

National Prion Disease Pathology Surveillance Center Cases Examined<sup>1</sup>  
(September 20, 2022)

Year	Total Neuropath Referrals <sup>2</sup>	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 <sup>3</sup>
2005	328	179	157	21	1	0
2006	365	179	159	17	1	2 <sup>4</sup>
2007	374	210	191	19	0	0
2008	384	221	205	16	0	0
2009	397	231	210	20	1	0
2010	401	246	218	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 <sup>5</sup>
2015	401	263	243	20	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	365	252	227	24	1	0
2021	343	248	223	22	0	0
2022	213	124	98	9	0	0
TOTAL	8211 <sup>6</sup>	5082 <sup>7</sup>	4556 <sup>8</sup>	488 <sup>9</sup>	14	4

Year	CSF Only & RT-QuIC Positive <sup>10</sup>
2015	140
2016	183
2017	227
2018	266
2019	311
2020	310
2021	341
2022	262
TOTAL	2040

<sup>1</sup> Listed based on the year of death or, if not available, on year of referral; <sup>2</sup> Cases with suspected prion disease for which brain tissue and/or blood (in familial cases) were submitted; <sup>3</sup> Disease acquired in the United Kingdom; <sup>4</sup> Disease acquired in the United Kingdom in one case and in Saudi Arabia in the other; <sup>5</sup> Disease possibly acquired in a Middle Eastern or Eastern European country; <sup>6</sup> Includes **28** cases in which the diagnosis is pending (**1** from **2020**, **3** from **2021** and **24** from **2022**), and **20** inconclusive cases;<sup>7</sup> Includes **20** (**3** from **2021** and **17** from **2022**) cases with type determination pending in which the diagnosis of vCJD has been excluded. <sup>8</sup> The sporadic cases include **4437** cases of sporadic Creutzfeldt-Jakob disease (sCJD), **82** cases of Variably Protease-Sensitive Prionopathy (VPSPr) and **37** cases of sporadic Fatal Insomnia (sFI). <sup>9</sup> Total does not include **300** Familial cases diagnosed by blood only. <sup>10</sup> Lists number of patients (deceased and alive) who have had a positive RT-QuIC and no neuropath examination.