

Creutzfeldt-Jakob Disease Incidence, South Korea, 2001–2019

Appendix

Appendix Table 1. Classification of germline mutations of familial Creutzfeldt-Jakob disease, South Korea, 2008–2019

| Year | Total, no. (%) | Mutation, no. (%) | | | | |
|-----------|----------------|-------------------|------------|------------|----------|-----------|
| | | D178N-M129V | V180I | E200K | V203I | M232R |
| 2008–2010 | 4 (12.90) | 0 | 1 (25) | 1 (25) | 1 (25) | 1 (25) |
| 2011–2015 | 12 (38.71) | 1 (8.33) | 5 (41.67) | 6 (50) | 0 | 0 |
| 2016–2019 | 15 (48.39) | 0 | 5 (33.33) | 4 (26.67) | 0 | 6 (40) |
| Total | 31 (100) | 1 (3.23) | 11 (35.48) | 11 (35.48) | 1 (3.23) | 7 (22.58) |

Appendix Table 2. Annual number of Creutzfeldt-Jakob disease cases by age group, South Korea, 2011–2019

| Year | Total, no. (%) | Age group, y; no. (%) | | | | | p value |
|-------|----------------|-----------------------|-----------|------------|-------------|-------------|---------------|
| | | 30–39 | 40–49 | 50–59 | 60–69 | >70 | |
| 2011 | 29 (7.36) | 0 | 5 (17.24) | 3 (10.34) | 14 (48.28) | 7 (24.14) | 0.0307 |
| 2012 | 45 (11.42) | 2 (4.44) | 2 (4.44) | 9 (20) | 13 (28.89) | 19 (42.22) | 0.5733 |
| 2013 | 34 (8.63) | 1 (2.94) | 2 (5.88) | 6 (17.65) | 10 (29.41) | 15 (44.12) | 0.9342 |
| 2014 | 65 (16.5) | 0 | 2 (3.08) | 16 (24.62) | 18 (27.69) | 29 (44.62) | 0.5429 |
| 2015 | 33 (8.38) | 0 | 0 | 5 (15.15) | 14 (42.42) | 14 (42.42) | 0.4847 |
| 2016 | 43 (10.91) | 0 | 4 (9.3) | 8 (18.6) | 10 (23.26) | 21 (48.84) | 0.5660 |
| 2017 | 38 (9.64) | 0 | 2 (5.26) | 9 (23.68) | 11 (28.95) | 16 (42.11) | 0.9022 |
| 2018 | 54 (13.71) | 1 (1.85) | 3 (5.56) | 10 (16.67) | 20 (35.19) | 20 (38.89) | 0.9640 |
| 2019 | 53 (13.45) | 1 (1.89) | 3 (5.66) | 9 (16.98) | 19 (35.85) | 21 (39.62) | 0.9823 |
| Total | 394 (100) | 5 (1.27) | 23 (5.84) | 75 (19.04) | 129 (32.74) | 162 (41.12) | – |

*Bold text indicates statistical significance (p<0.05). –, not applicable.

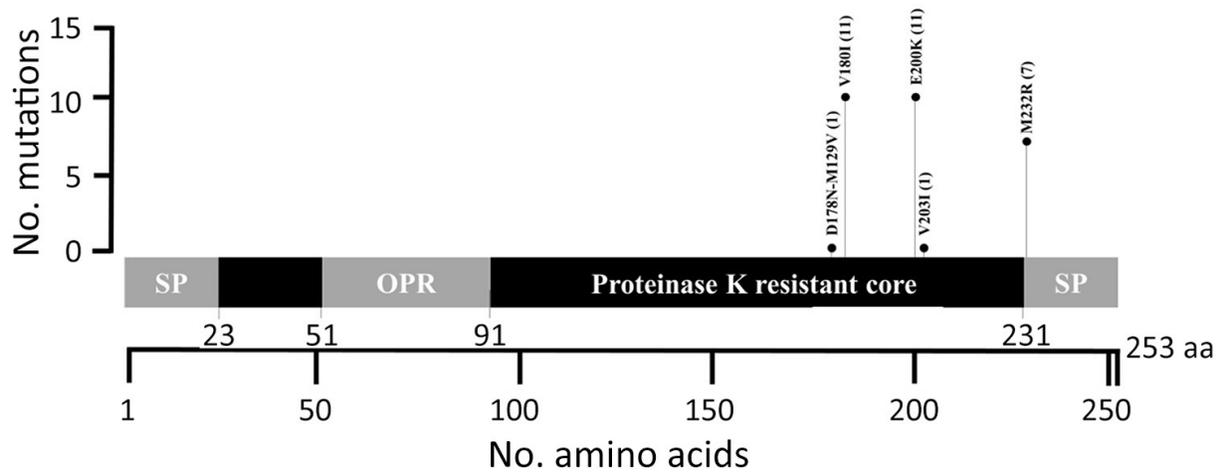
Appendix Table 3. Annual number of Creutzfeldt-Jakob disease cases by city and province, South Korea, 2011–2019

