



CENTAUR GLOBAL NETWORK

2011-06-16-110 Prion disease update 2011 (06)  
To: (06) Transmissible spongiform encephalopathies  
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PRION DISEASE UPDATE 2011 (06)  
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A ProMED-mail post  
<<http://www.promedmail.org>>

In this update:

- [1] UK: National CJD Surveillance Unit - monthly statistics as of Mon 6 Jun 2011 - no new vCJD cases
- [2] France: Institut de Veille Sanitaire - monthly statistics as of Thu 26 May 2011 - no new vCJD cases
- [3] USA: National Prion Disease Pathology Surveillance Center - cumulative case numbers for 2011 up to 28 Feb 2011 - no vCJD cases [not updated since 28 Feb 2011]
- [4] TSE overview
- [5] Switzerland: 2nd case of BSE

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[1] UK: National CJD Surveillance Unit - monthly statistics as of Mon 6 Jun 2011 - no new vCJD cases  
Date: Mon 6 Jun 2011  
Source: UK National CJD Surveillance Unit, monthly statistics [edited]  
<<http://www.cjd.ed.ac.uk/figures.htm>>

The number of deaths due to definite or probable vCJD as of 6 Jun 2011 remains 171. A total of 4 definite/probable patients remain alive, so the total number of definite or probable vCJD cases remains 175.

The overall picture remains consistent with the view that the vCJD outbreak in the UK is in decline, albeit now with a pronounced tail.

The 1st cases were observed in 1995, and the peak number of deaths was 28 in the year 2000, followed by 20 in 2001, 17 in 2002, 18 in 2003, 9 in 2004, 5 in 2005, 5 in 2006, 5 in 2007, 1 in 2008, 3 in 2009, 3 in 2010, and one so far in 2011.

Totals for all types of CJD cases in the UK in the year 2011

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During 2011 so far [as of 6 Jun 2011], there have been 64 referrals, 26 fatal cases of sporadic CJD, one case of GSS, 3 cases of familial CJD, one case of vCJD, and none of iatrogenic CJD.

Since records began in 1990, there have been 27 897 referrals, 1237 fatal cases of sporadic CJD, 171 cases of vCJD, 85 cases of familial CJD, 65 cases of iatrogenic CJD, and 44 cases of GSS.

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[2] France: Institut de Veille Sanitaire - monthly statistics as of Thu 26 May 2011 - no new vCJD cases  
Date: Thu 26 May 2011  
Source: IVS - Maladie de Creutzfeldt-Jakob et maladies apparentees [in French, trans. & summ. Mod.CP, edited] <[http://www.invs.sante.fr/display/?doc=publications/mcj/donnees\\_mcj.html](http://www.invs.sante.fr/display/?doc=publications/mcj/donnees_mcj.html)>

During the 1st 5 months of 2011, there were 672 referrals, 18 confirmed fatal cases of sporadic CJD, one of familial CJD, and none of iatrogenic CJD, or vCJD.



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A total of 25 cases of confirmed or probable vCJD have been recorded in France since records began in 1992. There was one case in 1996, one in 2000, one in 2001, 3 in 2002, 2 in 2004, 6 in 2005, 6 in 2006, 3 in 2007, 2 in 2009, and none in 2010 and 2011.

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[3] USA: National Prion Disease Pathology Surveillance Center - cumulative case numbers for 2011 up to 28 Feb 2011 - no vCJD cases [not updated since 28 Feb 2011]

Date: Mon 28 Feb 2011

Source: US National Prion Disease Pathology Surveillance Center [edited]

<<http://www.cjdsurveillance.com/pdf/case-table.pdf>>

Cumulative data 1 Jan to 28 Feb 2011, the 12 months of 2010, and overall since 1996 [unchanged since previous update]

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During the 1st 2 months of 2011, there have been 52 referrals, 28 of whom were classified as prion disease cases, comprising 10 cases of sporadic CJD, 3 of familial CJD, and none of iatrogenic CJD or vCJD.

During the 12 months of 2010, there were 403 referrals, of whom 251 were classified as prion disease cases, comprising 208 cases of sporadic CJD, 40 of familial CJD, and none of iatrogenic or vCJD.

Overall, 3952 referrals have been examined since screening began in 1996 or thereabouts, with diagnoses pending in 13 cases or inconclusive in 18 cases, a measure of the difficulty in achieving unequivocal diagnoses.

During this 16 year period, a total of 2329 prion disease cases have been screened, a figure which includes 18 (15 during 2010) cases with type determination pending, but in which a diagnosis of vCJD has been excluded. Overall, there were 1965 cases of sporadic CJD, 338 cases of familial CJD, 5 cases of iatrogenic CJD, and 3 cases of vCJD. The 3 cases of vCJD recorded in the USA have been attributed to infection occurring in the United Kingdom in 2 cases, and in Saudi Arabia in the other.

