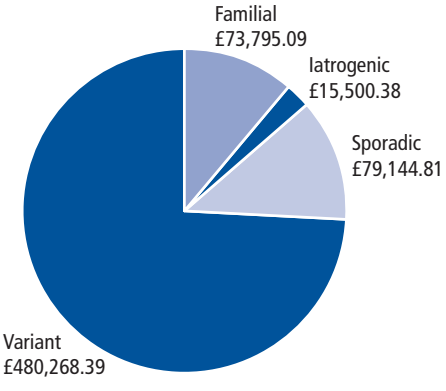
CJD Care Fund expenditure

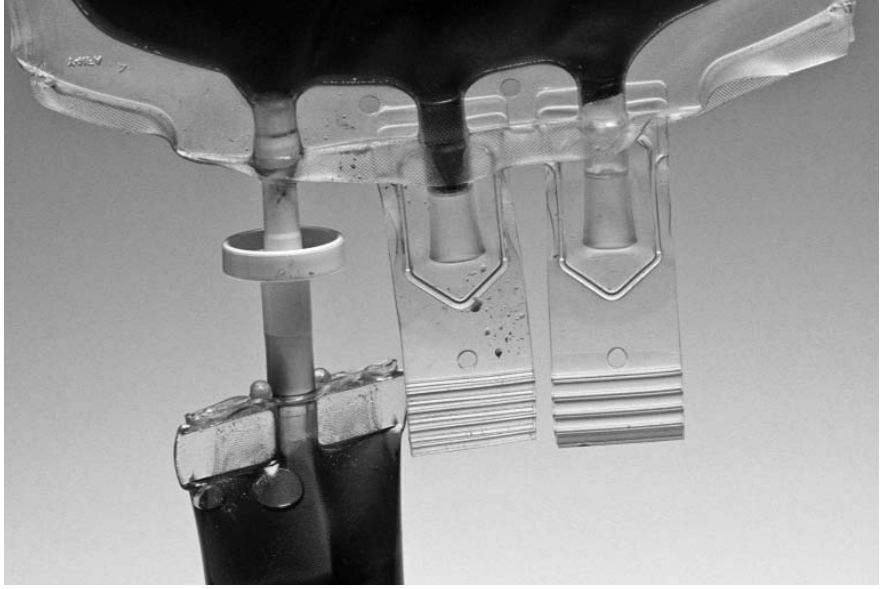
Total Care Fund Expenditure October 2000 to December 2003

Nursing	349,300.25
Equipment	141,598.20
Adaptations	60,684.84
Car Hire	35,432.91
Respite	19,957.03
Alternative Therapy	18,075.19
Accommodation	9,133.43
Social Care	6,011.93
Childcare	2,970.50
Transport	2,274.00
Physiotherapy	1,970.36
Counselling	1,300.00
TOTAL	£648,708.67

Expenditure by type of CJD

4 vCJD patients, 14 sCJD patients, 8 familial patients and 4 iatrogenic CJD patients accessed the fund between October 2000 and December 2003





Blood and CJD

Written by the National Blood Service

Q Is UK blood safe?

A The UK has an exceptionally good track record of safe blood. But like any medical procedure transfusion does have a small risk associated with it. To make sure blood is as safe as possible we have extremely sensitive tests and controls. We constantly review our safety methods to ensure that they are appropriate, in the light of new developments such as vCJD.

Any risk associated with receiving blood must be balanced against the risk of not receiving that blood when it is most needed.

Q With the first reported case of BSE in the USA, will the NBS continue to import plasma from the USA?

A Yes. The original risk assessment never assumed that there would be zero BSE cases in the US. It is important to remember that in comparison there have been over 750,000 BSE cases and 145 vCJD cases in the UK. Therefore the scale of the problem within the USA and UK is entirely different.

Q Should patients be worried about receiving a blood transfusion?

A Like all medical treatments a blood transfusion should only be administered when really necessary. The decision to give a blood transfusion to a patient is made only after careful consideration. In making that decision the doctor will have had to balance the risk of having a blood transfusion against the risk of not having one.

Patients still worried should contact NHS direct on 0845 46 47.

Q How many patients have been exposed to blood components from donors who went on to develop vCJD and will they be informed?

A There are currently 16 living recipients in the UK of blood components from donors who went on to develop vCJD. All have been informed of the circumstances

The CJD Incidents Panel is calculating which groups of patients will need to be contacted following receipt of plasma products from batches contributed to by donors who went on to develop vCJD. Some patients have already been contacted.

