ADVISORY COMMITTEE ON DANGEROUS PATHOGENS

vCJD position paper

Issue

This update paper:

- provides ACDP with an overview of the Department of Health's response to the public health challenge of TSEs in animals and humans
- sets out current issues
- identifies key issues going forward.

Background on the position up to the late 1990s, and on BSE and vCJD, is at **Annex A**.

Action

ACDP are asked to note:

- this update paper;
- the statement on prevalence recently published by the TSE Risk Assessment Sub-Group;
- appointment of Dr Roland Salmon as Chair of the TSE Risk Management Sub-Group;
- abolition of the CJD Incidents Panel at the end of March 2013.

SUMMARY
The primary route of variant Creutzfeldt-Jakob disease (vCJD) infection, from BSE/cattle to humans, appears to have been resolved through extensive meat hygiene and other controls. In addition a significant number of actions have been taken, based on available evidence and careful risk assessments, to minimise the potential for secondary (person to person) spread of abnormal prion infection. Nevertheless considerable scientific uncertainties remain, especially around the prevalence of infection in the wider population and the risk that this may pose for further transmission.

It is therefore important that measures should continue to:

- maintain barriers against the possibility of further primary spread;
- ensure proportionate actions to reduce potential risks of secondary infection via donations and transplants or surgery are in place and kept under regular expert review; and
- support funding for ongoing surveillance and research.

vCJD: RECENT HISTORY
In 1996, with strong epidemiological and laboratory evidence for a causal link between vCJD and BSE, the committee advising on all TSEs, the Spongiform Encephalopathy Advisory Committee (SEAC), concluded that the most likely explanation for the emergence of vCJD was that it had been transmitted to people who had eaten cattle meat, organs or tissue infected with BSE. In the late 1990s new research provided further evidence that the agent which causes BSE is the same as that which causes vCJD.

From May 1997 UK Governments continued to work to reduce risk, and following SEAC's advice 'beef on the bone' was banned from December 1997. This policy was reviewed and eventually lifted in December 1999. The beef export ban was lifted for all UK beef from August 1999, although there was resistance from France to this action, which did not lift the ban until October 2002.

In December 1997, the then Government set up the Phillips Inquiry into BSE and published its report in October 2000. The Government's response to the report was published in September 2001.

Approach to vCJD risk reduction
Following identification of the link between BSE and vCJD, successive UK Governments and UK Health Departments have followed a precautionary approach to reduce the risk of contracting vCJD from meat and meat products, and the risk of person to person (secondary) transmission of CJD through clinical interventions, such as blood/blood products and surgery.

Shortly after vCJD was first identified, the possibility of human-to-human transmission via blood was considered, and the Department implemented precautionary measures to reduce what was, at that time, a theoretical risk. The measures were tightened as evidence of transmission via blood began to emerge from animal studies. These measures included removing white blood cells from blood, and importing blood products from countries unaffected by vCJD. The first possible case of transfusion associated transmission in humans was identified in

late 2003. An important additional step, introduced in March 2004, was to exclude from blood donation all those who had themselves received a blood transfusion since January 1980 (a list of blood risk reduction measures is at **Annex B**).

Whilst there is to date no evidence that vCJD has been transmitted via surgical instruments, there is some evidence that sporadic CJD has been transmitted in certain very particular circumstances, such as neurosurgery. Therefore, the Advisory Committee on Dangerous Pathogens (ACDP) advised from the mid 1990s on the decontamination, quarantining and appropriate use of surgical equipment (including endoscopes), and on pre-surgical assessment of patients to indentify and act on those with, or at risk of, all forms of human prion disease.

Prions are difficult to remove from instruments and so high quality decontamination across surgical and dental practice is essential, and health Department work with the health care providers to drive up standards. Because of the difficulties in effectively removing prions from, particularly metal, surfaces it is important that safe, effective optimum methods of decontamination are used. NICE have provided specific advice on instrument decontamination in respect of neuro and ophthalmic surgery, which are associated with the greatest potential risk of human prion disease transmission via surgical instruments.

There is evidence from animal models of potential dental routes of transmission of infection. In April 2007 the Chief Dental Officer advised all dentists in the UK to use only single-use endodontic (root canal) reamers and files, and to ensure that the highest standards of decontamination are observed for all dental instruments.

Government Funding

In 2002, following the Phillips Inquiry, a no-fault payment scheme (the vCJD Trust) was established, operated by an independent Board of Trustees, for all vCJD patients and their families. Compensation for vCJD patients is paid in recognition of their wholly exceptional situation. The scheme has distributed nearly £40m to patients and their families.

