information before going into the archive. Therefore if abnormal prion proteins are found in a tonsil sample, the results cannot be passed back to the patient.

This anonymous procedure is used because the significance for an otherwise well person of finding abnormal prion protein in their tonsil tissue is unknown at present. The Research Ethics Committee that reviewed the study supported the view that the tonsils should be tested anonymously.

5. Since 1995 there have been 168 definite or probable cases of vCJD in Britain, resulting in 115 deaths from vCJD and 49 deaths thought likely to be due to vCJD. Back calculation based on these cases would suggest between 10 and 190 further clinical cases over the next ten years.

6. The NATA study is able to detect presence of the prion protein regardless of the genotype of the prion protein gene.

7. For further information on this press release please contact the Health Protection Agency's Centre for Infections press office on:

Kate Swan 020 8327 7097
Georgina Fletcher 020 8327 6690
Louise Brown 020 8327 7080
Alex Baker 020 8327 7098
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Last reviewed: 21 May 2009

別紙様式第2-1

No. 23

医薬品 研究報告 調査報告書

識別番号・報告回数			報告日	第一報入手日	新医薬品等の区分	総合機構処理欄
				2009. 3. 18	該当なし	

一般的名称	解凍人赤血球濃厚液	研究報告の公表状況		公表国
販売名(企業名)	解凍赤血球濃厚液「日赤」(日本赤十字社) 照射解凍赤血球濃厚液「日赤」(日本赤十字社) 解凍赤血球-LR「日赤」(日本赤十字社) 照射解凍赤血球-LR「日赤」(日本赤十字社)		Ferguson-Smith MA, Richt JA. Nature. 2009 Feb 26;457(7233):1079.	英国

研究報告の概要

○稀なBSE突然変異により公衆衛生リスクが懸念される
最近、非定型(H-型、L-型)のウシ海綿状脳症(BSE)が、日本、カナダ、米国に加え、複数のヨーロッパ諸国で発生した。これにより、ヒトの変異型クロイツフェルトヤコブ病(vCJD)が増加するというありがたくない可能性が浮上している。これまで検査された非定型BSE症例のうち、プリオンタンパク遺伝子(PRNP)の突然変異が検出されたのは1例(アラバマ州のBSE牛)のみで、このウシの健常な仔ウシにも突然変異が存在した。これは当該疾患が遺伝性である可能性を示す。実際、2000年のUK BSE Inquiryの報告では、英国のBSE流行はこうした変異による可能性が高いことが示され、スクレイピー関連とする仮説に反対の見解を示した。
非定型BSEを発症させる可能性のある稀なPRNP変異は、オーストラリアとニュージーランドのようなBSEが発生していないと考えられている国々でも起こる可能性がある。このため、ウシに対する厳しいBSE調査を継続し、反すう動物の厳密な飼料規制を行うことが重要である(現在でも多くの国がブタに反すう動物性タンパク質を与えている)。食肉処理時にウシの特定危険部位(脳や脊髄など)を除去することで、感染部位がヒトの食物連鎖に入り込むことを回避できる。
現在利用可能なウシのPRNP突然変異を調べるルーチン遺伝子スクリーニング検査により、公衆リスクについてさらなるデータが得られるだろう。アラバマのウシに同定された点突然変異は、ヒトで最も一般的な型の家族性(遺伝的)CJDの原因と同一であるため、これによって生じる感染性プリオンタンパク質は、より容易にウシ-ヒト関門を通過する可能性が考えられる。vCJD患者の特定は今後も続くだろう。発症頻度が減少しているからといって、将来のアウトブレイクの防止に必要な規制を緩和すべきではない。

