**CREUTZFELDT-JAKOB DISEASE (CJD) AND RELATED DISEASES IN ITALY**

**(As at30September 2021)**

|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **REFERRALS OF SUSPECTED**  **CASES** | | **DEATHS OF DEFINITE AND PROBABLE CJD** | | | | | | | |
| *Year* | *Referrals* | *Year* | *Sporadic CJD* | *Genetic forms* | | | *Iatrogenic CJD3* | *Variant CJD4* | ***Total Deaths*** |
| *Genetic CJD* | *GSS1* | *FFI2* |
| 1993 | 51 | 1993 | 27 | 6 | 1 | 2 | 0 | 0 | **36** |
| 1994 | 62 | 1994 | 33 | 7 | 0 | 1 | 0 | 0 | **41** |
| 1995 | 52 | 1995 | 28 | 6 | 2 | 1 | 0 | 0 | **37** |
| 1996 | 77 | 1996 | 51 | 8 | 1 | 1 | 0 | 0 | **61** |
| 1997 | 139 | 1997 | 49 | 12 | 0 | 1 | 1 | 0 | **63** |
| 1998 | 143 | 1998 | 63 | 10 | 0 | 1 | 2 | 0 | **76** |
| 1999 | 192 | 1999 | 74 | 20 | 0 | 0 | 0 | 0 | **94** |
| 2000 | 177 | 2000 | 60 | 23 | 4 | 3 | 0 | 0 | **90** |
| 2001 | 219 | 2001 | 87 | 13 | 1 | 0 | 0 | 0 | **101** |
| 2002 | 201 | 2002 | 76 | 14 | 3 | 0 | 0 | 0 | **93** |
| 2003 | 192 | 2003 | 81 | 9 | 1 | 1 | 0 | 1 | **93** |
| 2004 | 176 | 2004 | 77 | 21 | 3 | 0 | 0 | 0 | **101** |
| 2005 | 236 | 2005 | 108 | 20 | 4 | 0 | 1 | 0 | **133** |
| 2006 | 238 | 2006 | 97 | 32 | 6 | 2 | 1 | 0 | **138** |
| 2007 | 208 | 2007 | 99 | 18 | 2 | 0 | 1 | 0 | **120** |
| 2008 | 217 | 2008 | 96 | 12 | 1 | 1 | 0 | 0 | **110** |
| 2009 | 191 | 2009 | 109 | 14 | 2 | 0 | 0 | 0 | **125** |
| 2010 | 216 | 2010 | 111 | 16 | 8 | 0 | 1 | 0 | **136** |
| 2011 | 220 | 2011 | 121 | 24 | 3 | 0 | 0 | 1 | **149** |
| 2012 | 230 | 2012 | 117 | 26 | 1 | 0 | 1 | 0 | **145** |
| 2013 | 234 | 2013 | 103 | 30 | 2 | 0 | 0 | 0 | **135** |
| 2014 | 245 | 2014 | 112 | 11 | 4 | 1 | 1 | 0 | **129** |
| 2015 | 280 | 2015 | 124 | 31 | 6 | 0 | 0 | 0 | **161** |
| 2016 | 244 | 2016 | 109 | 22 | 2 | 0 | 0 | 1 | **134** |
| 2017 | 305 | 2017 | 156 | 25 | 2 | 1 | 1 | 0 | **185** |
| 2018 | 295 | 2018 | 144 | 20 | 2 | 1 | 0 | 0 | **167** |
| 2019 | 306 | 2019 | 111 | 19 | 0 | 1 | 0 | 0 | **131** |
| 2020 | 275 | 2020 | 97 | 13 | 3 | 0 | 1 | 0 | **114** |
| 2021 | 246 | 2021 | 28 | 3 | 0 | 1 | 0 | 0 | **32** |
| ***Total referrals*** | **5868** | ***Total deaths*** | **2548** | **485** | **64** | **19** | **11** | **3** | **3130** |

1GSS, Gerstmann-Sträussler-Scheinker disease

2FFI, Fatal Familial insomnia

3All iatrogenic CJD cases are related to dura mater implants.

4All patients were likely infected in Italy through the consumption of BSE-contaminated food. However, the patient deceased in 2016 had been working in a prion research laboratory where he/she had been in contact with human brain tissues infected with variant CJD and bovine brain tissues infected with typical and atypical BSE. Therefore, it cannot be excluded that this patient was infected during laboratory routine activities, although epidemiological investigations failed to reveal any reported laboratory accident.