Variant Creutzfeldt–Jakob disease

Reporting on 2014 data retrieved from TESSy* on 10 October 2015

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Key facts

- No new confirmed cases of variant Creutzfeldt–Jakob disease (vCJD) were reported in 2014.
- The disease is today extremely rare, which is consistent with the current understanding of the underlying epidemiology of vCJD and with the positive impact of risk mitigation measures to remove potential infectious animal material from the human food chain introduced in the EU from the late 1980s.
- The long incubation period – which can last years before the infected person will demonstrate physical symptoms –, the associated risk of secondary transmission from pre-clinically infected individuals, and the possible relaxation of feed control measures means that continued surveillance is crucial. Surveillance is also essential in order to monitor the gradual elimination of the disease and assess the impact of control measures at the EU level.

Methods

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In 2012, ECDC took over the surveillance for vCJD cases in the EU. The ECDC-operated TESSy database includes individual case data from all vCJD cases diagnosed in the EU. Prospective reporting of 'probable' or 'confirmed' new cases is now done in TESSy, in accordance with to the EU-2012 case definition.

The clinical presentation and associated diagnostic criteria for vCJD are relatively unusual. Suspected cases are typically reported to national surveillance centres. Typically, the centres then offer diagnostic support and, if needed, post-mortem analysis. Ultimately, successful vCJD surveillance requires the identification of patients as 'possible' CJD cases, supported by accurate differential diagnosis between vCJD and other more common forms of CJD (sporadic, iatrogenic, and familial).

A further diagnostic constraint is the need to obtain appropriate tissue samples post mortem to determine neuropathological characteristics associated with vCJD. In many cases, such tissue is not available, and in these situations, cases can only be classified as 'possible' or 'probable' based on the clinical and diagnostic criteria available.

Please note that the cases reported here are restricted to 'confirmed' and 'probable' cases; cases classified as 'possible' are not included.

Epidemiology

No cases of vCJD were reported in the EU/EEA in 2014. The overall mortality rate remains below 0.01 per 1 000 000 population in this long post-epidemic tail.

Table 1. Reported confirmed vCJD cases: number and rate per 100 000 population, EU/EEA, 2010–2014

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Country	2010	2011	2012	2013	2014			
	Confirmed Cases	Confirmed Cases	Confirmed Cases	Confirmed Cases	National data	Report type	Reported cases	Confirmed cases
Austria	0	0	0	0	Y	C	0	0
Belgium	.	.	0	0	Y	C	0	0
Bulgaria	0	0	0	0	Y	C	0	0
Croatia	.	.	0	0	.	.	.	.
Cyprus	.	.	0	0	Y	C	0	0
Czech Republic	0	0	0	0	Y	C	0	0
Denmark	0	0	0	.	Y	C	0	0
Estonia	0	0	0	0	Y	C	0	0
Finland	.	.	.	.	.	.	.	.
France	0	0	1	0	.	.	.	.
Germany	.	.	.	.	.	.	.	.
Greece	.	.	.	.	Y	C	0	0
Hungary	0	0	0	0	.	.	.	.
Iceland	0	0	0	0	Y	C	0	0
Ireland	0	0	0	0	Y	C	0	0
Italy	.	0	.	.	.	.	.	.
Latvia	0	0	0	0	.	.	.	.
Liechtenstein	.	.	.	.	.	.	.	.
Lithuania	0	0	0	0	Y	C	0	0
Luxembourg	0	0	0	0	Y	C	0	0
Malta	.	.	.	0	Y	C	0	0
Netherlands	.	.	.	.	.	.	.	.
Norway	.	.	.	0	.	.	.	.
Poland	0	0	0	0	Y	C	0	0
Portugal	0	0	0	0	Y	C	0	0
Romania	0	0	0	0	Y	C	0	0
Slovakia	.	.	0	0	Y	C	0	0
Slovenia	.	.	0	0	Y	C	0	0
Spain	0	0	0	0	Y	C	0	0
Sweden	.	.	0	0	Y	C	0	0
United Kingdom	3	1	0	1	.	.	.	.
EU / EEA	**3**	**1**	**1**	**1**	**.**	**C**	**0**	**0**