報告企業の意見	今後の対応
非定型ウシ海綿状脳症(BSE)が、日本、カナダ、米国に加え、複数のヨーロッパ諸国で発生し、オーストラリアとニュージーランドのようなBSEが発生していないと考えられている国々でも起こる可能性があるとの報告である。	日本赤十字社は、vCJDの血液を介する感染防止の目的から、献血時に過去の海外渡航歴(旅行及び居住)を確認し、欧州36ヶ国に一定期間滞在したドナーを無期限に献血延期としている。また、英国滞在歴を有するvCJD患者が国内で発生したことから、平成17年6月1日より1980～96年に1日以上の英国滞在歴のある人の献血を制限している。今後もCJD等プリオン病に関する新たな知見及び情報の収集に努める。

使用上の注意記載状況・その他参考事項等

解凍赤血球濃厚液「日赤」
照射解凍赤血球濃厚液「日赤」
解凍赤血球-LR「日赤」
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血液を介するウイルス、
細菌、原虫等の感染
vCJD等の伝播のリスク

(18)

CORRESPONDENCE

Human dignity must be basis for debate on primate research

SIR — Bill Crum emphasizes a fundamental keynote of biomedical-research ethics in his Correspondence 'It should be possible to replace animals in research' (*Nature* **457**, 657; 2009) by stating that "good medical science" is not necessarily "morally justifiable or morally acceptable". On the other hand, many states and societies claim 'freedom of research' — meaning research being free from the need for justification — as a basic right. On the face of it, this looks like a discrepancy.

However, we have to recognize the fact that this freedom, like every other kind of freedom, has its ethical limits. Research can only be a right as long as it is not acting against our fundamental moral value: respect for human dignity. This is the basic point that we should agree on, regardless of our different opinions on what might constitute a breach of that principle.

With human dignity in mind, the ethical discussion about research on non-human primates has to focus on answering two questions. First, would prohibiting studies on primates constitute a threat to the human dignity of future generations, by reducing their chances of what we could consider a good life, as Roberto Caminiti states in his Correspondence 'Replacement of animals in research will never be possible' (*Nature* **457**, 147; 2009)? Second, is performing "invasive medical experiments" on creatures that "provide excellent experimental models of human cognition", as Crum states, a threat to our own dignity and our vision of how a good life should be led?

Only by using human dignity as the normative correlate for ethical decisions can we ensure that these decisions will be made on a basis that is equally important to all parties in this debate.

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Readers are welcome to comment at http://tinyurl.com/c62pgf

Rare BSE mutation raises concerns over risks to public health

SIR — Atypical forms (known as H- and L-type) of bovine spongiform encephalopathy (BSE) have recently appeared in several European countries as well as in Japan, Canada and the United States. This raises the unwelcome possibility that variant Creutzfeldt–Jakob disease (vCJD) could increase in the human population.

Of the atypical BSE cases tested so far, a mutation in the prion protein gene (*PRNP*) has been detected in just one, a cow in Alabama with BSE; her healthy calf also carried the mutation (J. A. Richt and S. M. Hall *PLoS Pathog.* **4**, e1000156; 2008). This raises the possibility that the disease could occasionally be genetic in origin. Indeed, the report of the UK BSE Inquiry in 2000 suggested that the UK epidemic had most likely originated from such a mutation and argued against the scrapie-related assumption.

Such rare potential pathogenic *PRNP* mutations could occur in countries at present considered to be free of BSE, such as Australia and New Zealand. So it is important to maintain strict surveillance for BSE in cattle, with rigorous enforcement of the ruminant feed ban (many countries still feed ruminant proteins to pigs). Removal of specified risk material, such as brain and spinal cord, from cattle at slaughter prevents infected material from entering the human food chain.

Routine genetic screening of cattle for *PRNP* mutations, which is now available, could provide additional data on the risk to the public. Because the point mutation identified in the Alabama animals is identical to that responsible for the commonest type of familial (genetic) CJD in humans, it is possible that the resulting infective prion protein might cross the bovine–human species barrier more easily. Patients with vCJD continue to be identified. The fact that this is happening less often should not lead to relaxation of the controls necessary to prevent future outbreaks.