The Government has an on-going commitment to provide resources for CJD surveillance and research. This currently includes annual funding of £2.2m for the National CJD Research and Surveillance Unit (NCJDRSU), as part of an £5.5m ring-fenced budget for research and surveillance funding (the only ring-fenced amount in the Department's Policy Research Programme (PRP) budget). The PRP currently funds projects on epidemiology/prevalence and decontamination of instuments. The Surveillance Unit's current funding contract is until March 2015.

The Department also funds a care package to ensure that patients with all forms of CJD have support in the community, and to provide for those elements of their care that cannot be readily supplied by local health and social services. Expenditure from the National CJD Care Fund from 2000-2010 was £3,216,362.

CURRENT ISSUES

Surveillance

Surveillance is the cornerstone of the Government's policy to monitor and control the spread of vCJD. The incidence of all forms of CJD is monitored in the UK by

the NCJDRSU. The Unit brings together a team of clinical neurologists, neuropathologists and scientists specialising in this group of conditions. Whilst the number of clinical cases of vCJD has fallen (there have been no new onsets in the UK for two years) the potential for long incubation periods for these conditions and lack of knowledge about the implications of prevalence of sub-clinical infection (see below) means that surveillance will need to continue in the coming years.

Prevalence of vCJD in the population

A major strand of the Department's funded work on vCJD has been devoted to the commissioning of prevalence studies, to try and better understand the number of people who may be infected but show no symptoms, so that appropriate evidence based and cost effective risk management strategies can be developed. Estimates of the number of people who may be incubating vCJD have been made based on modelling and vary widely.

The most recent prevalence study results are provided at **Annex C**, and a ACDP TSE Risk Assessment Sub-Group statement on the occurrence of vCJD and prevalence of infection in the UK population is at **Annex D**. This study tested 32,441 appendix samples, collected during surgery since 2000 on patients born between 1941 and 1985. Of these, 16 samples were judged to be "positive"[1]. This indicates a central prevalence estimate very close to 1 in 2,000 in the age cohort covered, with a 95% Confidence Interval running from approximately 1 in 3,500 to 1 in 1,250.

The Sub-Group concluded that the survey "provides the most reliable available indication of the prevalence of asymptomatic vCJD infection within the UK population[2]".

vCJD and Blood

A blood test for vCJD may be advantageous for both health protection and possibly prevalence study purposes. Unfortunately, there are no current tests suitable for use. A joint meeting of the TSE Risk Assessment Sub-Group and the Blood Services Prion Working Group will meet to review current test developments on 25 October 2012.

The potential for a blood test was heightened in December 2003, when the then Secretary of State made a statement to Parliament on the first possible case of vCJD possibly acquired through transfusion of whole blood. Following this other transfusion recipients identified to be at increased risk were been notified. This enables public health precautions to be taken to reduce possible onward transmissions. Currently there are 17 living patients identified to be at increased risk of vCJD as a result of receiving blood transfusion from donors who are known

[1] Of these 12 showed indicative patterns of staining with more than one antibody.

[2] Like the Hilton *et al* study, the new appendix survey used Immunohistochemisty (IHC) to screen samples. A large scale prospective survey of tonsil samples found no positives in 95,672 tested using a high throughput enzyme immunoassay technique (Frosh *et al*, 2004; Clewley *et al*, 2009). However re-testing of 9,672 samples using IHC revealed one specimen with a strongly positive follicle (de Marco *et al*, 2010). It is therefore considered that IHC provides the more reliable method of detection.

to have subsequently developed clincal vCJD. Four occurrences of possible transfusion associated transmission have now been identified: in three cases, the recipients developed clinical vCJD, in the other case the patient died of unrelated causes but evidence of abnormal prion was found at autopsy.

In light of the potential for blood transmissions in 2004, the Department commissioned an assessment of the vCJD risks from blood products used to treat bleeding disorders such as haemophilia. As a result, all people with haemophilia and blood disorders treated with UK derived products between 1980 and 2001 were designated 'at risk for public health purposes' in relation to vCJD. Haemophilia patients are now provided, wherever suitable, with a centrally funded synthetic product.

