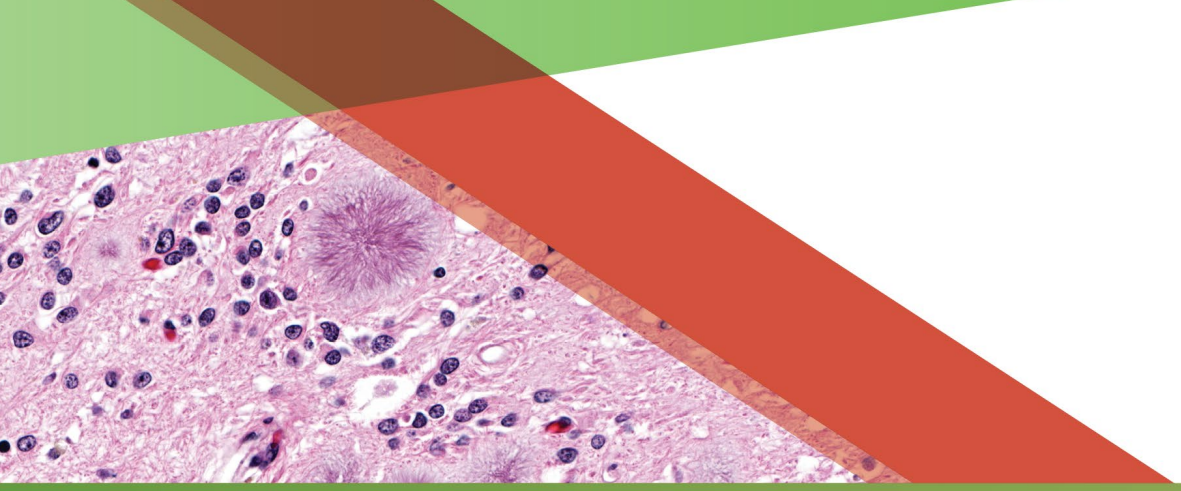

SURVEILLANCE REPORT

Variant Creutzfeldt–Jakob disease

Annual Epidemiological Report for 2017

Key facts

- No cases of variant Creutzfeldt–Jakob disease (vCJD) were identified in the EU/EEA in 2017.
- The 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 in the late 1980s to remove potential infectious animal material from the human food chain.

Methods

This report is based on data for 2017 retrieved from The European Surveillance System (TESSy) on 30 October 2019. 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 available clinical and diagnostic criteria.

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

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Stockholm, September 2020

Epidemiology

For 2017, no cases of confirmed or probable vCJD were reported in the EU. This is consistent with an overall mortality rate of 0.01 per 1 million population in this long post-epidemic tail.

Table 1. EU/EEA Member States reporting confirmed or probable vCJD cases, 2013–2017

Country	2013	2014	2015	2016	2017
	Reported cases	Reported cases	Reported cases	Reported cases	Reported cases
France	0	0	0	0	0
Italy	-	-	-	1	0
United Kingdom	1	0	0	1	0
Portugal	0	0	0	0	0
EU/EEA	**1**	**0**	**0**	**2**	**0**

-: rate not calculated.

Discussion

The vCJD epidemic peaked in the EU/EEA from 1999–2004 and has since tailed off [4], such that 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). The absence of cases of vCJD identified in 2017 is consistent with a declining and increasingly rare condition.

Some uncertainty remains regarding the future epidemiology and public health risk from vCJD. Studies on the prevalence of abnormal prion protein in human appendixes conducted in the United Kingdom (UK) suggest 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]. Furthermore, in 2016 the first confirmed vCJD case in a patient expressing heterozygosity at codon 129 of the prion protein gene was identified [9]. It has been hypothesised that heterozygotes with the methionine/valine (MV) genotype at 129, which make up approximately 50% of the EU population, may be potentially susceptible to infection, but that the MV genotype may confer longer incubation periods [10]. If the hypothesis were corroborated by empirical evidence in human populations, it would have 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].

Because vCJD is associated with the transmission of BSE from infected animals, assessing the ongoing epidemiology of prion diseases in animals and potential zoonotic transmission remain important for public health. Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy (TSE) that effects cervids (deer, elk and moose). The first cases of CWD in wild animal populations in the EU/EEA were reported in Norway in 2016 [11]. No cases of human prion disease have to date been associated with CWD, but in response to the identification of CWD-positive animals in the EU/EEA, the European Food Safety Authority (EFSA) produced a scientific opinion in January 2017 to identify monitoring activities and measures to prevent the introduction and spread of chronic wasting disease into and within the EU [12]. In July 2017, EFSA published a scientific opinion on the origin of the 60 cases of classical BSE reported in cattle born after the enforcing of the EU ban on the use of animal proteins in livestock feed in 2001. Experts concluded that contaminated feed is the most likely source of infection. A second possibility was that contaminated feed ingredients have been imported from non-EU countries. The experts could not rule out other causes due to the difficulty of investigating individual cases [13].

The nature of CJD infection suggests that the clinical presentation of the disease in infected patients exposed through non-dietary routes or from a source other than BSE 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 other possible sources of public health risk.

Public health conclusions

Public health measures are developed on the basis that all population groups are equally susceptible to infection and clinical disease, but some uncertainties on CJD infection and transmission risk remains, including human prion disease aetiology, potential zoonotic risk from animal TSEs, and potentially changing risk profiles around all TSEs and other neurodegenerative diseases. Surveillance at the national and EU levels provides confidence that the public health risk profile of vCJD remains unaltered, and that any changes can be detected [4].

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