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Scientific links with Cuba flourished despite US embargo

SIR — In your Editorial 'Cuba's biotech boom' (*Nature* **457**, 130; 2009), you state that "despite many constraints on interaction between Cuban and US scientists, biotech has prospered". In fact, US biotechnologists contributed in no small way to its development.

At the start, during the early 1980s, Cuban biotechnology was confined to a small house in a Havana suburb. An American group organized by Harlyn Halvorson, then director of Brandeis University's Rosenstiel Center and an inspirational leader, stepped in to help the venture. We were received warmly in Cuba whenever we visited.

The biotechnology effort soon transferred to a larger house across the street and from 1986 was housed in the majestic Center for Genetic Engineering and Biotechnology. The Cuban scientists set up symposia where one or more of us would speak.

The US government allowed us to travel to Cuba on the condition that we spent no American dollars there. We therefore continued to advise this fledgling group until the Soviet Union ceased to support Cuba financially and they could no longer pay for our visits.

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Idea of a love drug was no mystery to Shakespeare

SIR — In his Essay 'Love: neuroscience reveals all' (*Nature* **457**, 148; 2009), Larry Young claims that the biochemical understanding of love is not poetry. But at least one poet, namely William Shakespeare, foretold the application of drugs to manipulate the brain systems associated with pair bonding.

In *A Midsummer Night's Dream*, Oberon maintains that topical applications of the juice of the wild pansy (*Viola tricolor*, called 'love-in-idleness' in the play) "Will make or man or woman madly dote Upon the next live creature that it sees" (Act 2, Scene 1). The potion proves highly effective, supplying much of the humour in the play as Titania falls in love with the donkey-headed Bottom. Shakespeare also suggests that other substances from "Dian's bud" — variously identified as a species of wormwood (*Artemisia* spp.) or chaste tree (*Vitex agnus-castus*, a species not native to England but long known for its anti-libidinal properties) — could reverse the neurobiological results of the pansy. Perhaps poets have something to teach us about neurobiology and love after all.

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別紙様式第2-1

No. 25

医薬品 研究報告 調査報告書

識別番号・報告回数			報告日	第一報入手日	新医薬品等の区分	総合機構処理欄
				2009. 4. 15	該当なし	

一般的名称	人赤血球濃厚液	研究報告の公表状況		公表国
販売名(企業名)	赤血球濃厚液-LR「日赤」(日本赤十字社) 照射赤血球濃厚液-LR「日赤」(日本赤十字社)		ProMED 20090108.0076, 2009 Jan 8. 情報源:UK: National CJD Surveillance Unit – monthly statistics as of 5 Jan 2009, 2009 Jan 5.	英国

研究報告の概要

○プリオン病最新情報 英国:国立CJDサーベイランスユニット、月次vCJD・CJD統計、2009年1月5日時点
英国のCJDサーベイランスユニットから公表されたvCJDを始めとするプリオン病の患者数に関する最新情報である。2008年は、12月31日時点で140名の照会があった。内訳は、孤発性CJDによる死亡患者:73名、医原性CJDによる死亡患者:5名、GSS:3名、家族性CJD:2名、vCJD:1名。vCJD確定例または可能性例総数は前月から変化なく167名のままである。このデータは英国におけるvCJD流行は減少しつつあるとする見解に一致する。死亡患者数のピークは2000年の28名であり、その後2001年に20名、2002年に17名、2003年に18名、2004年に9名、2005年に5名、2006年に5名、2007年に5名、2008年に1名と減少している。

報告企業の意見	今後の対応
英国CJDサーベイランスユニットの統計によると、2009年1月5日の時点で、vCJD死亡患者総数には前月から変化なく167名のままであり、英国におけるvCJD流行は減少しつつあるとする見解に一致するとの報告である。	日本赤十字社は、vCJDの血液を介する感染防止の目的から、献血時に過去の海外渡航歴(旅行及び居住)を確認し、欧州36ヶ国に一定期間滞在したドナーを無期限に献血延期としている。また、英国滞在歴を有するvCJD患者が国内で発生したことから、平成17年6月1日より1980～96年に1日以上の英国滞在歴のある人の献血を制限している。今後もCJD等プリオン病に関する新たな知見及び情報の収集に努める。

