Elsevier Editorial System(tm) for The Lancet Infectious Diseases

Manuscript Draft

Manuscript Number:

Title: HUMAN and ANIMAL TSE Classifications i.e. mad cow disease and the UKBSEnvCJD only theory

Article Type: Personal View

Corresponding Author: Mr. Terry S. Singeltary,

Corresponding Author's Institution: na

First Author: Terry S Singeltary, none

Order of Authors: Terry S Singeltary, none; Terry S. Singeltary

Abstract: TSEs have been rampant in the USA for decades in many species, and they all have been rendered and fed back to animals for human/animal consumption. I propose that the current diagnostic criteria for human TSEs only enhances and helps the spreading of human TSE from the continued belief of the UKBSEnvCJD only theory in 2007.

HUMAN and ANIMAL TSE Classifications i.e. mad cow disease and the UKBSEnvCJD only theory August 2007

August 2007

HUMAN and ANIMAL TSE Classifications i.e. mad cow disease and the UKBSEnvCJD only theory

TSEs have been rampant in the USA for decades in many species, and they all have been rendered and fed back to animals for human/animal consumption. I propose that the current diagnostic criteria for human TSEs only enhances and helps the spreading of human TSE from the continued belief of the UKBSEnvCJD only theory in 2007.

With all the science to date refuting it, to continue to validate this myth, will only spread this TSE agent through a multitude of potential routes and sources i.e. consumption, surgical, blood, medical, cosmetics etc. I propose as with Aguzzi, Asante, Collinge, Caughey, Deslys, Dormont, Gibbs, Ironside, Manuelidis,
Marsh, et al and many more, that the world of TSE Transmissible Spongiform Encephalopathy is far from an exact science, but there is enough proven science to date that this myth should be put to rest once and for all, and that we move forward with a new classification for human and animal TSE that would properly identify the infected species, the source species, and then the route.

This would further have to be broken down to strain of species and then the route of transmission would further have to be broken down. Accumulation and Transmission are key to the threshold from sub-clinical to clinical disease, and key to all this, is to stop the amplification and transmission of this agent, the spreading of, no matter what strain. In my opinion, to continue with this myth that the U.K. strain of BSE (one strain TSE in cows), and the nv/v CJD (one strain TSE humans) and that all the rest of human TSE are just one single strain i.e. sporadic CJD (when to date there are 6 different phenotypes of sCJD, and growing per Gambetti et al), and that no other animal TSE transmits to humans, to continue with this masquerade will only continue to spread, expose, and kill, who knows how many more in the years and decades to come. ONE was enough for me, My Mom, hvCJD i.e. Heidenhain Variant CJD, DOD 12/14/97 confirmed, which is nothing more than another mans name added to CJD, like CJD itself, Jakob and Creutzfeldt, or Gerstmann-Straussler-Scheinker syndrome, just another CJD or human TSE, named after another human.

WE are only kidding ourselves with the current diagnostic criteria for human and animal TSE, especially differentiating between the nvCJD vs the sporadic CJD strains and then the GSS strains and also the FFI fatal familial insomnia strains or the ones that mimics one or the other of those TSE? Tissue infectivity and strain typing of the many variants

of the human and animal TSEs are paramount in all variants of all TSE. There must be a proper classification that will differentiate between all these human TSE in order to do this. With the CDI and other more sensitive testing coming about, I only hope that my proposal will some day be taken seriously. ...

Terry S. Singeltary Sr.
P.O. Box 42
Bacliff, Texas USA 77518
flounder9@verizon.net

SOURCE

Research Project: Transmission, Differentiation, and Pathobiology of Transmissible Spongiform Encephalopathies
Location: Virus and Prion Diseases of Livestock

Title: Pathobiology and diagnosis of animal transmissible spongiform encephalopathies: current knowledge, research gaps, and opportunities

Authors

Kehrli, Marcus
O`rourke, Katherine
Hamir, Amirali
Richt, Juergen
Nicholson, Eric
Silva, Christopher
Edelman, Daniel - FOOD AND DRUG ADMINISTRAT
Gay, Cyril

Submitted to: Government Publication/Report
Publication Type: Government Publication
Publication Acceptance Date: May 1, 2007
Publication Date: July 1, 2007

Citation: Kehrli, Jr., M.E., O'Rourke, K.I., Hamir, A.N., Richt, J.A., Nicholson, E.M., Silva, C.J., Edelman, D., Gay, C.G. 2007.

Pathobiology and diagnosis of animal transmissible spongiform encephalopathies: current knowledge, research gaps, and opportunities [government white paper]. Beltsville, MD: Interagency Working Group on Prion Science, Subcommittee on Pathobiology and Diagnostics. USDA, Agriculture Research Service. 33 p.