Source: Country reports. Legend: Y = yes, N = no, C = case based, A = aggregated, · = no data reported, ASR: age-standardised rate, - = no report

The vCJD epidemic peaked in the EU between 1999 and 2004 and has now reached its tail; vCJD has become a very rare neurodegenerative disease in the EU, due to the successful implementation of prevention and control measures aimed at the cattle trade (1989) and animal feed production (since 1994).

The estimated prevalence of vCJD infection is considered to be higher than the clinical case numbers suggest: A study on prevalence of abnormal prion protein in human appendixes conducted in the United Kingdom suggests a high prevalence of infections (493 cases per one million population) with abnormal PrP, indicating a higher than expected potential vCJD carrier status in the population [1].

All tested probable and confirmed vCJD clinical cases to date have been limited to a specific genotypic group: all are methionine homozygotes at codon 129, which typically is represented by approximately 40% of the European population. However, it remains uncertain if clinical cases of vCJD may also be seen in population groups with alternative codon 129 genotypes, and it remains a hypothesis that other polymorphisms may require prolonged incubation periods [2]. If this is the case, one could expect increasing vCJD cases numbers as the population that is potentially infected – but with genotypes that may confer longer incubation periods – grows older. It is therefore increasingly important that EU surveillance systems are able to capture CJD cases in all populations. This is challenging given the high background prevalence of dementia and other conditions characterised by neurological deterioration, which may mask vCJD clinical presentation.

The possibility remains that there is a silent pool of infected individuals that may be a source of secondary transmission through blood/organ donations. This has important implications in areas such as the management

of blood and blood products, tissue transplantation, cellular therapies, and the handling of surgical instruments [3-5].

The wider issues related to human transmissible spongiform encephalopathies (TSEs) continue to reveal issues of potential concern. For example, there is increasing evidence that protein misfolding is central in the causation of a range of other neurodegenerative disorders, including Alzheimer's and Parkinson's diseases [3,6]. The recent evidence suggests that such diseases could be 'seeded', raising the hypothesis that they could, in theory, be transmissible [7]. In addition, amendment of the TSE regulations and partial lifting of feed bans may also pose a renewed risk for human exposure in the long run. A study reporting that scrapie-infected transgenic mice present with phenotypic characteristics that are more aligned to sporadic CJD (sCJD) than vCJD [8] prompted EFSA to review their earlier scientific opinion concerning the zoonotic potential of ovine scrapie prions in 2015 [9]. The report concluded that the evidence for any causal link between scrapie and human TSEs remains absent, and no consistent risk factors have been identified for sCJD, but the possibility of scrapie-related public health risks from the consumption of ovine products could not be assessed. While the reporting of cases to TESSy is currently restricted for vCJD, the EuroCJD network continues to monitor occurrence of other forms of CJD as well as other human prion diseases.

Public health conclusions

Given the long incubation period of vCJD (over 10 years), continued TSE surveillance at the national and EU levels ensures that any variance in vCJD epidemiology can be detected. More generally, the remaining underlying uncertainties related to human prion disease aetiology – including the potential zoonotic risk from animal TSEs – and potentially changing risk profiles around all TSEs and other neurodegenerative diseases means that the continuation of detailed surveillance for all human prion diseases remains prudent [10]. From an EU perspective, there may be a need to discuss whether expanding mandatory EU surveillance and reporting to other forms of CJD would be helpful.

References

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Additional information

ECDC Surveillance Atlas of Infectious Diseases

Annex

Table. vCJD, surveillance systems overview, 2014

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* The European Surveillance System (TESSy) is a system for the collection, analysis and dissemination of data on communicable diseases. EU Member States and EEA countries contribute to the system by uploading their infectious disease surveillance data at regular intervals.