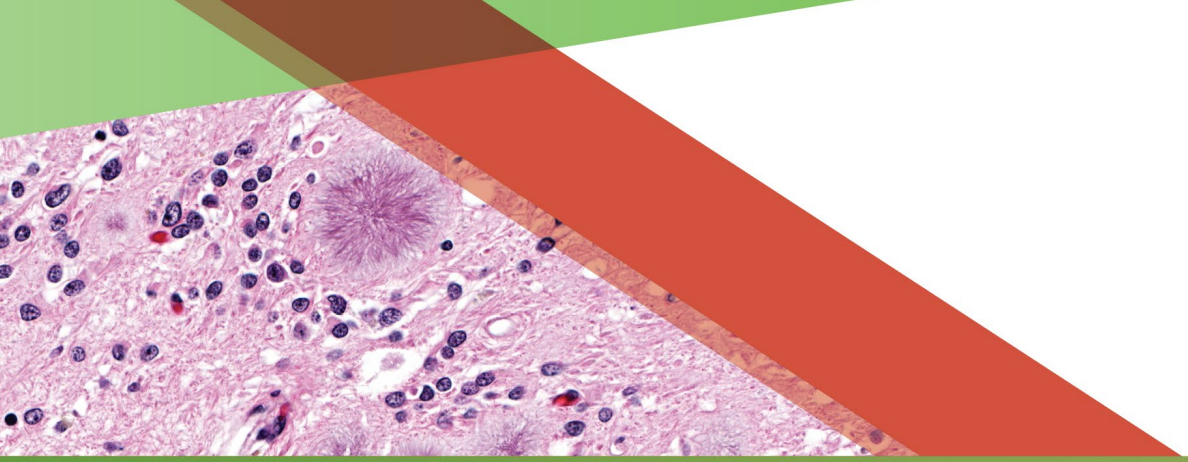

SURVEILLANCE REPORT

Variant Creutzfeldt–Jakob disease

Annual Epidemiological Report for 2018

Key facts

- One confirmed case of variant Creutzfeldt–Jakob disease (vCJD) was identified in the EU/EEA (France) in 2018.
- 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 potentially infectious animal material from the human food chain.

Introduction

Variant Creutzfeldt–Jakob disease (vCJD) is a rare neurological disease caused by prions composed of misfolded prion proteins (PrPSc). This form of prion aggregates in neurological tissue leading to progressive brain damage, causing psychiatric or sensory symptoms, neurological abnormalities and eventual death. The disease was first identified in the United Kingdom, and in March 1996, an association was identified between vCJD and the consumption of products from animals infected with bovine spongiform encephalopathy (BSE) or the mad cow disease [1].

Methods

This report is based on data for 2018 retrieved from The European Surveillance System (TESSy) on 17 September 2019. TESSy is a system for the collection, analysis and dissemination of data on communicable diseases.

For a detailed description of the methods used to produce this report, refer to the 'Methods' chapter in the 'Introduction to the Annual Epidemiological Report' [2].

An overview of the national surveillance systems is available online [3].

A subset of the data used for this report is available through ECDC's online *Surveillance Atlas of Infectious Diseases* [4].

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 2018 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).

Suggested citation: European Centre for Disease Prevention and Control. Variant Creutzfeldt–Jakob disease. In: ECDC. Annual Epidemiological Report for 2018. Stockholm: ECDC; 2022.

Stockholm, October 2022

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.

Epidemiology

For 2018, France reported one confirmed case of vCJD. The case first displayed symptoms and thereafter underwent initial investigation and diagnosis in 2018, succumbing to the disease in 2019. A full case study and post-mortem neuropathological investigation confirmed the presence of variant CJD. A possible source of exposure in this case was plausibly related to accidental occupational exposure in a laboratory. The individual had handled murine samples contaminated with the agent that causes bovine spongiform encephalopathy (BSE) 7.5 years earlier [5].

The overall mortality rate remains below 0.01 cases per one million population in this long post-epidemic tail.