使用上の注意記載状況・その他参考事項等

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照射赤血球濃厚液-LR「日赤」

血液を介するウイルス、
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Archive Number 20090108.0076
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PRION DISEASE UPDATE 2009 (01)
****
A ProMED-mail post
<http://www.promedmail.org>
ProMED-mail is a program of the
International Society for Infectious Diseases
<http://www.isid.org>

[With the continuing decline in the number of cases in the human population of variant Creutzfeldt-Jakob disease -- abbreviated previously as vCJD or CJD (new var.) in ProMED-mail -- it has been decided to broaden the scope of the occasional ProMED-mail updates to include other prion-related diseases. Data on vCJD cases and other forms of CJD: sporadic, iatrogenic, familial, and GSS (Gerstmann-Straussler-Scheinker disease) are included also when they have some relevance to the incidence and etiology of vCJD. - Mod.CP

In this update:
[1] UK: National CJD Surveillance Unit - monthly statistics as of 5 Jan 2009
[2] France: Institut de Veille Sanitaire - as of 30 Dec 2008
[3] US National Prion Disease Pathology Surveillance Center - as of 30 Nov 20
[4] and [5] Prion protein function
[6] CJD Update

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[1] UK: National CJD Surveillance Unit - monthly statistics as of 5 Jan 2009
Date: Mon 5 Jan 2009
Source: UK National CJD Surveillance Unit, monthly statistics [edited]
<http://www.cjd.ed.ac.uk/figures.htm>

The number of suspect cases of vCJD referred to the CJD surveillance unit in Edinburgh and the number of deaths of definite and probable variant Creutzfeldt-Jakob disease [abbreviated in ProMED-mail as CJD (new var.) or vCJD], the form of the disease thought to be linked to BSE (bovine spongiform encephalopathy), remain unchanged since the previous monthly report; that is, the number of definite or probable vCJD cases (dead and alive) remains 16

This situation is consistent with the view that the vCJD outbreak in the UK is in decline. 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, and only one so far (up to the end of 2008).

Totals for all types of CJD cases in the year 2008

As of 31 Dec 2008 in the UK, so far there have been 140 referrals, 73 deaths from sporadic CJD, 5 deaths from iatrogenic CJD, 3 from GSS, 2 from familial CJD, and one from vCJD.

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Communicated by:

ProMED-mail
<promed@promedmail.org>

******
[2] France: Institut de Veille Sanitaire - as of 30 Dec 2008
Date: 30 Dec 2008
Source: IVS - Maladie de Creutzfeldt-Jakob et
maladies apparentees [French, trans. & summ. Mod.CP, edited]
<http://www.invs.sante.fr/display/?doc=publications/mcj/donnees_mcj.html>

During the period 1992 to 2008, there were 23
cases of vCJD, all now deceased. They occurred
between 1996 and 2007: one case in 1996, one in
2000, one in 2001, 3 in 2002, none in 2003, 2 in
2004, 6 in 2005, 6 in 2006, 3 in 2007, and none
so far in 2008. There were 12 male and 11 female patients.

Their ages at time of death ranged from 19 to 58
years (mean 39); 6 of the patients resided in the
Ile-de-France [Paris area] and 17 in the
provinces. All the cases were met-met homozygotes
for codon 129 of the prion protein gene. No
special risk factors were evident, which
distinguished these patients from those with
other forms of CJD (sporadic, genetic,
iatrogenic). However, one patient had visited the UK at regular intervals.