Q Are there any additional precautions that can be applied?

A Expert advisors continue to review current precautions and possible future precautions.

The majority of blood components are derived from UK donors. It is unlikely that such quantities of blood could be sourced from non-remunerated donors outside of the UK. Even if this was possible, it could increase the risk of exposure to other infectious agents, would be difficult to implement for components with short shelf lives, and could precipitate shortages.

Q Is there a blood test available for vCJD?

A No. There is no blood test available world wide for vCJD.

Q Can blood donors contract vCJD from giving blood?

A No. Blood donations are taken through sterile, non-reusable, disposable needles and equipment. It is safe to give blood – you cannot catch anything from giving blood.

Q What is being done to ensure that blood is used appropriately?

A National programmes for good transfusion practice have been established, supporting the work of local hospital transfusion committees. Transfusion practitioners have been or are being appointed to many hospitals. There is increasing use in the NHS of techniques that can, for some patients, reduce the need for donor blood.

Q If someone has been infected with vCJD because of a blood transfusion, when would they become ill?

A As there has only been one possible transmission of vCJD by a blood transfusion, it is impossible to say whether someone will develop vCJD or how long it would take.

Q Can people have a test to find out if they are going to get vCJD?

A As yet, there is no screening test for people concerned that they may develop vCJD

Q What about members of the family of someone who received blood?

A No special precautions are needed for family or other household members. There is no evidence that vCJD can be passed on between people by:

- Living in the same house
- Sharing utensils
- Kissing
- Sexual contact
- From mother to baby through childbirth or breastfeeding.

Q Do people who have received a transfusion after 1st January 1980 have to inform people who treat them in the future?

A At the moment there is no requirement for people to inform those who treat them that they received a transfusion since 1st January 1980

Q What is this test assessment facility?

A This facility will enable the UK to be able to test any blood test for vCJD may emerge over the next few years. Having this facility will ensure that a potential test can be quickly verified and validated, thus avoiding potential delays.

Q Should recipients of blood be worried?

A It is important to put this context into issue. No medical product can be 100% safe, but people must weigh up the balance of risks. In addition there has only been one possible case of vCJD being passed through blood, yet three million units of blood are issued to hospitals every year.

If someone needs a blood transfusion, then the risk to that person of not having a blood transfusion far outweighs any possible risk of contracting vCJD. This is a further precaution, in addition to existing safety measures, against the possible risk of vCJD. Recipients of blood who are still worried should contact NHS Direct on 0845 46 47.

Q Are the steps being taken enough?

A There remains uncertainty over the number of people who will develop vCJD from eating meat and whether vCJD can definitely be transmitted by blood. Blood transfusion saves and improves many thousands of patients' lives every year. There has to be a balance between what measures should be implemented and the sufficiency of the blood supply.

The measures in place or being implemented by the NBS are as a direct response to expert advice. These measures continue to be reviewed.

Q What other precautions are in place?

A In view of the uncertainty as to whether vCJD could be transmitted by blood or blood products, the UK Blood Services have taken a number of precautionary measures:

- Withdrawal and recall of any blood components, plasma derivatives or tissues obtained from any individual who later develops vCJD (December 1997).
- Import of plasma from the US for fractionation to manufacture plasma derivatives (announced May 1998, implemented October 1999).
- Leucodepletion of all blood components (announced July 1998, implemented Autumn 1999).
- Importation of clinical FFP from the US for patients born on or after 1st January 1996 (announced August 2002, to be implemented spring 2004).
- Promotion of appropriate use of blood and tissues and alternatives throughout the NHS.

Q How can people find out more about vCJD?

A More information about vCJD is available from the following organisations:

- National CJD Surveillance Unit at www.cjd.ed.ac.uk (0131 5372128)
- Department of Health at www.doh.gov.uk/cjd/index.htm (0207 2105025)
- CJD Support Network at www.cjdsupport.net (01630 673973973)
- MRC Prion Unit at www.st-marys.org.uk/specialist/prion/index_prion.htm (0207 8866883)
- Human BSE Foundation at www.hbsef.org.uk (0191 389 4157

Statistics

CJD AND TRANSFUSIONS

- **Total number of cases of vCJD in UK as of 31st December 2003: 145**
- **Number of reported cases to have received a blood transfusion: 9**
- **Number of reported cases who were blood donors: 17**
- **Number of these blood donors from whom components were issued: 15**
- **Number of components issued from these 15 donors: 55**
- **Number of identified recipients who received these blood components: 48**
- **Number of recipients still alive: 16 (14 in England and Wales, 2 in Scotland)**

NATIONAL BLOOD SERVICE

- **Approximately 800,000 patients receive a blood transfusion every year**
- **In 2002/03, the NBS issued the adult equivalent of 2,186,000 units of red cells**
- **In 2002/03, the NBS issued the adult equivalent of 398,000 units of frozen plasma products**
- **In 2002/03, the NBS collected 2,332,371 donations of blood**

BSE in America

This article, written by Prof John Collinge OBE, appeared in The Times newspaper 3 January 2004.