Technical Abstract:

Transmissible spongiform encephalopathies (TSEs) are fatal neurologic diseases that can affect several animal species and human beings. There are four animal TSE agents found in the United States: scrapie of sheep and goats; chronic wasting disease (CWD) of deer, elk, and moose; transmissible mink encephalopathy (TME) and bovine spongiform encephalopathy (BSE). Although the animal TSEs do not cause major death losses among US livestock populations, they are important because of international trade issues. The experience of the United Kingdom and Europe in dealing with the vast majority of the world's BSE cases, serves as a reminder of the need for continuing vigilance in monitoring risks for public health and research to answer remaining questions around the pathogenesis and transmission of these diseases. There remain questions on 1) cross-species transmissibility of TSEs in livestock and wildlife; 2) the pathobiology of TSEs in natural and secondary hosts; pathogenesis and transmission of CWD; and 4) pathogenesis and ante mortem detection of typical and atypical BSE. Our understanding of the pathogenesis and transmission of these diseases continues to evolve as ongoing, global TSE research efforts focus on defining tissue sites of abnormal prion accumulation, routes of infection, methods of strain differentiation, genetics of susceptibility and ante-mortem diagnostics. In this paper, a Subcommittee on Pathobiology and Diagnostics of TSEs for an Interagency Working Group on Prion Science summarizes the science of animal TSEs in order to identify knowledge gaps for the purpose of prioritizing animal prion research needs. Because of substantial losses involving international trade and potential risk for interspecies transmission to susceptible livestock and possibly humans, the presence of BSE, CWD, scrapie and TME in the United States presents a liability to U.S. domestic and alternative livestock industries. In addition, the proven risk of BSE to agriculture and public health from subclinical or clinically sick animals requires science-based surveillance for any silent, unrecognized epizootic expansions of these diseases in populations of animals that could either directly or indirectly affect food animals. CWD is an example of an uncontrolled expanding epidemic that threatens not only cervids but possibly other livestock. CWD also has elicited public health surveillance programs to monitor for scientific evidence of a prion disease in humans that consume venison. Therefore, some of the research needs are precautionary, but the risks to animal and human health from being caught unaware are high. Efforts

are being made by both federal and state regulatory agencies to eradicate scrapie and CWD, and to determine the prevalence of BSE. The effectiveness of these programs will depend heavily on having accurate information about the nature of these diseases, not only in the original hosts, but also in other species that may be in contact with infected animals.

http://arsserv0.tamu.edu/research/publications/Publications.htm?seq_no_115=212488

USA MAD COW STRAIN MORE VIRULENT TO HUMANS THAN UK STRAIN

18 January 2007 - Draft minutes of the SEAC 95 meeting (426 KB) held on 7 December 2006 are now available.

snip...

64. A member noted that at the recent Neuroprion meeting, a study was presented showing that in transgenic mice BSE passaged in sheep may be more virulent and infectious to a wider range of species than bovine derived BSE.

Other work presented suggested that BSE and bovine amyloidotic spongiform encephalopathy (BASE) MAY BE RELATED. A mutation had been identified in the prion protein gene in an AMERICAN BASE CASE THAT WAS SIMILAR IN NATURE TO A
MUTATION FOUND IN CASES OF SPORADIC CJD.

snip...

http://www.seac.gov.uk/minutes/95.pdf

3:30 Transmission of the Italian Atypical BSE (BASE) in Humanized Mouse

Models Qingzhong Kong, Ph.D., Assistant Professor, Pathology, Case Western Reserve
University

Bovine Amyloid Spongiform Encephalopathy (BASE) is an atypical BSE strain discovered recently in Italy, and similar or different atypical BSE cases were also reported in other countries. The infectivity and phenotypes of these atypical BSE strains in humans are unknown. In collaboration with Pierluigi Gambetti, as well as Maria Caramelli and her co-workers, we have inoculated transgenic mice expressing human prion protein with brain homogenates from BASE or BSE infected cattle. Our data shows that about half

of the BASE-inoculated mice became infected with an average incubation time of about 19 months; in contrast, none of the BSE-inoculated mice appear to be infected after more than 2 years.

***These results indicate that BASE is transmissible to humans and suggest that BASE is more virulent than
classical BSE in humans.***

6:30 Close of Day One

http://www.healthtech.com/2007/tse/day1.asp

There is a growing number of human CJD cases, and they were presented last week in San Francisco by Luigi Gambatti(?) from his CJD surveillance collection.

He estimates that it may be up to 14 or 15 persons which display selectively SPRPSC and practically no detected RPRPSC proteins.

http://www.fda.gov/ohrms/dockets/ac/06/transcripts/1006-4240t1.htm

http://www.fda.gov/ohrms/dockets/ac/06/transcripts/2006-4240t1.pdf

If, on the other hand, atypical BSE continues to occur as typical BSE disappears, this would be a strong indication that it is indeed sporadic, and if in addition at least 1 form of what is presently considered as sporadic CJD (such as the type 2 M/V subtype shown to have a Western blot signature like BASE) were to increase, this would suggest (although not prove) a causal relationship (Figure 5).

http://www.cdc.gov/ncidod/EID/vol12no12/06-0965.htm

Creutzfeldt-Jakob Disease Mortality in Japan, 1979-2004: Analysis of National Death Certificate Data

Yuriko Doi1), Tetsuji Yokoyama2), Miyoshi Sakai2) and Yosikazu Nakamura3)

1) Department of Epidemiology, National Institute of Public Health.
2) Department of Technology Assessment and Biostatistics, National Institute of Public Health.
3) Department of Public Health, Jichi Medical University.

