



SURVEILLANCE REPORT

Annual Epidemiological Report for 2016

Creutzfeldt–Jakob disease

Key facts

- Two cases of variant Creutzfeldt–Jakob disease (vCJD) were identified in the EU/EEA in 2016: one probable case from Italy and one confirmed case from the UK.
- The UK case was the first reported clinical vCJD patient with heterozygosity at codon 129 of the prion protein gene confirming that population groups carrying polymorphisms other than MM genotype are susceptible to clinical disease.
- vCJD disease remains extremely rare. This is consistent with the current understanding of the underlying epidemiology of vCJD and with the positive impact of risk mitigation measures introduced in the EU from the late 1980s to remove potential infectious animal material from the human food chain.

Methods

This report is based on data for 2016 retrieved from The European Surveillance System (TESSy) on 21 February 2018. TESSy is a system for the collection, analysis and dissemination of data on communicable diseases.

For a detailed description of methods used to produce this report, refer to the *Methods* chapter [1].

An overview of the national surveillance systems is available online [2].

A subset of the data used for this report is available through ECDC's online *Surveillance atlas of infectious diseases* [3].

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 done in accordance with the 2012 EU case definition.

The clinical presentation and associated diagnostic criteria for vCJD are relatively unusual. Suspected cases are typically reported to national surveillance centres. The centres offer diagnostic support and post-mortem analysis when needed. 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.

Cases reported here are restricted to 'confirmed' and 'probable' cases.

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Epidemiology

For 2016, Italy and the UK each reported a single case of vCJD. The overall mortality rate remains below 0.01 per 1 million population in this long post-epidemic tail.

Table 1. EU/EEA Member States reporting confirmed or probable vCJD cases, 2012–2016

Country	2012	2013	2014	2015	2016
	Reported cases	Reported cases	Reported cases	Reported cases	Reported cases
France	1	0	0	0	0
Italy	-	-	-	-	1
United Kingdom	0	1	0	0	1
EU/EEA	1	1	0	0	2

-: rate not calculated.

Discussion

The vCJD epidemic peaked in the EU from 1999–2004 and has now reached its tail [4]. vCJD has become a very rare neurodegenerative disease in the EU/EAA. This is due to the successful implementation of prevention and control measures to remove Bovine Spongiform Encephalopathy (BSE) prions from the animal and human food chains aimed at the cattle trade (1989) and animal feed production (since 1994). Although two cases were identified in 2016, this is consistent with a declining and increasingly rare condition.

The estimated prevalence of vCJD infection may be higher than the clinical case numbers suggest. A study on prevalence of abnormal prion protein in human appendixes conducted in the UK suggests a high prevalence of infection (493 cases per one million population) with abnormal prion protein, indicating a higher-than-expected potential vCJD carrier status in the population [5]. Hence, there may be a hidden population of infected individuals that may cause secondary transmission through blood and/or 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 [6–8].

In 2016, the United Kingdom reported the first confirmed vCJD case in a clinical patient expressing heterozygosity at codon 129 of the prion protein gene [9]. All previous vCJD cases have been restricted to populations that were homozygous for the methionine (M) allele at codon 129, carried by approximately 40% of the EU population. This first clinical case carrying both the M and valine (V) allele at codon 129 was expected as animal transmission studies had shown that heterozygote populations could become ill with vCJD [7]. There was also evidence that heterozygosity in infected individuals may extend the preclinical incubation period. Accordingly, the MV case could represent the first case of a so-called 'second wave' of cases as the population potentially infected, but with genotypes that may confer longer incubation periods, grows older [10].

Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy (TSE) that effects cervids (deer, elk and moose). The first two cases of CWD in wild animal populations in the EU/EEA were reported in Norway in 2016. There is no current scientific evidence that humans can contract the disease through eating meat from infected animals, but to address animal health concerns, EFSA adopted a scientific opinion on CWD in cervids in December 2016 [11] that recommended monitoring activities and measures to prevent the introduction and spread of the disease in EU animal populations.

The nature of CJD infection implies that the clinical presentation of disease in infected patients exposed through non-dietary routes or an infectious agent that is not BSE-derived may differ from that of vCJD. Although TESSy supports data collection of vCJD cases, continued monitoring of the occurrence of other forms of CJD and other human prion diseases is important in order to identify possible sources of public health risk.

Public health conclusions

Public health measures are developed on the basis that all population groups are susceptible to infection and clinical disease, so the finding of the first clinical patient with an MV genotype requires no change in public health action at this stage. However, the long incubation period of vCJD (over 10 years) requires continued surveillance at the national and EU levels and the MV case emphasises the need to monitor disease epidemiology and pathology to ensure that the risk profile of the vCJD does not alter and any change in vCJD epidemiology can be detected. More generally, given the remaining 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, it is prudent to continue detailed surveillance for all human prion diseases [4].

References

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