Watching Ann Veneman, US Agriculture Secretary, announce the first case of BSE in the United States at Christmas was profoundly disappointing, not simply because another country was blighted by this degenerative brain disease, but the presentation was all too familiar. That agriculture rather than health secretary sought to reassure about risk to human health, and that she said she would be feeding her own children beef, was as reminiscent of the early stages of the outbreak in Britain as it is scientifically inane. There are many lessons the Americans could learn from the British experience, and the following months will prove crucial.

The first, and most important, is that infection with the human form of so-called 'mad cow disease' may be more widespread than commonly assumed. To date around 140 people have died of vCJD (the human form of BSE). While each case is a tragedy, this relatively small number in public health terms has been interpreted to mean the human cost will be small. The largest number of reported cases was in 2000, leading to assumptions that vCJD has peaked and will now decline with a death toll of a couple of hundred. While everyone hopes that this proves correct, I must point out some uncomfortable facts that challenge such optimism.

We are used to infectious disease epidemics, such as influenza or foot and mouth disease, that arise and decline over weeks and months. However, we know from the disease kuru, transmitted amongst the Fore in Papua New Guinea during cannibalistic feasts with devastating effects, that human prion disease epidemics can span decades with incubation periods that can exceed forty years, with an average of around 12 years.

We also need to think about the effect of the so-called 'species barrier'. When prions from one species infect another species, the incubation periods seen are typically very much longer. Average incubation periods of BSE prions in humans of 30 years or more would not be surprising based on these sorts of comparisons.

By definition, those who have already got vCJD are those with the shortest incubation periods. These people do not seem to have been exposed to more BSE in their lives, suggesting they may be particularly vulnerable as a result of their genes, or the presence of unknown environmental factors. We know that our genes have a major bearing on incubation periods, but until we understand more about this, we cannot know if the current vCJD patients represent a first 'wave', with other peaks, perhaps much larger, to follow.

In short, while it would be marvellous if it were true, the science suggests much caution in concluding the human BSE problem is receding at what is, in the timescale of human prion diseases, this early stage.