In February 2009 the first finding of abnormal prion protein at autopsy in a spleen sample of a 73 year old man with haemophilia (in the designated "at risk" group, but who died of causes unrelated to vCJD or any other neurological disorder) was announced. It is known that the patient had received product manufactured from a large pool known to have contained at least one donation from a person who later developed vCJD. It is not possible to tell with any certainty where the abnormal prion protein originated and a risk assessment concluded that:

- The most likely cause of infection for this patient was from the factor VIII product used to treat haemophilia.
- That it was more likely that the infection came from a product that was not made from a pool known to contain a donation from someone who later went on to die from vCJD because of the large overall number of product units the patient had received.

A further risk group are those who have had a large number of donor exposures through use of blood components. It has been agreed that a donor exposure limit of 300 will categorise an individual as at increased risk of vCJD, and that in principle that those affected should be identified and notified where practicable. However before such notification is performed, the HPA has been asked to consult with haematologists and other specialists caring for those likely to be notified, and appropriate patient representative groups, and prepare a report to the Chief Medical Officer on the potential impact of such an exercise on individuals and services

One potential method of reducing the blood transmission risk are prion filters which aim to remove potential infectivity from donated blood. The Advisory Committee on the Safety of Blood, Tissues and Organs is reviewing evidence of prion filter efficacy at the next meeting in December 2012.

Decontamination of Surgical and Dental instruments

ACDP has provided advice on the handling of instruments and devices in procedures on patients with known or suspected CJD/vCJD. Specific guidance is provided by NICE for neuro and ophthalmology surgery and the Chief Dental Officer on single-use endodontic (root canal) reamers and files.

The Code of Practice for health and adult social care on the prevention and control of infections and related guidance requires providers of healthcare to have a policy on decontamination of reusable medical devices and thus ensures that decontamination is part of the infection prevention and control programme. Management systems should be in place to ensure adequate supplies of instruments and to minimise the risk of transmission of infection to patients and staff. For example endoscopes and surgical instruments, are required to be individually identifiable, or identified to a set of which they are a consistent member, throughout the use and decontamination cycle in order to ensure subsequent traceability. Systems should also be implemented to enable the identification of service users on whom the medical devices have been used.

Treatment for vCJD

There are no effective treatments for vCJD. The only therapeutic agent recently used was Pentosan, which whilst permitting the survival of some vCJD patients for over 6 years provided limited, if any, therapeutic benefit.

Government Expert Advice on TSEs

The Government continues to be advised on CJD and other TSEs by the Advisory Group on Dangerous Pathogens, particularly through its TSE Risk Management and TSE Risk Assessment Sub-Groups, they provide advice on all matters related to TSE risk assessment and management. SEAC was abolished from March 2011 and the CJD Incidents Panel, which currently provides operational advice on action to handle specific incidents involving potential transmission of CJD between patients, will be wound up in March 2013.

From April 2013 the TSE Risk Management Sub-Group will take on all the "generic" TSE risk management functions currently performed by the Risk Management Sub Group and the CJD Incidents Panel. Responsibility for actions on individual CJD incidents are to be managed at the local level, in the same way as most other incidents that place patients at risk. Local teams will apply national guidance on handling such incidents published by ACDP. Generic guidance on patient notification processes following incidents is currently subject to work by a group, chaired by Paul Johnstone, DPH Yorks & Humber.

The vast majority of CJD related incidents involve sporadic or familial CJD. After abolition of the Panel, local incident teams would still be able to refer novel issues to the ACDP Risk Management Sub-Group where necessary. The Sub-Group would provide advice based on its own or co-opted expertise. Local teams would be asked to let PHE know where a CJD patient notification has been made to enable any appropriate enhanced surveillance of those notified.

THE FUTURE

We cannot be sure that the number of vCJD cases is tailing off, especially because of the potential length of the disease incubation period, which may be several decades, and other scientific uncertainties still surrounding the behaviour of the causative prion agent (such as the nature and pattern of infectivity, and the influence of genotype variations in the population). BSE and vCJD will continue to be of concern to health professionals and the public for the foreseeable future.

Issues

Over the next 12-18 months, the key CJD issues are likely to be:

- the need to maintain investment in CJD surveillance and research because of the long-term uncertainties;
- ensuring a balance is maintained in the cost/benefit of additional risk reduction measures including the potential use of additional blood safety precautions;
- provision of revised advice on decontamination science and technologies to ensure optimum levels of prion removal from instruments, and a continuing drive to raise decontamination standards in all sectors;
- appropriate definition and handling of populations defined as “at increased risk”, such as those exposed to multiple blood exposures, so that whilst the approach is precautionary the actions are measured and proportionate;
- maintaining public health input to FSA processes and assurance controls which impact on TSEs – with the apparent reduction of risk from BSE and diminution of public health skills within FSA, both DH and HPA will need to ensure continued engagement; and
- updating prevalence as results from current studies accumulate, and taking forward additional prevalence studies as resources permit.

