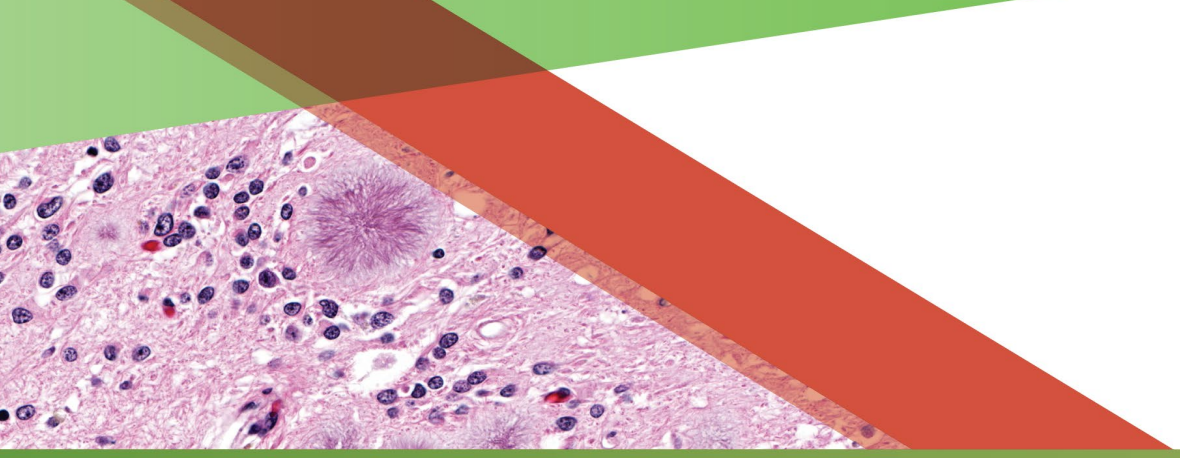

SURVEILLANCE REPORT

Variant Creutzfeldt–Jakob disease

Annual Epidemiological Report for 2020

Key facts

- No cases of variant Creutzfeldt–Jakob disease (vCJD) were identified in the EU/EEA in 2020.
- vCJD disease remains extremely rare. This is consistent with the current understanding of the underlying epidemiology of the disease, 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 prion disease, a group of rare neurological diseases caused by abnormal misfolded prion proteins (PrPSc). These abnormal prions accumulate in the brain and lead to progressive brain damage, causing psychiatric or sensory symptoms, neurological abnormalities and eventual death. The disease vCJD 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 mad cow disease [1].

Methods

This report is based on data for 2020 retrieved from The European Surveillance System (TESSy) on 5 November 2021. 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 [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 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).

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Stockholm, March 2023.

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 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

No cases of vCJD were reported in the EU/EEA in 2020. The overall mortality rate remains below 0.01 cases per one million population at the tail-end of this long post-epidemic period.

Table 1. Distribution of confirmed variant Creutzfeldt-Jakob disease cases by country and year, EU/EEA, 2016–2020

Country	2016	2017	2018	2019	2020
	Number		Number		Number
Austria	0	0	0	0	0
Belgium	0	0	0	0	0
Bulgaria	0	0	0	NR	NR
Croatia	0	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	1	0	0
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	0	0
Latvia	0	0	0	0	0
Liechtenstein	ND	ND	ND	ND	ND
Lithuania	0	0	0	NR	0
Luxembourg	0	0	0	0	0
Malta	0	0	0	NR	NR
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
UK	1	0	0	0	NR
EU-EEA	**2**	**0**	**1**	**0**	**0**

Source: Country reports.
ND: no data reported
NR: Not reported.

Discussion

The vCJD epidemic peaked in the EU during the period 1999–2004, but the number of cases have subsequently decreased. No cases were reported in 2020 and 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, mainly by targeting the cattle trade (since 1989) and animal feed production (since 1994).

There remains some uncertainty concerning the 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 was identified in a clinical patient expressing heterozygosity at codon 129 of the prion protein gene [6]. 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 [7]. Therefore there may be a hidden population of infected individuals who 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 [8-10].

As vCJD is associated with the transmission of BSE from infected animals, assessment of ongoing epidemiology of prion diseases in animals, and potential zoonotic transmission remains important for public health. Consequently, the EU Member States continue to implement an annual targeted surveillance programme to assess the prevalence of transmissible spongiform encephalopathy (TSE) infection in animal populations, coordinated by the European Food Safety Authority (EFSA). In 2020, a total of 1 122 671 cattle were tested by EU Member States and the UK, and a total of four animals were identified with atypical BSE by three reporting countries: France (one H-BSE and one L-BSE), Spain (one H-BSE) and Ireland (one H-BSE). These animals were not destined for the human food chain, and so presented no direct risk to public health. Overall, the low prevalence of positive cases identified by EU surveillance strongly indicates that there are very few BSE-infected animals in EU cattle populations. Therefore the public health risk of vCJD infection from consumption of beef in the EU appears low. All four cases are also classified as 'atypical BSE'. The origin of such cases is unclear, but the pathology differs from the 'classical BSE' associated with consumption of contaminated feed, which is believed to be the source of the primary BSE epidemic and subsequent causal association with vCJD cases.

Following the identification in 2016 of the first case of chronic wasting disease (CWD) – a transmissible spongiform encephalopathy (TSE) that affects cervids (deer, elk and moose) in wild European cervid populations [11], six Member States (Estonia, Finland, Latvia, Lithuania, Poland and Sweden) began EU-mandated surveillance in cervid populations in 2020 as part of a three-year targeted surveillance programme [12]. This resulted in the testing of 6 974 cervids in the EU, and confirmation of two CWD cases in wild moose (Eurasian/European elk) in 2020; one in Sweden and one in Finland. Further details of TSE surveillance in EU animal populations in 2020 is available in 'The European Union summary report on surveillance for the presence of transmissible spongiform encephalopathies (TSE) in 2020' [13]. In 2020, EFSA also launched a storymap and interactive dashboards presenting data on TSEs in the EU Member States and other reporting countries.

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 extended incubation periods mean there might be decades between infection and clinical manifestation of vCJD. The absence of a rapid diagnostic test means that the infection status of the EU population is unclear. In addition, 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. Therefore there is still a potential risk prion of transmission and circulation within human populations.

The evolving epidemiology of TSEs in animal populations and the potential zoonotic risk from animal TSEs has also created some uncertainty for public health. Although TESSy supports data collection of vCJD cases, continued human and animal surveillance at the national and EU level is important to monitor all forms of CJD and other human prion diseases in order to identify possible sources of public health risk. Monitoring will provide assurances that public health measures to minimise risk of vCJD infection in EU populations are effective, that risk profiles from vCJD and other prion diseases remain unaltered and that changes with potential impact on public health can be detected [14].

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