(Received: September 13, 2006)
(Accepted: March 18, 2007)

Abstract
BACKGROUND: Trend of the mortality rate of Creutzfeldt-Jakob disease (CJD) in Japan is still unclear. This study aimed to estimate annual crude mortality rates due to CJD and examine the CJD mortality trend in Japan during the period of 1979-2004.
METHODS: National death certificate data on CJD were used (CJD coded as 046.1 for ICD-9 and A81.0 for ICD-10). Trends in age-standardized mortality rates for CJD were examined by using time series analyses including the joinpoint regression analysis.
RESULTS: A total of 1,966 deaths (862 males and 1,104 females) were identified with CJD coded as the underlying-cause-of-death. The annual number of deaths and crude mortality rates peaked in 2004 at 163 (66 for males and 97 for females) deaths and 1.28 (1.06 for males and 1.48 for females) deaths per million population per year, respectively. The age-specific mortality rates rapidly increased with age between 50 and 74 years, especially among females, and sharply declined at 80+ years. Throughout the observed period, there were no significant change points, and the annual percentage changes (95% confidence intervals) were +3.09 (2.18 - 4.02) % for males and +3.90 (2.98-4.83) % and females. The total number of CJD deaths under 50 years of age was 131, and there was found no increase in the annual number of deaths for the past few years in this age group.
CONCLUSION: CJD mortality in trend data based on death certificates has significantly increased in Japan during the period of 1979-2004.
J Epidemiol 2007; 17: 133-139.

Key words: Creutzfeldt-Jakob Syndrome; Regression Analysis; Mortality; Death Certificate; Japan

snip...

AS demonstrated in this study, we found a significant linear increase in trends for age standardized mortality rates from the disease, with +3-4% of annual percentage change, between 1979 and 2004. In interpreting the results, we should consider some factors that might contribute to a false increase in mortality, such as the change of ICD codes and the enhancement

of case findings (e.g., physicians9 recognition of the disease, diagnostic tests, and quality of health care). No revolutionary new diagnostic test for CJD became available throughout the observational period. On the other hand, there were a few critical points of time to consider: in 1991, patients with CJD transmitted by cadaveric dura transplants were identified in Japan9, in 1995, the ICD code for CJD was changed from 9th to 10th version in Japan; and in 1996, a new case of vCJD causally linked to BSE was reported from the United Kingdom.6 Without an abrupt rise of age-standardized mortality rates from CJD after these years for both sexes, however, it is unlikely that these events artificially affected the increase in CJD mortality.

Rather, it may be the true fact that in Japan our results reflect to a large extent a genuine increase in CJD. The number of iCJD cases may still increase even after the total ban on the practice of causal grafts.5,8 Regarding sporadic CJD (sCJD), a recent report from the European Unions collective study on CJD suggests that the mortality rates from sCJD increased with time between 1993 and 2002.20 It is quite probable that this temporal increase of sCJD may also exist in Japan. The increase may have been accompanied to some extent by the improvement of physicians diagnostic skills for CJD since 1997 when a manual for clinical practice on CJD was introduced in our country.20,21

J Neurol Neurosurg Psychiatry. Published Online First: 23 May 2007.
doi:10.1136/jnnp.2006.104570

Original articles

Sporadic creutzfeldt-jakob disease in two adolescents

K Murray 1, D L Ritchie 1, M Bruce 2, C A Young 3, M Doran 3, J W Ironside 4 and R G Will 4*
1 NationalCJD Surveillance Unit, United Kingdom
2 Neuropathogenesis Unit, United Kingdom
3 Walton Centre for Neurology and Neurosurgery, United Kingdom
4 National CJD Surveillance Unit, United Kingdom

* To whom correspondence should be addressed. E-mail: [log in to unmask]

Accepted 15 April 2007

Abstract

Background: Sporadic Creutzfeldt-Jakob disease (CJD) is a condition predominantly affecting older age groups, with cases aged less than 45 years rare and an age at onset or death of less than 20 years exceptional.

Methods: Data from the systematic study of sporadic CJD in the UK are available from 1970 onwards. Clinical and pathological data are reviewed in order to identify atypical cases, including those at the extremes of the age range of sporadic CJD. Detailed analysis of atypical cases is undertaken and in selected cases laboratory transmission studies are carried out in order to provide information on the characteristics of the infectious agent.

Results: In the UK two cases of sporadic CJD in adolescents have been identified, dying aged 16 and 20 years. The first case predated the epidemic of bovine spongiform encephalopathy and the characteristics of the second case, including laboratory transmission studies, are consistent with a diagnosis of sporadic rather than variant CJD.

Conclusion: The cases in this report indicate that sporadic CJD can develop at a very young age, that variant CJD is not the only form of CJD occurring in this age group and that neuropathological examination is essential to accurate diagnosis of human prion disease.

http://jnnp.bmj.com/cgi/content/abstract/jnnp.2006.104570v1

Coexistence of multiple PrPSc types in individuals with Creutzfeldt-Jakob disease

Magdalini Polymenidou, Katharina Stoeck, Markus Glatzel, Martin Vey, Anne Bellon, and Adriano Aguzzi

Summary

Background The molecular typing of sporadic Creutzfeldt-Jakob disease (CJD) is based on the size and glycoform ratio of protease-resistant prion protein (PrPSc), and

on PRNP haplotype. On digestion with proteinase K, type 1 and type 2 PrPSc display unglycosylated core fragments of 21 kDa and 19 kDa, resulting from cleavage around amino acids 82 and 97, respectively.

Methods

We generated anti-PrP monoclonal antibodies to epitopes immediately preceding the differential proteinase K cleavage sites. These antibodies, which were designated POM2 and POM12, recognise type 1, but not type 2, PrPSc.

