

National Prion Disease Pathology Surveillance Center Cases Examined¹

(September 2nd, 2023)

Year	Total Referrals ²	Prion Disease	Sporadic	Familial	iCJD	vCJD
1999 & earlier	383	232	202	27	3	0
2000	145	102	90	12	0	0
2001	209	118	110	8	0	0
2002	241	144	124	18	2	0
2003	259	160	137	21	2	0
2004	315	180	163	16	0	1 ³
2005	330	179	157	21	1	0
2006	365	179	159	17	1	2 ⁴
2007	374	210	191	19	0	0
2008	384	221	205	16	0	0
2009	397	231	210	20	1	0
2010	402	247	219	28	0	0
2011	392	238	214	24	0	0
2012	413	244	221	23	0	0
2013	416	258	223	34	1	0
2014	355	208	185	21	1	1 ⁵
2015	401	262	243	19	0	0
2016	395	277	248	29	0	0
2017	375	266	247	19	0	0
2018	308	221	202	18	1	0
2019	434	281	259	22	0	0
2020	367	253	228	24	1	0
2021	344	248	224	22	0	0
2022	337	228	206	21	0	0
2023	193	114	101	8	0	0
TOTAL	8534⁶	5301⁷	4768⁸	507⁹	14	4

Year	CSF Only & RT-QuIC Positive ¹⁰
2015	140
2016	187
2017	227
2018	266
2019	310
2020	309
2021	331
2022	347
2023	240
TOTAL	2357

¹ Listed based on the year of death or, if not available, on year of referral; ² Cases with suspected prion disease for which brain tissue and/or blood (in familial cases) were submitted; ³ Disease acquired in the United Kingdom; ⁴ Disease acquired in the United Kingdom in one case and in Saudi Arabia in the other; ⁵ Disease possibly acquired in a Middle Eastern or Eastern European country; ⁶ Includes 28 cases in which the diagnosis is pending (1 from 2022, and 27 from 2023), and 20 inconclusive cases; ⁷ Includes 8 (2 from 2021, 1 from 2022 and 5 from 2023) cases with type determination pending in which the diagnosis of vCJD has been excluded. ⁸ The sporadic cases include 4641 cases of sporadic Creutzfeldt-Jakob disease (sCJD), 88 cases of Variably Protease-Sensitive Prionopathy (VPSPr) and 39 cases of sporadic Fatal Insomnia (sFI). ⁹ Total does not include 312 Familial cases diagnosed by blood only. ¹⁰ Lists number of patients who have had a positive RT-QuIC and no neuropath examination.