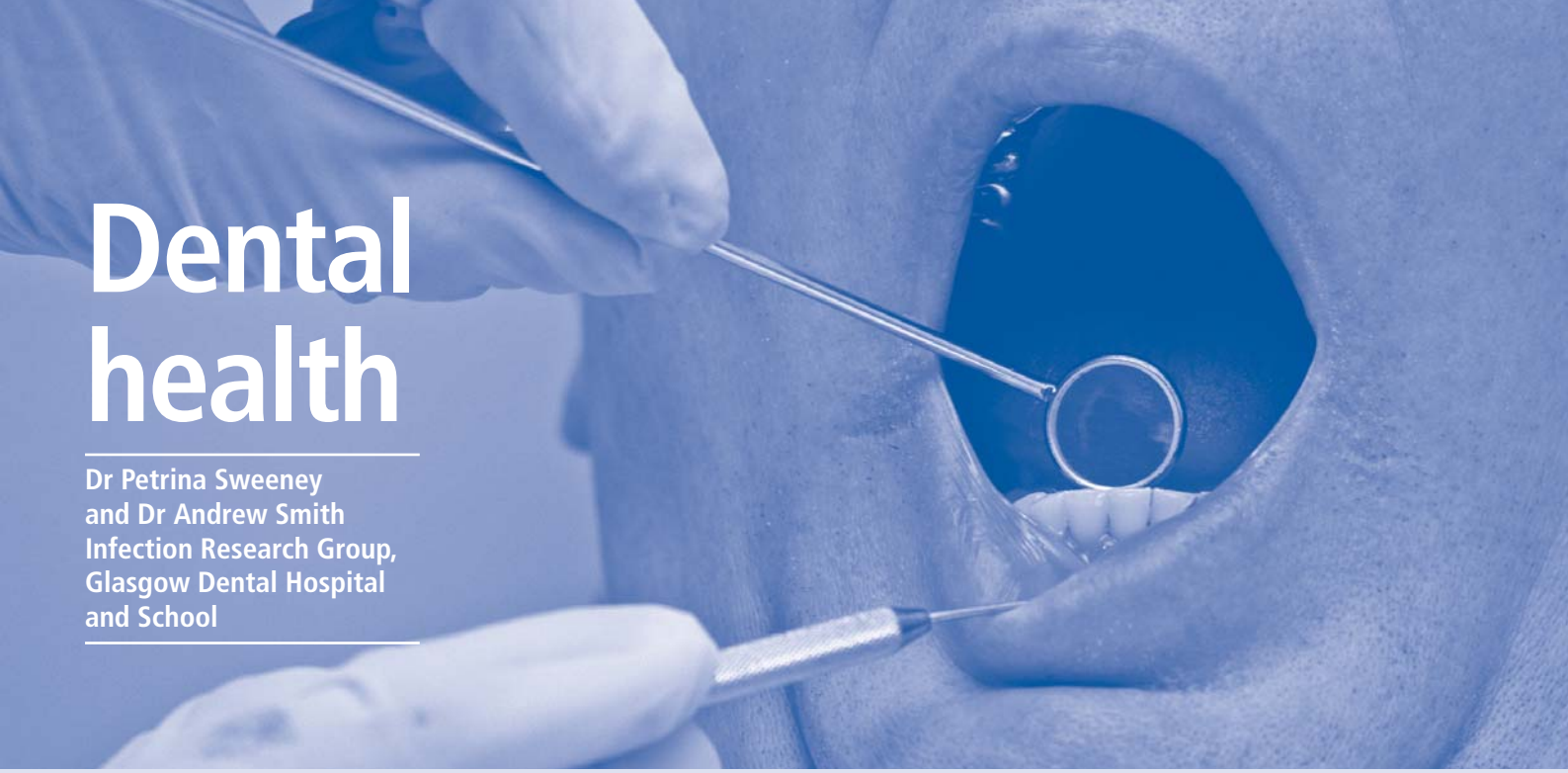
So has America taken this problem seriously enough? Secretary Veneman's later statement announced some sound measures to protect US consumers – notably bans on high risk cattle tissues in human food, on air-injection stunning of cattle and the use of mechanically recovered meat.

However, some of her comments resonated in the British ear: 'I will stress again that our food supply and the public health remain safe' and 'Our goal is to see trade resume as quickly as possible.' But one piece of scientific advice given repeatedly to the British Government with respect to the risks of BSE is that 'absence of evidence is not evidence of absence' and no doubt our US scientific colleagues will advise likewise.

What would reassure is proper surveillance of animals, using diagnostic tests at abattoirs and of fallen stock. It will be interesting to see if Veneman's scientific advisors agree that testing 20,000 cattle per year in the US (out of a total of more than 35 million slaughtered annually) is the 'very aggressive surveillance programme' that she described. If the US BSE case is due to contaminated feed, it is hard to imagine that there will not be others in other herds.

While agriculture ministers the world over will quite properly defend their important beef industries, it is to be hoped they learn the UK lesson. Openness about the uncertainties from the beginning is in the end less damaging to commerce than denial and misleading statements on risk that cannot be scientifically substantiated and that, if proved wrong, severely damage public confidence in Government (and science). In these post-9/11 times, restoring and maintaining public confidence in Government's management of risk on both sides of the Atlantic should be paramount.

Editor's note *Professor John Collinge is the clinical director of the specialist Prion Clinic at St Mary's Hospital which was set up to provide a clinical service for patients with or suspected to have any form of prion disease. We were very pleased to learn that John had been awarded the OBE in the New Year's Honour's list. We would like to send John our warmest congratulations for this well deserved recognition of his personal efforts and importance working for families affected by all strains of CJD.*



Dental health

Dr Petrina Sweeney
and Dr Andrew Smith
Infection Research Group,
Glasgow Dental Hospital
and School

- **A healthy mouth is important in maintaining quality of life for patients with CJD.**
- **A preventive approach is essential particularly if communication between patient and carers is difficult.**
- **Key workers assigned to a family should be aware of the importance of preventive dental advice as part of the overall care of such patients. Early intervention will minimise future oral disease and pain.**
- **Appropriate treatment planning for this group of patients is best performed by dentists with experience of patients with special needs.**
- **Current evidence suggests that normal social or routine clinical contact does not present a risk to healthcare workers, families or others, this includes dentists. There is no evidence of infectivity in saliva.**

DOH guidelines on oral health care

The four key points for an oral health plan for patients with CJD are:

1 **Early assessment of oral health**
Individual care plans should be drawn up following liaison with family and other health care workers for each case.