ANNEX A

BACKGROUND – TSEs

Transmissible spongiform encephalopathies (TSEs), or prion diseases, are a group of very rare degenerative brain disorders characterised in clinical cases by the "spongy" appearance of the brain. All can have very long incubation periods and cause severe and irreversible damage to the central nervous system. There are so far no proven effective treatments for any TSE and they are inevitably fatal. They include the various forms of Creutzfeldt-Jakob disease (CJD) and other TSEs in humans, bovine spongiform encephalopathy (BSE) in cattle, scrapie in sheep and goats and chronic wasting disease (CWD) which affects deer in North America.

Studies suggest that TSEs are caused by abnormal prion protein, which when it occurs in a normal form is a harmless protein found in the body's cells. The harmless and infectious forms of the prion protein are nearly identical, but the infectious form takes on a different folded shape from the normal protein. Sporadic Creutzfeldt-Jakob disease (sCJD), first described in the 1920s, is the most well-known of the human TSEs, that affects about one in every one million people each year. Other human TSEs include kuru, fatal familial insomnia (FFI), and Gerstmann-Straussler-Scheinker disease (GSS). Kuru, identified in the Fore people of Papua New Guinea and associated with past ritualistic consumption of human tissues has now almost disappeared. FFI and GSS are extremely rare hereditary diseases, found in just a few families.

The range of human TSEs can occur three ways: sporadically; as hereditary diseases; or through transmission from infected individuals. Sporadic TSEs may develop because some of a person's normal prions spontaneously change into the infectious form of the protein and then alter the prions in other cells in a chain reaction. Inherited cases arise from a change, or mutation, in the prion protein gene that causes the prions to be shaped in an abnormal way. This genetic change may be transmitted to an individual's offspring. Transmission of TSEs from infected individuals is relatively rare.

TSEs cannot be transmitted through normal person-to-person contact or through the air. However, they may be transmitted through contact with infected tissue, body fluids, or contaminated medical instruments. Normal sterilization procedures such as boiling or irradiating materials do not prevent transmission of TSEs.

Symptoms of TSEs vary, but they commonly include personality changes, psychiatric problems such as depression, lack of coordination, and/or an unsteady gait. Patients also may experience involuntary jerking movements called myoclonus, unusual sensations, insomnia, confusion, or memory problems. In the later stages of the disease, patients have severe mental impairment and lose the ability to move or speak.

A new variety of CJD, called variant CJD (vCJD), was first identified when scientists at the UK National CJD Surveillance Unit first described it in 1996. The initial symptoms of vCJD are different from those of sCJD and the disorder

typically occurs in younger patients. Evidence suggests that primary vCJD may have resulted from human consumption of beef from BSE infected cattle.

TSEs in the UK

Bovine Spongiform Encephalopathy (BSE) has been the most highly publicised of the animal TSEs because of its links to variant Creutzfeldt–Jakob Disease (vCJD), in humans; and its estimated £38 billion cost to the UK economy. BSE is a progressive, lethal central nervous system disease of cattle, which exhibit changes in behaviour, loss of weight, and loss of coordination, and like all TSEs there is no effective treatment. The first case in UK cows was identified in 1985 and BSE was first recognised as a new disease in 1986. In 1992 the number of cases in cattle reached a peak of 36,680 and the annual number has fallen steadily since (there were only 8 cases identified in 2009). At the height of the epidemic in British cattle the incidence of BSE was almost 1,000 cases per week, and by 2012, more than 181,000 cases had been confirmed in British cattle.

When first identified BSE seemed similar to scrapie, a disease in sheep that had been around for 200 years with no apparent effect on human health. Few cattle seemed to be infected, and the possibility that it might be transmissible to humans was regarded by most scientists as remote. It is now generally agreed that the BSE epidemic in cattle was exacerbated by feeding rendered meat- and bone-meal (MBM) to cattle. Measures to control the BSE epidemic in cattle were first introduced in 1988 with the ban on feeding MBM and policy to slaughter BSE infected cattle, and further actions in response to BSE followed in the early 1990's.

