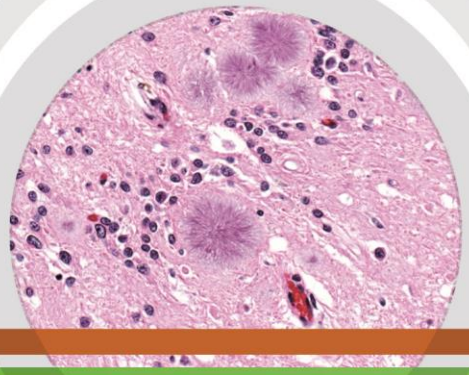

SURVEILLANCE REPORT

Annual Epidemiological Report for 2015

Variant Creutzfeldt–Jakob disease

Key facts

- No new confirmed cases of variant Creutzfeldt–Jakob disease (vCJD) were reported in 2015.
- The disease is extremely rare, which is consistent with the current understanding of the underlying epidemiology of vCJD and the positive impact of risk mitigation measures introduced in the EU in the late 1980s to remove potentially infectious animal material from the human food chain.
- 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 mean that continued disease 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

This report is based on data for 2015 retrieved from The European Surveillance System (TESSy) on 15 November 2016. 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, please 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].

In 2012, ECDC took over disease 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 carried out through TESSy, in accordance with to the EU-2012 case definition.

The clinical presentation and associated diagnostic criteria for vCJD are rather unusual. Suspected cases are typically reported to national surveillance centres. The centres offer diagnostic support and post-mortem analysis if 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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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, which means that 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 2015. The overall mortality rate remains below 0.01 cases per 1 000 000 population in this long post-epidemic tail.

Table 1. Distribution of confirmed cases of vCJD, EU/EEA, 2011–2015

Country	2011		2012		2013		2014		2015				
	Confirmed cases		Confirmed cases		Confirmed cases		Confirmed cases		National	Confirmed	Reported cases		
	Number	Rate	Number	Rate	Number	Rate	Number	Rate	coverage	cases	Number	Rate	ASR
France	1	-	1	-	0	-	0	-	Y	0	0	-	-
Romania	0	-	0	-	0	-	1	-	Y	0	0	-	-
United Kingdom	2	-	0	-	1	-	0	-	Y	0	0	-	-
EU/EEA	3	-	1	-	1	-	1	-	.	0	0	-	-

Source: Country reports. Legend: Y = yes, N = no, C = case based, A = aggregated, · = no data reported, ASR = age-standardised rate, - = no report
[1] Provisional data for 2015. Notification rates not calculated.

Discussion

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 (PrP) 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 [4].

Virtually all probable and confirmed vCJD clinical cases to date have been limited to a specific genotypic group, which typically is represented by approximately 40% of the European population. However, populations with other polymorphisms are also suspected to develop the disease [5]. It has been hypothesised that other polymorphisms confer extended incubation periods in those infected and 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 [6-8].

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 [6,9]. The recent evidence suggests that such diseases could be 'seeded', raising the hypothesis that they could, in theory, be transmissible [10]. In addition, amendment of the TSE regulations and partial lifting of feed bans may also pose a renewed risk for human exposure. A study reporting that scrapie-infected transgenic mice present with phenotypic characteristics that are more aligned to sporadic CJD (sCJD) than vCJD [11] prompted EFSA to review their earlier scientific opinion concerning the zoonotic potential of ovine scrapie prions in 2015 [12]. The report concluded that there is no evidence of a causal link between scrapie and human TSEs. No consistent risk factors were identified for sCJD, and it was not possible to assess scrapie-related public health risks related to the consumption of ovine products. While TESSy currently only records vCJD case reports, the EuroCJD network also monitors other forms of CJD and human prion diseases in general.

Public health implications

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 [13]. 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.

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