2 **Establish realistic methods of oral hygiene**

– Teeth should be cleaned at least twice dailyl, using fluoridated toothpaste and a small-headed toothbrush. If the patient is unable to rinse and spit, Corsodyl(r) gel (chlorhexidine gluconate) should be used in place of toothpaste. There are various aids available to help patients or their carers maintain a clean, comfortable oral environment and a dental hygienist can be particularly helpful delivering advice and support.

– Dentures should be assessed for fit and comfort as ill-fitting dentures can rub the mouth and cause discomfort and ulceration.

– Dentures (complete and partial) should be removed after every meal, rinsed in water to remove food debris and checked for sharp edges and cracks. They should be replaced in the mouth after the mouth has been checked for food debris and wiped or rinsed clean. At night, dentures should be cleaned with a toothbrush and left to soak in fresh tap water overnight.

3 **Establish a good diet**
Liase with dietician.

4 **Regular oral examination**
It is important that the mouth is checked regularly by appropriate dental team members. Many drugs can have adverse effects on the mouth leading to oral dryness, painful mouth ulcers and infections, such as thrush. The frequency of these examinations must be individually assessed.

The Department of Health has published guidance for healthcare workers on-line at www.dh.gov.uk/

Good news

Photo: Elagante Wedding

Sarah Shadbolt and Roger Tomkins were married in St John's Church, Hoveton in Norfolk, on Saturday 4 October 2003, having been together for five years. The church service was designed to reflect both their lives and lost loved ones. The reception was held in a local hotel and the next day a four-hour river cruise was organised for some 120 guests, including Gillian Turner and her husband David.

Sarah and Roger have been members of the CJD Support Network for four years both as full and co-opted members of the committee. Over this period they have given numerous talks on their experiences of CJD at seminars and conferences, and recently a talk to medical students at the University of East Anglia in Norwich.

They are always willing to talk to family members, professionals and the media about CJD and they operate the CJD helpline when Gillian is on holiday.

Sarah had been married to her husband Edward for 30 years and lived in Horning on the Norfolk Broads. Sadly Edward died of Classical CJD on the 19 December 1997.

Roger lived in Kent with his family. Roger sadly lost his daughter Clare, age 24, to vCJD on April 22 1998. Roger's wife Dawn contracted ovarian cancer during Clare's illness and sadly died 6 weeks after Clare on 9th June 1998. Roger and Dawn had been married for 30 years. Roger has an elder daughter who lives in Kent with her husband and two young daughters.

Sarah and Roger were introduced through a mutual friend, although Sarah had known Dawn for some years because she would frequent the craft shop in Horning, which Sarah and her husband Edward owned. Horning was where Roger and Dawn had kept their motor cruiser for many years.

Sarah and Roger now live by the river in Horning where they run their 80 berth Marina in the beautiful setting of the Norfolk Broads. They are now very happily married and enjoy a busy life together with their various activities and many good friends.

With Roger having survived bowel cancer during 2003 with the never-ending support of Sarah, they both feel that they are well equipped to focus on the real priorities of life.

CJD Support Network membership application

Becoming a member of the CJD Support Network 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 CJD Support Network newsletter.
☐ I would like to become a member but **not** receive the CJD Support Network newsletter.

There is no fixed subscription, but you may wish to make a small donation to cover the cost of production and postage of our newsletter.

☐£8 ☐£12 ☐£25 ☐£50 ☐Other .....

Please make cheques payable to **CJD Support Network**
However, if you are a carer and would appreciate free membership, please tick the box ☐

Name ..... Title .....

Address .....

..... Postcode .....

Telephone ..... Email .....

I am caring for someone with CJD: ☐ at home ☐ in residential care
I am: ☐ a concerned relative/friend ☐ former carer ☐ professional ☐ interested

MAR04

☐ I want all my donations to the CJD Support Network from 6 April 2000 to be Gift Aid until I notify you otherwise.
Use Gift Aid and you can make your donation worth more. For every pound you give to us, we get an extra 28 pence from the Inland Revenue.To qualify for Gift Aid, what you pay in UK income tax, capital gains tax or tax on interest from savings must at least equal the amount we will claim in the tax year. Please let us know if you move or if you no longer pay enough tax to cover the money we claim back from the Inland Revenue.

CJD SUPPORT NETWORK

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

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