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[4] TSE overview

Date: Wed 19 Jan 2011

Source: EFSA (European Food Safety Authority) Journal 2011; 9(1):1945 [edited]

<<http://www.efsa.europa.eu/de/efsajournal/doc/1945.pdf>>

[Ref: EFSA Panel on Biological Hazards (BIOHAZ): Joint Scientific opinion on any possible epidemiological or molecular association between TSEs in animals and humans. EFSA Journal 2011; 9(1): 1945.  
doi:10.2903/j.efsa.2011.1945]

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Abstract

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The existing scientific evidence that links animal and human TSEs [transmissible spongiform encephalopathies] is reviewed and discussed.



The challenges involved in identifying TSEs as zoonoses are described and the example of the process that led to the establishment of a link between bovine spongiform encephalopathy (BSE) and variant Creutzfeldt-Jakob disease (vCJD) is reviewed. The strain diversity of animal and human TSE agents and the factors influencing the capacity of TSE agents to cross the species transmission barrier are also discussed. The scientific opinion critically assesses the tools and methodologies currently available to study and evaluate the possible association of animal and human TSEs, focussing on epidemiological and laboratory methods. The available scientific evidence on classical BSE, typical BSE (H-type and L-type), classical scrapie, atypical scrapie, chronic wasting disease (CWD), transmissible mink encephalopathy (TME), and human TSEs is reviewed.

The conclusions state that, at present, the only TSE agent demonstrated to be zoonotic is the classical BSE agent. Active screening has allowed the identification of 3 new forms of animal TSEs (H-type atypical BSE, L-type atypical BSE, and atypical scrapie), but the information obtained has major limitations due to the unknown sensitivity of the current monitoring system for these TSEs. There is no epidemiological evidence to suggest that classical scrapie is zoonotic. The epidemiological data are too limited to conclude whether the atypical scrapie agent has a zoonotic potential. Transmission experiments to human PrP transgenic mice or primates suggest that some TSE agents other than the classical BSE agent in cattle (namely L-type atypical BSE, classical BSE in sheep, TME, CWD agents) might have zoonotic potential and indicate that that of the L-type atypical BSE agent appears similar or even higher than that of the classical BSE agent. A single study reported efficient transmission of a natural sheep classical scrapie isolate to primates.

#### Commentary

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Following to a request from the European Commission, the Panel on Biological Hazards (BIOHAZ) and the European Centre for Disease Prevention and Control (ECDC) were asked to deliver a scientific opinion on any possible epidemiological or molecular association between transmissible spongiform encephalopathies (TSEs) in animals and humans. The opinion reviews and discusses the existing scientific evidence that links animal and human TSEs currently known.

The opinion first considers the definition of zoonoses and the principles for the identification of zoonotic diseases, which can be based on evidence gathered from both epidemiological and laboratory studies. The opinion describes the challenges involved in identifying TSEs as zoonoses, due to the specific characteristics of TSE infections/diseases, such as the nature of TSE agents, the occurrence of animal and human TSEs, and the type of monitoring applied, the long incubation period of TSEs etc. The example of the process that led to establishing a link between bovine spongiform encephalopathy (BSE) and variant Creutzfeldt-Jakob disease (vCJD) is reviewed. The epidemiological and laboratory criteria that can be used to investigate such a link are described in detail, since those criteria might be useful for the identification of links between other animal and human TSEs.

The opinion discusses the strain diversity of the TSE agents described in sheep, goats, cattle, cervids, and humans, based on the current knowledge, which highlights that multiple TSE agents exist in each species. The factors influencing the capacity of TSE agents to cross the species transmission barrier are then considered in detail, including the variability in host and donor PrP gene and protein, the TSE strain type involved and its interaction with the host PrP, and the route of infection.

The opinion critically assesses the tools and methodologies currently available to study and evaluate the possible association between animal and human TSEs. The use of epidemiology is discussed for TSEs in both animals and humans, and the possibility to compare the 2 sources of information is presented as a possible method to study the possible links.

Both in vivo and in vitro laboratory methods are considered and discussed, including neuropathology, transmission experiments involving different animal models (wild type and transgenic mice, primates and other species), biochemical methods, cell-free conversion assays, protein misfolding cyclic amplification (PMCA), and cell culture assays. Characteristics, advantages, and disadvantages of the different methods are reviewed, including the opportunity to collate data from different types of experiments for the study of potential associations between animal and human TSEs.



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The opinion then reviews the scientific evidence currently available for the different animal and human TSEs, including classical BSE, atypical BSE (H-type and L-type), classical scrapie, atypical scrapie, chronic wasting disease (CWD), transmissible mink encephalopathy (TME), and human TSEs. In particular, the following aspects are systematically discussed for each TSE agent: epidemiology, pathogenesis, and in vivo and in vitro transmission experiments.

The opinion concludes that, at present, the only TSE agent demonstrated to be zoonotic is the classical BSE agent. With regard to human TSEs, detected cases of sporadic CJD are randomly distributed in time and geographical location. These observations have been interpreted as a supportive argument that sporadic CJD is not environmentally acquired. However, the epidemiological evidence in relation to sporadic CJD cannot be regarded as definitive, and the possibility that a small proportion of cases are zoonotic cannot be excluded.