Findings

We studied 114 brain samples from 70 patients with sporadic CJD and three patients with variant CJD. Every patient classified as CJD type 2, and all variant CJD patients, showed POM2/POM12 reactivity in the cerebellum and other PrPSc-rich brain areas, with a typical PrPSc type 1 migration pattern.

Interpretation

The regular coexistence of multiple PrPSc types in patients with CJD casts doubts on the validity of electrophoretic PrPSc mobilities as surrogates for prion strains, and questions the rational basis of current CJD classifications.

snip...

The above results set the existing CJD classifications

into debate and introduce interesting questions about

human CJD types. For example, do human prion types

exist in a dynamic equilibrium in the brains of affected

individuals? Do they coexist in most or even all CJD

cases? Is the biochemically identified PrPSc type simply

the dominant type, and not the only PrPSc species?

Published online October 31, 2005

http://neurology.thelancet.com

Detection of Type 1 Prion Protein in Variant Creutzfeldt-Jakob Disease

Helen M. Yull,* Diane L. Ritchie,*

Jan P.M. Langeveld,? Fred G. van Zijderveld,?

Moira E. Bruce,? James W. Ironside,* and

Mark W. Head*

From the National CJD Surveillance Unit,* School of Molecular

and Clinical Medicine, University of Edinburgh, Edinburgh,

United Kingdom; Central Institute for Animal Disease Control

(CIDC)-Lelystad, ? Lelystad, The Netherlands; Institute for Animal

Health, Neuropathogenesis Unit, ? Edinburgh, United Kingdom

Molecular typing of the abnormal form of the prion

protein (PrPSc) has come to be regarded as a powerful

tool in the investigation of the prion diseases. All evidence

thus far presented indicates a single PrPSc molecular

type in variant Creutzfeldt-Jakob disease (termed

type 2B), presumably resulting from infection with a single strain of the agent (bovine spongiform encephalopathy).

Here we show for the first time that the PrPSc that accumulates in the brain in variant Creutzfeldt-Jakob disease also contains a minority type 1 component. This minority type 1 PrPSc was found in all 21 cases of variant Creutzfeldt-Jakob disease tested, irrespective of brain region examined, and was also present in the variant Creutzfeldt-Jakob disease tonsil. The quantitative balance between PrPSc types was maintained when variant Creutzfeldt-Jakob disease was transmitted to wild-type mice and was also found in bovine spongiform encephalopathy cattle brain, indicating that the agent rather than the host specifies their relative representation. These results indicate that PrPSc molecular typing is based on quantitative rather than qualitative phenomena and point to a complex relationship between prion protein biochemistry, disease phenotype and agent strain. (Am J Pathol 2006, 168:151-157;

DOI: 10.2353/ajpath.2006.050766)

snip...

Discussion

Irrespective of whether this proves to be the case, the results shown here point to further complexities in the relationship between the physico-chemical properties of the prion protein, human disease phenotype, and prion agent strain.

Acknowledgments

snip...

Type 1 PrPSc in Variant Creutzfeldt-Jakob Disease 157

AJP January 2006, Vol. 168, No. 1 ...TSS

http://ajp.amjpathol.org/cgi/content/abstract/168/1/151maxtoshow=&HITS=10&hits=10&RESULTFORMAT=&fulltext=prion&searchid=1136646133963_237&FIRSTINDEX=0&volume=168&issue=1&journalcode=amjpathol

Neuropathology and Applied Neurobiology

(2005),

31

, 565-579 doi: 10.1111/j.1365-2990.2005.00697.x

565

Blackwell Science, LtdOxford, UKNANNeuropathology and Applied Neurobiology0305-1846Blackwell Publishing Ltd, 2005

316565579

Review article

Phenotypic variability in human prion diseases

J. W. Ironside, D. L. Ritchie and M. W. Head

National Creutzfeldt-Jakob Disease Surveillance Unit, Division of Pathology, University of Edinburgh, Edinburgh, UK

J. W. Ironside, D. L. Ritchie and M. W. Head (2005)

Neuropathology and Applied Neurobiology

31,

565-579

Phenotypic variability in human prion diseases

Human prion diseases are rare neurodegenerative disorders that can occur as sporadic, familial or acquired disorders. Within each of these categories there is a wide range of phenotypic variation that is not encountered in other neurodegenerative disorders. The identification of the prion protein and its key role in the pathogenesis of this diverse group of diseases has allowed a fuller understanding of factors that influence disease phenotype. In particular, the naturally occurring polymorphism at codon 129

in the prion protein gene has a major influence on the disease phenotype in sporadic, familial and acquired prion diseases, although the underlying mechanisms remain unclear. Recent technical advances have improved our ability to study the isoforms of the abnormal prion protein in the brain and in other tissues. This has lead to the concept of molecular strain typing, in which different isoforms of the prion protein are proposed to correspond to individual strains of the transmissible agent, each with specific biological properties. In sporadic Creutzfeldt-Jakob disease there are at least six major combinations of codon 129 genotype and prion protein isotype, which appear to relate to distinctive clinical subgroups of this disease. However, these relationships are proving to be more complex than first considered, particularly in cases with more than a single prion protein isotype in the brain. Further work is required to clarify these relationships and to explain the mechanism of neuropathological targeting of specific brain regions, which accounts for the diversity of clinical features within human prion diseases.

