

National Prion Disease Pathology Surveillance Center Cases Examined¹

(September 18, 2018)

Year	Total Referrals ²	Prion Disease	Sporadic	Familial	Iatrogenic	vCJD
1998 & earlier	259	157	135	20	2	0
1999	121	73	65	7	1	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	316	181	164	16	0	1 ³
2005	327	178	156	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	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 ⁵
2015	402	264	244	20	0	0
2016	397	278	248	29	0	0
2017	373	266	243	19	0	0
2018	188	129	114	7	0	0
TOTAL	6734	4085⁷	3656⁸	400⁹	12	4

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 was 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 12 cases in which the diagnosis is pending, and 20 inconclusive cases; ⁷ Includes 13 (8 from 2018) cases with type determination pending in which the diagnosis of vCJD has been excluded. ⁸ The sporadic cases include 3560 cases of sporadic Creutzfeldt-Jakob disease (sCJD), 63 cases of Variably Protease-Sensitive Prionopathy (VPSPr) and 33 cases of sporadic Fatal Insomnia (sFI). ⁹ Total does not include 247 Familial cases diagnosed by blood test only.