

# National Prion Disease Pathology Surveillance Center Cases Examined<sup>1</sup>

(January 11, 2023)

Year	Total Referrals <sup>2</sup>	Prion Disease	Sporadic	Familial	iCJD	vCJD
<b>1999 &amp; earlier</b>	383	232	202	27	3	0
<b>2000</b>	145	102	90	12	0	0
<b>2001</b>	209	118	110	8	0	0
<b>2002</b>	241	144	124	18	2	0
<b>2003</b>	259	160	137	21	2	0
<b>2004</b>	315	180	163	16	0	1 <sup>3</sup>
<b>2005</b>	330	179	157	21	1	0
<b>2006</b>	365	179	159	17	1	2 <sup>4</sup>
<b>2007</b>	374	210	191	19	0	0
<b>2008</b>	384	221	205	16	0	0
<b>2009</b>	397	231	210	20	1	0
<b>2010</b>	402	246	218	28	0	0
<b>2011</b>	392	238	214	24	0	0
<b>2012</b>	413	244	221	23	0	0
<b>2013</b>	416	258	223	34	1	0
<b>2014</b>	355	208	185	21	1	1 <sup>5</sup>
<b>2015</b>	401	263	243	20	0	0
<b>2016</b>	395	277	248	29	0	0
<b>2017</b>	375	266	247	19	0	0
<b>2018</b>	308	221	202	18	1	0
<b>2019</b>	433	280	259	21	0	0
<b>2020</b>	366	252	227	24	1	0
<b>2021</b>	343	248	223	22	0	0
<b>2022</b>	307	199	165	13	0	0
<b>TOTAL</b>	<b>8308<sup>6</sup></b>	<b>5156<sup>7</sup></b>	<b>4623<sup>8</sup></b>	<b>491<sup>9</sup></b>	<b>14</b>	<b>4</b>

Year	CSF Only & RT-QuIC Positive <sup>10</sup>
<b>2015</b>	241
<b>2016</b>	360
<b>2017</b>	406
<b>2018</b>	431
<b>2019</b>	538
<b>2020</b>	494
<b>2021</b>	516
<b>2022</b>	492
<b>TOTAL</b>	<b>3478</b>

<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 **25** cases in which the diagnosis is pending (**1** from **2020**, **2**, **2** from **2021**, and **21** from **2022**), and **20** inconclusive cases; <sup>7</sup> Includes **24** (**3** from **2021** and **21** from **2022**) cases with type determination pending in which the diagnosis of vCJD has been excluded. <sup>8</sup> The sporadic cases include **4504** 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 **301** Familial cases diagnosed by blood only. <sup>10</sup> Lists number of patients who have had a positive RT-QuIC and no neuropath examination.