BSE prions propagate as either variant CJD-like or sporadic CJD-like prion strains in transgenic mice expressing human prion protein

The EMBO Journal Vol. 21 No. 23 pp. 6358±6366, 2002

Emmanuel A.Asante, Jacqueline M.Linehan,

Melanie Desbruslais, Susan Joiner,

Ian Gowland, Andrew L.Wood, Julie Welch,

Andrew F.Hill, Sarah E.Lloyd,

Jonathan D.F.Wadsworth and

John Collinge1

MRC Prion Unit and Department of Neurodegenerative Disease,

Institute of Neurology, University College, Queen Square,

London WC1N 3BG, UK

1Corresponding author

e-mail: j.collinge@prion.ucl.ac.uk

Variant Creutzfeldt±Jakob disease (vCJD) has been recognized to date only in individuals homozygous for methionine at PRNP codon 129. Here we show that

transgenic mice expressing human PrP methionine 129, inoculated with either bovine spongiform encephalopathy (BSE) or variant CJD prions, may develop the neuropathological and molecular phenotype of vCJD, consistent with these diseases being caused by the same prion strain. Surprisingly, however, BSE transmission to these transgenic mice, in addition to producing a vCJD-like phenotype, can also result in a distinct molecular phenotype that is indistinguishable from that of sporadic CJD with PrPSc type 2. These data suggest that more than one BSEderived prion strain might infect humans; it is therefore possible that some patients with a phenotype consistent with sporadic CJD may have a disease arising from BSE exposure.

snip...

The EMBO Journal Vol. 21 No. 23 pp. 6358±6366, 2002

http://embojournal.npgjournals.com/cgi/reprint/21/23/6358

J Neuropsychiatry Clin Neurosci 17:489-495, November 2005
doi: 10.1176/appi.neuropsych.17.4.489

Psychiatric Manifestations of Creutzfeldt-Jakob Disease: A 25-Year Analysis

Christopher A. Wall, M.D., Teresa A. Rummans, M.D.,
Allen J. Aksamit, M.D.,
Lois E. Krahn, M.D. and V. Shane Pankratz, Ph.D.
Received April 20, 2004; revised September 9, 2004;
accepted September 13,
2004. From the Mayo Clinic, Department of Psychiatry and Psychology,
Rochester, Minnesota; Mayo Clinic, Department of Neurology, Rochester,
Minnesota. Address correspondence to Dr. Wall, Mayo Clinic, Department of
Psychiatry and Psychology, Mayo Building-W11A, 200 First St., SW, Rochester,
MN 55905; wall.chris@mayo.edu (E-mail).

This study characterizes the type and timing of psychiatric manifestations in sporadic Creutzfeldt-Jakob disease (sCJD). Historically, sCJD has been characterized by prominent neurological symptoms, while the variant form (vCJD) is described as primarily psychiatric in presentation and course: A retrospective review of 126 sCJD patients evaluated at the Mayo Clinic from 1976-2001 was conducted. Cases were reviewed for symptoms of depression, anxiety, psychosis, behavior dyscontrol, sleep disturbances, and neurological signs during the disease course. Eighty percent of the cases demonstrated psychiatric symptoms within the first 100 days of illness, with 26% occurring at presentation. The most commonly reported symptoms in this population included sleep disturbances, psychotic symptoms, and depression. Psychiatric manifestations are an early and prominent feature of sporadic CJD, often occurring prior to formal diagnosis.

snip...

CONCLUSIONS

Historically, psychiatric manifestations have been described as a relatively infrequent occurrence in the sporadic form of creutzfeldt-Jakob disease.

However, our findings suggest otherwise. In this study, a vast majority of the cases were noted to have at least one psychiatric symptom during the course of illness, with nearly one-quarter occurring in the prodromal or presenting phase of the illness. After comparing the frequency of neuropsychiatric symptoms in sporadic CJD to studies describing the variant form of CJD, we found that there are fewer clinical differences than previously reported.5-7 While the age of patients with vCJD presentation is significantly younger and the course of illness is longer, the type and timing of psychiatric manifestations appear similar between these two diseases. ...snip...

http://neuro.psychiatryonline.org/cgi/content/abstract/17/4/489

Polish Journal of Neurology and Neurosurgery 6/2005

abstract:

CASE REPORT
Mental disorders in a female patient with probable Creutzfeldt-Jakob disease

Neurol Neurochirur Pol 2005; 39, 6: 520–523

authors: Marek Gronkowski, Bozena Spila, Alina Nowicka, Piotr Machala, Grzegorz Przywara,

The paper presents an overview of the current knowledge about the etiology, classification of Creutzfeldt-Jakob disease, abnormalities in the results of the EEG, MR and laboratory examinations in patients with this disease. The diagnostic value of the CSF examination for presence of protein 14-3-3 is underlined. The article is based on both Polish and foreign literature, describing mainly the diagnostics of CJD. The case of a female patient with dementia, mental disorders and neurological symptoms in the course of probable CJD, who was hospitalized at the Psychogeriatric Department of the Neuropsychiatric Hospital in Lublin is described.