Totals for all types of CJD cases in the year 2008

As of 30 Dec 2008 in France, during the course of
2008 there have been 1438 referrals, 76 deaths
from sporadic CJD, 3 deaths from iatrogenic CJD,
8 from familial CJD, none from GSS, and none from vCJD.

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Communicated by:
ProMED-mail
<promed@promedmail.org>

******
[3] US National Prion Disease Pathology Surveillance Center - as of 30 Nov 20(
Date: 30 Nov 2008
Source: US National Prion Disease Pathology Surveillance Center [edited]
<http://www.cjdsurveillance.com/resources-casereport.html>

Cases examined - as of 30 Nov 2008

During the period 1997 to 30 Nov 2008, 2 cases of
vCJD were reported, both contracted overseas. The
1st case was recorded in 2004, disease contracted
in the UK, and the 2nd in 2006, disease contracted in Saudi Arabia.

Totals for all types of CJD cases in the year 2008 as of 30 Nov 2008

So far in 2008 there have been 332 referrals, 199
cases of prion disease, including 151 cases of
sporadic CJD, 21 cases of familial CJD, no cases
of atrogenic CJD and no indigenous cases of vCJD.

Overall during the period 1997 to 2008, there
have been 3018 referrals, 1745 cases of prion
disease, 1456 cases of sporadic CJD, 252 cases of
familial CJD, 4 cases of iatrogenic CJD and no indigenous cases of vCJD.

[During 2008 so far the USA with approximately
2.5x the combine populations of the UK and France
have reported a similar number of cases of
sporadic CJD (149 versus 151). Whether this is
due ot a difference in surveillance procedure or
actual disease incidence is unclear at the present time. - Mod.CP]

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Communicated by:

ProMED-mail
<promed@promedmail.org>

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[4] Prion protein function
Date: Sun 21 Dec 2008
Source: BBC News online [edited]
<http://news.bbc.co.uk/1/hi/health/7788444.stm>

Scientists sniff out prion secret

The brain protein which has a hand, when defective, in the lethal disease CJD may also be involved in aiding our sense of smell. Mice bred to lack the prion protein could not find buried food or choose between smells. Columbia University scientists said some symptoms of prion disease might be due to the loss of the protein's original role. The study was published in the journal Nature Neuroscience [see below].

The prion protein has historically received something of a bad press, being blamed in its misshapen form for degenerative brain diseases in humans and other animals. However, many scientists have been trying to uncover what it actually does when it is behaving correctly. Dr Stuart Firestein's team believe that one of these roles is to help us smell. While his prion-protein free mice were still able to detect scents, they had lost some higher functions which required that smell information to be analysed and processed by the brain. The scientists found changes in the communication between neurons in the nerve cells of the olfactory bulb, part of the forebrain which deals with odours. When the protein was restored to this part of the brain, the ability to discriminate between odours came back.

The scientists said that while the discovery had no direct link to the diseases caused by faulty prion proteins, it might help account for some of the symptoms experienced by patients, which might be due to the failure of the proteins to do their normal job properly, rather than the damage caused by accumulation of defective prions.

This is not the 1st suggested role for the prion protein -- in 2007, Leeds University scientist Professor Nigel Hooper said that it might help reduce the formation of "plaques" linked to the onset of Alzheimer Disease. He said of the newly-reported research: "It's likely that these proteins have a number of roles in various different body systems, including the olfactory system, as suggested here. "I don't think you can say that it is so mysterious any more, or that we do not understand what it does."

[Reference: Nature Neuroscience, Published online: 21 December 2008 doi:10.1038/nn.2238
<http://www.nature.com/neuro/journal/v12/n1/abs/nn.2238.html>

Title: Olfactory behavior and physiology are disrupted in prion protein knockout mice
Authors: Claire E Le Pichon1, Matthew T Valley1, Magdalini Polymenidou2,3, Alexander T Chesler1, Botir T Sagdullaev1,3, Adriano Aguzzi2 & Stuart Firestein1

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