

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

(As at 31 August 2021)

REFERRALS OF SUSPECTED CASES		DEATHS OF DEFINITE AND PROBABLE CJD							
Year	Referrals	Year	Sporadic CJD	Genetic forms			Iatrogenic CJD ³	Variant CJD ⁴	Total Deaths
				Genetic CJD	GSS ¹	FFI ²			
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	154	25	2	1	1	0	183
2018	295	2018	143	20	2	1	0	0	166
2019	306	2019	106	17	0	1	0	0	124
2020	275	2020	94	12	3	0	1	0	110
2021	221	2021	21	3	0	1	0	0	25
Total referrals	5842	Total deaths	2530	482	64	19	11	3	3109

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.