Polish Journal of Neurology and Neurosurgery 6/2005

full text of the article:

http://www.termedia.pl/showpdf.php?article_id=4105&filename=Zaburzenia%20psych.pdf&priority=1

First case of vCJD reported in a Japanese patient: update

Editorial team (eurosurveillance.weekly@hpa.org.uk), Eurosurveillance editorial office

A detailed description of the first case of variant Creutzfeldt-Jakob disease (vCJD) in Japan, originally reported in February 2005, has just been published [1,2]. The patient was a 51 year old man, who had spent around 24 days in the United Kingdom in 1990, during the bovine spongiform encephalopathy (BSE) outbreak. He is known to have eaten mechanically recovered meat during his visit, and although exposure in other European countries he visited, including France and Japan, cannot be excluded, it is thought that he may have been exposed to the BSE agent during his UK visit. If exposure in the UK was the source of his infection, then the incubation period to illness onset was 11.5 years.

It is also noted that the patient's illness duration was unusually long, at 42 months, and that periodic synchronous discharges (PSD), which have not been reported in other vCJD cases, appeared on the patient's electroencephalogram, 12 months before death. The working group reporting on the case suggest that the World Health Organization vCJD case definition [3] be revised to state that PSD does not exclude the possibility of vCJD.

This article is adapted from reference 1

References:

Yamada M, Variant CJD Working Group. The first Japanese case of variant Creutzfeldt-Jakob disease showing periodic electroencephalogram. Lancet 2006; 367: 874.

Eurosurveillance. First case of vCJD reported in a Japanese patient. Eurosurveillance 2005; 10(2): 050210. (http://www.eurosurveillance.org/ew/2005/050210.asp#1)

The Revision of the Surveillance Case Definition for Variant Creutzfeldt-Jakob Disease (vCJD). Report of a WHO consultation, Edinburgh, United Kingdom 17 May 2001. WHO/CDS/CSR/EPH/2001.5. Geneva: World Health Organization; 2001 (http://www.who.int/csr/resources/publications/bse/whocdscsreph20015.pdf)

http://www.eurosurveillance.org/ew/2006/060316.asp#3

sporadic cjd

http://lists.ifas.ufl.edu/cgi-bin/wa.exe?A2=ind0705&L=sanet-mg&P=25276

http://lists.ifas.ufl.edu/cgi-bin/wa.exe?A2=ind0705&L=sanet-mg&T=0&P=25276

Colorado Surveillance Program for Chronic Wasting Disease
Transmission to Humans (TWO SUSPECT CASES)

http://lists.ifas.ufl.edu/cgi-bin/wa.exe?A2=ind0704&L=sanet-mg&T=0&P=1165

An evaluation of scrapie surveillance in the United States
From: Terry S. Singeltary Sr.
Date: Sun, 5 Aug 2007 13:05

http://lists.ifas.ufl.edu/cgi-bin/wa.exe?A2=ind0708&L=sanet-mg&T=0&P=3427

SEAC New forms of Bovine Spongiform Encephalopathy 1 August 2007
From: Terry S. Singeltary Sr.
Date: Sun, 5 Aug 2007 13:09:38 -0500

http://lists.ifas.ufl.edu/cgi-bin/wa.exe?A2=ind0708&L=sanet-mg&T=0&P=3573

Owens, Julie
From: Terry S. Singeltary Sr. [flounder9@verizon.net]
Sent: Monday, July 24, 2006 1:09 PM
To: FSIS RegulationsComments
Subject: [Docket No. FSIS-2006-0011] FSIS Harvard Risk Assessment of Bovine Spongiform Encephalopathy (BSE)
Page 1 of 98
8/3/2006

Greetings FSIS,

I would kindly like to comment on the following ;

[Federal Register: July 12, 2006 (Volume 71, Number 133)]
[Notices]
[Page 39282-39283]

From the Federal Register Online via GPO Access [wais.access.gpo.gov]
[DOCID:fr12jy06-35]

DEPARTMENT OF AGRICULTURE
Food Safety and Inspection Service
[Docket No. FSIS-2006-0011]
Harvard Risk Assessment of Bovine Spongiform Encephalopathy (BSE)
Update; Notice of Availability and Technical Meeting

snip...

http://a257.g.akamaitech.net/7/257/2422/01jan20061800/edocket.access.gpo.gov/2006/E6-10928.htm

MY comments/questions are as follows ;

1. SINCE the first Harvard BSE Risk Assessment was so flawed and fraught with error after the PEER REVIEW assessment assessed this fact, how do you plan on stopping this from happening again, will there be another peer review with top TSE Scientist, an impartial jury so-to-speak, to assess this new and updated Harvard BSE/TSE risk assessment and will this assessment include the Atypical TSE and SRM issues ?