Variant Creutzfeldt-Jakob disease

Variant CJD (vCJD), as described by the NCJDSU, is distinguishable from the classical (sporadic) form in a number of ways. It tends to affect younger people with an average (median) age of onset of around 26 years (median age at death 28 years). The predominant initial clinical symptom is of psychiatric or sensory problems, with brain changes shown as coordination problems, dementia and muscle twitching occurring later. The illness usually lasts between six months and two years before death (median 14 months). The definitive diagnosis of vCJD can only be confirmed by looking at brain tissue usually at post-mortem and requires the exclusion of sporadic and familial CJD. There have been 168 deaths from definite or probable cases of vCJD in the United Kingdom and an additional four people alive with the disease (**Annex E**). The peak year of deaths was 2000, since when numbers of cases have fallen. The graph at **Annex F** shows UK data for date onsets, diagnosis and deaths for all 176 UK vCJD cases 1994-2012.

ANNEX B

BLOOD RISK REDUCTION MEASURES to 2012.

Since the theoretical risk of vCJD transmission through blood was first identified as a possibility in 1996, a series of precautionary measures have been implemented to protect the blood supply and products made by fractionating plasma, including:

Applicable to all blood/blood products

- From December 1997, blood components, plasma products or tissues obtained from any individual who later develops vCJD, have been withdrawn/recalled to prevent their use;
- From October 1999, white blood cells (which may carry a risk of transmitting vCJD) have been reduced in all blood used for transfusion, a process known as leucodepletion or leucoreduction;
- Following the report of the first possible case of transmission of vCJD by blood transfusion in December 2003, individuals who had themselves received a transfusion of blood components since January 1980 were excluded from donating blood. This took effect from April 2004;
- In July 2004, this exclusion criterion for blood donation was extended to include two new groups, who had received transfusions of blood components since 1980:
    - Previously transfused platelet donors,
    - Donors who were unsure if they had previously had a blood transfusion. This now applies to donors who have been transfused anywhere in the world;
- In July 2005, the Department of Health announced further precautionary measures for around 100 individuals who donated blood to three people who later developed vCJD. The notified people have been asked not to donate blood, tissues or organs and to inform health care professionals so extra precautions can be taken when they have surgery or other invasive procedures;
- In November 2005, the Department of Health announced an extension of the July 2005 notification exercise. A further 50 people who had received blood from some of the 100 or so donors notified since July 2005 were traced and notified of their potential exposure to vCJD, and asked to take similar precautions.

Plasma

- Since 1999, plasma for the manufacture of fractionated plasma products, such as clotting factors, has been obtained from non-UK sources;
- Since 2004, fresh frozen plasma for treating babies and young children born on or after 1 January 1996 has been obtained from the USA;
- Fresh frozen plasma for treating babies and young children born on or after 1 January 1996 has been obtained from the USA, and from July 2005 its use was extended to all children up to the age of 16;

- The NHS has been instructed to purchase imported solvent detergent-treated pooled plasma for adult patients with thrombotic thrombocytopenic purpura;
- Synthetic (recombinant) clotting factor for treatment of haemophilia has been provided to the under-16s since 1998 and for all patients for whom it is suitable since 2005.

Platelets

- To reduce donor exposure, the Advisory Committee on the Safety of Blood, Tissues and Organs in 2009 reiterated its predecessor committee's advice on increasing the percentage of platelets collected by apheresis to at least 80%.

Cryoprecipitate (a special cold-treated plasma preparation)

- Cryoprecipitate produced from methylene blue treated-plasma imported from the USA is being implemented for children up to the age of 16. The Advisory Committee on the Safety of Blood, Tissues and Organs is reviewing use of cryoprecipitate in older patients.

All of these recommendations were recommended or endorsed by the Advisory Committee on the Safety of Blood, Tissues and Organs (which first met in January 2008), or its predecessor committees.

Additionally, considerable effort is being extended to promote appropriate use of blood throughout the NHS, to target blood use to where it is clinically essential. This work has already achieved notable successes, especially in reducing the use of blood in surgery.

Annex C

HPR 32 !2

Summary results of the second national survey of abnormal prion prevalence in archived appendix specimens

In April 2008, the Spongiform Encephalopathy Advisory Committee (SEAC) considered available prevalence data for variant Creutzfeldt-Jakob Disease (vCJD) in the British population and advised that a second appendix survey, using the same approach as a previous appendix tissue survey [1] on samples from the 1941 to 1985 birth cohort, be undertaken to further refine the estimate for the prevalence of subclinical infection [2]. The second unlinked anonymous survey of the prevalence of abnormal prion protein in archived appendix tissues has now been completed and this summary provides an update to the interim results published in September 2011 [3,4].