It also concludes that a series of uncertainties in relation to the epidemiological patterns of animal and human TSEs indicate that even a rough comparison of the present epidemiological patterns of human and animal TSEs other than classical BSE is unlikely to be informative.

Because of these uncertainties, it is an imperative to continue to carry out systematic surveillance of human TSE diseases, and to continue and improve the surveillance of animal TSE diseases.

The opinion highlights that the active screening has allowed the identification of 3 new forms of animal TSEs (L-type atypical BSE, H-type atypical BSE, and atypical scrapie), but that the information obtained has major limitations due to the unknown sensitivity of the current monitoring system for these TSEs.

There is no epidemiological evidence to suggest that classical scrapie is zoonotic. The epidemiological data are too limited to conclude whether the atypical scrapie agent has a zoonotic potential.

Transmission experiments to human PrP transgenic mice suggest that some TSE agents other than the classical BSE agent in cattle (namely L-type atypical BSE and classical BSE in sheep agents) might have zoonotic potential, whereas for other agents there is no evidence provided of a zoonotic potential (H-type atypical BSE and CWD), or no published studies are available (classical and atypical scrapie). In addition, transmission experiments to primates suggest that some TSE agents other than the classical BSE agent in cattle (namely L-type atypical BSE, classical BSE in sheep, TME, CWD agents) might have zoonotic potential. In particular, primates are highly permissive to L-type atypical BSE, even by the oral route.

The opinion emphasizes that laboratory transmission experiments indicate that the L-type atypical BSE agent has a significant zoonotic potential, which appears similar or even higher than that of the classical BSE agent. While transmission data for evaluating the zoonotic potential of classical scrapie in primates and human PrP transgenic mice are extremely limited or not yet available, a single study reported efficient transmission of a natural sheep classical scrapie isolate to primates.

The opinion concludes that human PrP transgenic mice and primates are currently the most relevant models for investigating the human transmission barrier, but the extent to which such models are informative for measuring the zoonotic potential of an animal TSE under field exposure conditions is unknown. It is unpredictable whether a TSE agent will transmit to a new host, and if the transmission principally occurs, what the transmission rate will be.

Based on the results obtained with in vitro conversion assays, the opinion concludes that there is probably no absolute molecular barrier to transmission of TSE agents between mammalian species. Results also suggest that these assays may be developed as a tool for quantifying the transmission barriers between species for different TSE agent strains; however, there is no means at the moment to transpose in vitro results into the likelihood of in vivo interspecies transmission.

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[ProMED-mail thanks Terry S Singeltary Sr for drawing attention to this comprehensive document which provides a current evaluation of experimental work designed to explore the zoonotic potential of the various recently recognised TSEs of domestic and other animals.

It is concluded that at present the only TSE agent demonstrated to be zoonotic is the classical BSE agent. Nor can it be entirely excluded at the present time that a small proportion of cases of sporadic CJD may be environmentally acquired. - Mod.CP]

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[5] Switzerland: 2nd case of BSE

Date: Tue 24 May 2011

Source: Swiss Confederation, press release [in German, trans. & summ., Mod.AS, edited]

<<http://www.news.admin.ch/message/index.html?lang=de&msg-id=39298>>

A case of BSE in an old cow in the Canton of Bern

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The animal was born in 1995 -- that is still well before the total ban on feeding meat and bone meal to farm animals in 2001. The current case confirms that cases of BSE in older cattle must still be expected occasionally. There are still animals alive from the time before the total ban on feeding meat and bone meal (MBM) which came into force in 2001. That this case emerges only weeks after another case of BSE in the canton of St Gallen is pure coincidence. The case in St Gallen was a so-called atypical BSE case, which can occur sporadically and spontaneously, and most likely is not due to meat and bone meal feed.

The latest case concerns an old cow in the canton of Bern that had been infected before the total MBM feed ban in 2001. The cases have therefore nothing to do with each other.

The latest case shows that the monitoring function measures and the MBM feed ban for cattle continues to be important. Meat and bone meal from cattle must not be included in the animal food chain. For the safety of the people, since 1990 all potentially infectious parts of cattle, such as brains and spinal cords have been removed as SRM [specified risk material] from the food chain. This continues to be operational.

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[see also:

Prion disease update 2011 (05) 20110505.1393 Prion disease update 2011 (04) 20110406.1066 Prion disease update 2011 (03) 20110309.0764 Prion disease update 2011 (02) 20110211.0473 Prion disease update 2011 (01): correction 20110112.0140 Prion disease update 2011 (01) 20110110.0119 2010

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Prion disease update 2010 (11) 20101206.4364 Prion disease update 2010 (10) 20101105.4008 Prion disease update 2010 (09) 20101006.3622 Prion disease update 2010 (08) 20100911.3285 Prion disease update 2010 (07) 20100809.2720 Prion disease update 2010 (06) 20100706.2248 Prion disease update 2010 (05) 20100507.1488 Prion disease update 2010 (04) 20100405.1091 Prion disease update 2010 (03) 20100304.0709 Prion disease update 2010 (02) 20100205.0386 Prion disease update 2010 20100107.0076] .....