*** Suppressed peer review of Harvard study October 31, 2002 ***

http://www.fsis.usda.gov/oa/topics/BSE_Peer_Review.pdf

2. WITH A RECENT NATION WIDE MAD COW FEED BAN RECALL in the past few months that consisted of some 10,878.06 TONS, then another Mad Cow feed ban warning letter in May, IT should seem prudent to ask why our feed bans continue to fail in 2006, and continue to fail today ?

snip...

full text 98 pages ;

http://www.fsis.usda.gov/OPPDE/Comments/2006-0011/2006-0011-1.pdf

[Docket No. 03-025IFA] FSIS Prohibition of the Use of Specified Risk Materials for Human Food and Requirement for the Disposition of Non-Ambulatory Disabled Cattle

03-025IFA
03-025IFA-2
Terry S. Singeltary

From: Terry S. Singeltary Sr. [flounder9@verizon.net]

Sent: Thursday, September 08, 2005 6:17 PM

To: fsis.regulationscomments@fsis.usda.gov

Subject: [Docket No. 03-025IFA] FSIS Prohibition of the Use of Specified Risk Materials for Human Food and Requirements for the Disposition of Non-Ambulatory Disabled Cattle

Greetings FSIS,

I would kindly like to submit the following to [Docket No. 03-025IFA] FSIS Prohibition of the Use of Specified Risk Materials for Human Food and Requirements for the Disposition of Non-Ambulatory Disabled Cattle
THE BSE/TSE SUB CLINICAL Non-Ambulatory Disabled Cattle Broken bones and such may be the first signs of a sub clinical BSE/TSE Non-Ambulatory Disabled Cattle ;

SUB CLINICAL PRION INFECTION

MRC-43-00

Issued: Monday, 28 August 2000

NEW EVIDENCE OF SUB-CLINICAL PRION INFECTION: IMPORTANT RESEARCH

FINDINGS RELEVANT TO CJD AND BSE

A team of researchers led by Professor John Collinge at the Medical

Research Council Prion Unit1 report today in the Proceedings of the

National Academy of Sciences, on new evidence for the existence of a

"sub-clinical" form of BSE in mice which was unknown until now....

full text 17 pages ;

https://web01.aphis.usda.gov/regpublic.nsf/0/eff9eff1f7c5cf2b87256ecf000df08d?OpenDocument

Docket No, 04-047-l Regulatory Identification No. (RIN) 091O-AF46 NEW BSE SAFEGUARDS

Docket

No. 04-047-l

No. 04-021ANPR

No. 2004N-0264

NEW BSE SAFEGUARDS

Federal Measures to Mitigate BSE Risks: Considerations for Further Action

http://www.fda.gov/cvm/index/updates/bseanprm.htm

Greetings FDA, USDA and APHIS et al,

I would kindly like to comment on the continued delay of the regulations that have been proposed for years to reduce the risk of BSE/TSE in the USA. Each day that is wasted debating this issue allows this agent to spread,
and many many more humans and animals become needlessly exposed to this agent via a multitude of potential routes and sources right here in the USA. TO continue to ignore the new findings from several scientists about the fact that BSE is not the only strain of TSE in cattle, the fact that

new atypical strains of TSE are showing up in not only cattle, but sheep and the fact that the new strain of TSE in cattle seems to be more similar to sporadic CJD as opposed to the nv/v CJD, to continue to ignore these findings will only further spread this agent. .....

full text ;

https://web01.aphis.usda.gov/regpublic.nsf/168556f5aa7a82ba85256ed00044eb1f/eff9eff1f7c5cf2b87256ecf000df08d

Docket Management Docket: 02N-0273 - Substances Prohibited From Use in

Animal Food or Feed; Animal Proteins Prohibited in Ruminant Feed

Comment Number: EC -10

Accepted - Volume 2

http://www.fda.gov/ohrms/dockets/dailys/03/Jan03/012403/8004be07.html

PART 2

http://www.fda.gov/ohrms/dockets/dailys/03/Jan03/012403/8004be09.html

2003D-0186
Guidance for Industry: Use of Material From Deer and Elk In Animal Feed

EMC 7
Terry S. Singeltary Sr.
Vol #:
1

http://www.fda.gov/ohrms/dockets/dailys/03/oct03/100203/100203.htm

Docket Management Docket: 02N-0276 - Bioterrorism Preparedness ...
General Comments, Subject: Docket No: 02-088-1 RE-Agricultural ...
From: Terry S. Singeltary Sr.

To: regulations@aphis.usda.gov

Docket No: 02-088-1 Title: ...

Greetings FDA and public,

if you go to the below site, and search all BSE known countries and check out their air traffic illegal meat they have confiscated, and check out the low number checked, compared to actual passenger traffic, would not take too much for some nut to bring in FMD/TSEs into the USA as a 'suitcase bomb'.

[[Under APHIS-PPQ's agricultural quarantine inspection monitoring, 284 air passengers from Israel were sampled for items of agricultural interest in fiscal year 2001. Seven of these passengers, or 2 percent, carried a total of 11 kg of meat items that could potentially harbor the pathogen that causes BSE. None of these passengers from whom meat items were confiscated reported plans to visit or work on a ranch or farm during their visit to the U.S.]]

if they were to have questioned the terrorist that bombed the Twin Towers with jets, if they were to have questioned them at flight school in the USA, i am sure that they would have said they did not intend to visit the Twin Towers as a flying bomb either. what am i thinking, they probably did ask this? stupid me. ...

full text ;

http://www.fda.gov/ohrms/DOCKETS/dockets/02n0276/02N-0276-EC-254.htm

PDF]Freas, William TSS SUBMISSION

File Format: PDF/Adobe Acrobat -

Page 1. J Freas, William From: Sent: To: Subject: Terry S. Singeltary

Sr. [flounder@wt.net] Monday, January 08, 200l 3:03 PM freas ...

Greetings again Dr. Freas and Committee Members,

I wish to submit the following information to the
Scientific Advisors and Consultants Staff
2001 Advisory Committee (short version).