The survey examined appendices by immunohistochemistry from operations conducted between 2000 and 2012 and collected from 41 hospitals throughout England. Abnormal prion accumulation was detected within the follicular dendritic cells of 16 appendices out of 32,441 suitable samples examined. None of the positive appendices have come from the 176 known vCJD cases in the UK. In line with the interim findings, the final overall prevalence estimate, 493 per million (95% Confidence Interval (CI): 282 to 801 per million), remained statistically consistent with results from the earlier appendix survey (237 per million, 95%CI 49 to 692 per million) which examined samples from operations performed between 1995 and 1999 [1]. The prevalence estimates by birth cohort were 733 per million (95% CI: 269 to 1596 per million) in those born between 1941 and 1960 and 412 per million (95% CI: 198 to 758 per million) in those born between 1961 and 1985: these results were also in line with the interim findings [3,4].

The survey was conducted by a collaboration of the HPA, the Department of Neurodegenerative Diseases at the UCL Institute of Neurology, the Animal Health and Veterinary Laboratories Agency, the National Creutzfeldt-Jakob Disease Research and Surveillance Unit, the Histopathology Department of Derriford Hospital in Plymouth, and the MRC Prion Unit.

The final survey results have been considered by the Transmissible Spongiform Encephalopathies Risk Assessment Sub-Group of the Advisory Committee on Dangerous Pathogens, the successor to SEAC [5]. In summary, the estimated prevalence range largely overlaps that from the first survey, but *Health Protection Report* Vol 6 No. 32 - 10 August 2012 is narrower with a higher central estimate (around 1 in 2000 compared with around 1 in 4000). The new survey also demonstrates the presence of prion protein across a wider birth cohort than previously.

The hypothesis that the prevalence of abnormal prions found in both appendix surveys to date is linked to the epidemic of BSE in cattle in Britain can be tested directly by studying further appendix samples archived prior to the BSE outbreak

and samples from those born in 1996 or later by which time measures had been put in place to protect the food chain [5].

References

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Available at: http://www.dh.gov.uk/ab/ACDP/TSEguidance/DH_125868.

Annex D

https://www.wp.dh.gov.uk/transparency/files/2012/08/ACDP-statement-vCJD-occurrence-and-prevalence-Jul-2012.pdf

ANNEX E

Deaths from definite and probable cases of Creutzfeldt-Jakob Disease in the UK 1990-2012 (By Calendar Year)

REFERRALS OF SUSPECT CJD		DEATHS OF DEFINITE AND PROBABLE CJD						
Year	Referrals	Year	Sporadic	Iatrogenic	Familial	GSS	vCJD	**Total Deaths**
1990	[53]	1990	28	5	0	0	-	**33**
1991	75	1991	32	1	3	0	-	**36**
1992	96	1992	45	2	5	1	-	**53**
1993	79	1993	36	4	5	2	-	**47**
1994	119	1994	54	1	5	3	-	**63**
1995	87	1995	35	4	2	3	3	**47**
1996	133	1996	40	4	2	4	10	**60**
1997	162	1997	60	6	4	2	10	**82**
1998	154	1998	64	3	3	2	18	**90**
1999	170	1999	62	6	2	0	15	**85**
2000	178	2000	50	1	2	1	28	**82**
2001	179	2001	58	4	4	2	20	**88**
2002	163	2002	72	0	4	1	17	**94**
2003	162	2003	79	5	4	2	18	**108**
2004	114	2004	51	2	4	2	9	**68**
2005	124	2005	67	4	8	5	5	**89**
2006	111	2006	69	1	6	3	5	**84**
2007	115	2007	64	2	9	1	5	**81**
2008	147	2008	88	5	2	3	2	**100**
2009	145	2009	79	2	3	5	3	**92**
2010	150	2010	85	3	6	1	3	**98**
2011	154	2011	90	3	11	3	5	**112**
2012	72	2012	45	1	5	2	0	**68**
Total	**2967**	**Total Deaths**	**1368**	**69**	**99**	**48**	**176**	**1760**

As at 1st October 2012

Summary of vCJD case

Deaths

Deaths from definite vCJD (confirmed):	122
Deaths from probable vCJD (without neuropathological confirmation):	54
Deaths from probable vCJD (neuropathological confirmation pending):	0
Number of deaths from definite or probable vCJD (as above):	176

Alive

Number of definite/probable vCJD cases still alive:	0
Total number of definite or probable vCJD (dead and alive):	**176**

ANNEX F

UK vCJD cases 1994-2012