Table 1. Distribution of variant Creutzfeldt–Jakob disease cases and rates per 100 000 population by country, EU/EEA, 2014–2018

	2014	2015	2016	2017	2018
Country	Reported cases	Reported cases	Reported cases	Reported cases	Reported cases
Austria	0	0	0	0	0
Belgium	0	0	0	0	0
Bulgaria	0	0	0	0	0
Croatia	-	0	0	0	0
Cyprus	0	0	0	0	0
Czechia	0	0	0	0	0
Denmark	0	0	0	0	0
Estonia	0	0	0	0	0
Finland	ND	ND	ND	ND	ND
France	0	0	0	0	1
Germany	ND	ND	ND	ND	ND
Greece	0	0	0	0	0
Hungary	0	0	0	0	0
Iceland	0	0	0	0	0
Ireland	0	0	0	0	0
Italy	-	-	1	0	0
Latvia	0	0	0	0	0
Liechtenstein	ND	ND	ND	ND	ND
Lithuania	0	0	0	0	0
Luxembourg	-	-	0	0	0
Malta	0	0	0	0	0
Netherlands	0	0	0	0	0
Norway	0	0	0	0	0
Poland	0	0	0	0	0
Portugal	0	0	0	0	0
Romania	0	0	0	0	0
Slovakia	0	0	0	0	0
Slovenia	0	0	0	0	0
Spain	0	0	0	0	0
Sweden	0	0	0	0	0
United Kingdom	0	0	1	0	0
EU/EEA	**0**	**0**	**2**	**0**	**1**

Source: Country reports
-: no cases reported
ND: no data reported

Discussion

The vCJD epidemic peaked in the EU from 1999–2004 and has now reached its tail [6]. vCJD has become a very rare neurodegenerative disease in the EU/EEA. 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 (since 1989) and animal feed production (since 1994). The single case of reported vCJD in 2018 is consistent with a declining and increasingly rare condition.

Further investigations on the possible cause of the vCJD case reported by France in 2018 indicate that one plausible source of infection was an accidental occupational exposure in a laboratory environment. This is the second vCJD case in recent history that has an association with laboratory exposure to transmissible spongiform encephalopathy (TSE) agents following a vCJD case reported in Italy in 2016 who also had occupational contact with BSE-infected brain tissue. These two cases in a relatively close time span highlight that occupational exposure from transmissible prion agents remains a viable route of transmission [5].

More generally, there remains some uncertainty on the epidemiology and public health risk from vCJD. Studies on prevalence of abnormal prion protein in human appendixes conducted in the United Kingdom (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 [7].

Furthermore, in 2016, the first confirmed vCJD case in a clinical patient expressing heterozygosity at codon 129 of the prion protein gene was identified [8]. It is suggested that MV heterozygotes, 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 [9]. Hence, there may be a hidden population of infected individuals which may develop the disease or 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 [10-12].

Public health implications

Public health measures are developed on the basis that all population groups are susceptible to vCJD infection and clinical disease, and the continued absence of clinical cases of vCJD in the EU gives confidence that EU-wide protection measures against prion disease infection continue to be effective. However, some uncertainties remain. The consequences of the extended incubation periods and the absence of a rapid diagnostic test means that the infection status of the EU population is unclear. Hence prion transmission and circulation within human populations remains a potential risk.

The recent cases of vCJD infection associated with occupational exposure in laboratories also emphasise the potential risks from accidental infection when handling TSE-infected material, and the limitations of knowledge on optimal post-exposure decontamination procedures following accidental exposure. As no effective preventative treatment has been described following such exposure, there is a need for rigorous health and safety procedures to minimise the risk of accidental exposure for staff working in environments with TSE-infected material [5, 13].

The evolving epidemiology of TSEs in animal populations and potential zoonotic risk from animal TSEs, and the changing risk profiles around all TSEs and other neurodegenerative diseases also create some uncertainty for public health. Therefore, it is important that human and animal surveillance at the national and EU-levels continues to provide assurances that public health measures to minimise the risk of vCJD infection in EU populations are effective, risk profiles from vCJD and other prion diseases remain unaltered and changes can be detected [6].

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