I understand the reason of having to shorten my submission,
but only hope that you add it to a copy of the long version,
for members to take and read at their pleasure,
(if cost is problem, bill me, address below).
So when they realize some time in the near future
of the 'real' risks i speak of from human/animal TSEs and
blood/surgical products. I cannot explain the 'real' risk
of this in 5 or 10 minutes at some meeting,
or on 2 or 3 pages, but will attempt here:

remember AIDS/HIV, 'no problem to heterosexuals in the U.S.?
no need to go into that, you know of this blunder.

DO NOT make these same stupid mistakes again with
human/animal TSE's aka MADCOW DISEASE. I lost my Mom to hvCJD,
and my neighbor lost his Mother to sCJD as well (both cases
confirmed). I have seen many deaths, from many diseases.
I have never seen anything as CJD, I still see my Mom laying helpless,
jerking tremendously, and screaming "God, what's wrong
with me, why can't I stop this". I still see this, and will
never forget. Approximately 10 weeks from 1st of symptoms to death.
This is what drives me. I have learned more in 3 years about not only
human/animal TSE's but the cattle/rendering/feeding industry/government
than i ever wished to.

I think you are all aware of CJD vs vCJD, but i don't think
you all know the facts of human/animal TSE's as a whole,
they are all very very similar, and are all tied to the
same thing, GREED and MAN.

I am beginning to think that the endless attempt to track
down and ban, potential victims from known BSE Countries
from giving blood will be futile. You would have to ban
everyone on the Globe eventually? AS well, I think we
MUST ACT SWIFTLY to find blood test for TSE's,
whether it be blood test, urine test, eyelid test,
anything at whatever cost, we need a test FAST.

DO NOT let the incubation time period of these TSEs fool you.....

full text 6 pages ;

http://www.fda.gov/ohrms/dockets/ac/01/slides/3681s2_09.pdf

SEE STEADY INCREASE IN SPORADIC CJD IN THE USA FROM 1997 TO 2006. SPORADIC CJD CASES TRIPLED, with phenotype of 'UNKNOWN' strain growing. ...

http://www.cjdsurveillance.com/resources-casereport.html

Diagnosis and Reporting of Creutzfeldt-Jakob Disease

Singeltary, Sr et al. JAMA.2001; 285: 733-734.

http://jama.ama-assn.org/http://www.neurology.org/cgi/eletters/60/2/176#535

BRITISH MEDICAL JOURNAL

BMJ

http://www.bmj.com/cgi/eletters/319/7220/1312/b#5406

BMJ

http://www.bmj.com/cgi/eletters/320/7226/8/b#6117

JOURNAL OF NEUROLOGY

MARCH 26, 2003

RE-Monitoring the occurrence of emerging forms of Creutzfeldt-Jakob

disease in the United States

Email Terry S. Singeltary:

flounder@wt.net

I lost my mother to hvCJD (Heidenhain Variant CJD). I would like to

comment on the CDC's attempts to monitor the occurrence of emerging forms of CJD. Asante, Collinge et al [1] have reported that BSE transmission to the 129-methionine genotype can lead to an alternate phenotype that is indistinguishable from type 2 PrPSc, the commonest sporadic CJD. However, CJD and all human TSEs are not reportable nationally. CJD and all human TSEs must be made reportable in every state and internationally. I hope that the CDC does not continue to expect us to still believe that the 85%+ of all CJD cases which are sporadic are all spontaneous, without route/source. We have many TSEs in the USA in both animal and man. CWD in deer/elk is spreading rapidly and CWD does transmit to mink, ferret, cattle, and squirrel monkey by intracerebral inoculation. With the known incubation periods in other TSEs, oral transmission studies of CWD may take much longer. Every victim/family of CJD/TSEs should be asked about route and source of this agent. To prolong this will only spread the agent and needlessly expose others. In light of the findings of Asante and Collinge et al, there should be drastic measures to safeguard the medical and surgical arena from sporadic CJDs and all human TSEs. I only ponder how many sporadic CJDs in the USA are type 2 PrPSc?

http://www.neurology.org/cgi/eletters/60/2/176#535

THE PATHOLOGICAL PROTEIN
Hardcover, 304 pages plus photos and illustrations. ISBN 0-387-95508-9

June 2003

BY Philip Yam

CHAPTER 14 LAYING ODDS

Answering critics like Terry Singeltary, who feels that the U.S. under-counts CJD, Schonberger conceded that the current surveillance system has errors but stated that most of the errors will be confined to the older population.

http://www.thepathologicalprotein.com/

doi:10.1016/S1473-3099(03)00715-1

Newsdesk

Tracking spongiform encephalopathies in North America

Xavier Bosch

Available online 29 July 2003.

Volume 3, Issue 8, August 2003, Page 463

“My name is Terry S Singeltary Sr, and I live in Bacliff, Texas. I lost my mom to hvCJD (Heidenhain variant CJD)
and have been searching for answers ever since. What I have found is that we have not been told the truth. CWD
in deer and elk is a small portion of a much bigger problem.”

.....

http://www.thelancet.com/journals/laninf/article/PIIS1473309903007151/fulltext

http://download.thelancet.com/pdfs/journals/1473-3099/PIIS1473309903007151.pdf

see history of cjd questionnaire

http://brain.hastypastry.net/forums/showthread.php?t=2408

Terry S. Singeltary Sr.
P.O. Box 42
Bacliff, Texas USA 77518