| Year | Total, no. | | | | | | | | | | | | | | | | | |
|--------|------------------|-------------------|------------------|------------------|------------------|------------------|------------------|------------------|----------------|--------------------|------------------|------------------|------------------|------------------|------------------|------------------|------------------|----------------|
| | (%) | Seoul | Busan | Daegu | Incheon | Gwanju | Daejeon | Ulsan | Sejong | Gyeonggi | Gangwon | Chungbuk | Chungnam | Jeonbuk | Jeonnam | Gyeongbuk | Gyeongnam | Jeju |
| 2011 | 29 (7.36) | 7 (24.14) | 0 | 4 (13.79) | 3 (10.34) | 1 (3.45) | 1 (3.45) | 0 | 0 | 7 (24.14) | 1 (3.45) | 0 | 2 (6.9) | 1 (3.45) | 1 (3.45) | 0 | 0 | 1 (3.45) |
| 2012 | 45 (11.42) | 9 (20) | 2 (4.44) | 3 (6.67) | 1 (2.22) | 2 (4.44) | 2 (4.44) | 1 (2.22) | 0 | 10 (22.22) | 3 (6.67) | 1 (2.22) | 4 (8.89) | 3 (6.67) | 2 (4.44) | 0 | 2 (4.44) | 0 |
| 2013 | 34 (8.63) | 7 (20.59) | 4 (11.76) | 5 (14.71) | 1 (2.94) | 1 (2.94) | 1 (2.94) | 1 (2.94) | 0 | 1 (2.94) | 1 (2.94) | 1 (2.94) | 4 (11.76) | 1 (2.94) | 1 (2.94) | 3 (8.82) | 2 (5.88) | 0 |
| 2014 | 65 (16.5) | 12 (18.46) | 2 (3.08) | 3 (4.62) | 3 (4.62) | 0 | 2 (3.08) | 1 (1.54) | 0 | 15 (23.08) | 2 (3.08) | 3 (4.62) | 6 (9.23) | 1 (1.54) | 1 (1.54) | 9 (13.85) | 5 (7.69) | 0 |
| 2015 | 33 (8.38) | 5 (15.15) | 4 (12.12) | 2 (6.06) | 1 (3.03) | 0 | 1 (3.03) | 0 | 0 | 8 (24.24) | 3 (9.09) | 0 | 1 (3.03) | 2 (6.06) | 2 (6.06) | 1 (3.03) | 2 (6.06) | 1 (3.03) |
| 2016 | 43 (10.91) | 13 (30.23) | 3 (6.98) | 3 (6.98) | 3 (6.98) | 1 (2.33) | 1 (2.33) | 1 (2.33) | 1 (2.33) | 8 (18.6) | 0 | 1 (2.33) | 3 (6.98) | 2 (4.65) | 0 | 1 (2.33) | 2 (4.65) | 0 |
| 2017 | 38 (9.64) | 10 (26.32) | 3 (7.89) | 2 (5.26) | 1 (2.63) | 2 (5.26) | 1 (2.63) | 0 | 1 (2.63) | 8 (21.05) | 1 (2.63) | 2 (5.26) | 1 (2.63) | 0 | 1 (2.63) | 2 (5.26) | 3 (7.89) | 0 |
| 2018 | 54 (13.71) | 12 (22.22) | 5 (9.26) | 2 (3.7) | 1 (1.85) | 0 | 1 (1.85) | 3 (5.56) | 0 | 14 (25.93) | 2 (3.7) | 0 | 2 (3.7) | 3 (5.56) | 0 | 4 (7.41) | 5 (9.26) | 0 |
| 2019 | 53 (13.45) | 8 (15.09) | 2 (3.77) | 1 (1.89) | 2 (3.77) | 1 (1.89) | 3 (5.66) | 0 | 0 | 17 (32.08) | 2 (3.77) | 2 (3.77) | 0 | 5 (9.43) | 4 (7.55) | 4 (7.55) | 2 (3.77) | 0 |
| Total† | 394 (100) | 83 (21.07) | 25 (6.35) | 25 (6.35) | 16 (4.06) | 8 (2.03) | 13 (3.3) | 7 (1.78) | 2 (0.51) | 88 (22.34) | 15 (3.81) | 10 (2.54) | 23 (5.84) | 18 (4.57) | 12 (3.05) | 24 (6.09) | 23 (5.84) | 2 (0.51) |
| Pop‡ | 50,913,333 (100) | 9,887,778 (19.42) | 3,438,000 (6.75) | 2,464,889 (4.84) | 2,870,889 (5.64) | 1,500,556 (2.95) | 1,533,222 (3.01) | 1,146,111 (2.25) | 186,000 (0.37) | 12,476,111 (24.50) | 1,512,778 (2.97) | 1,587,222 (3.12) | 2,116,778 (4.16) | 1,822,000 (3.58) | 1,787,778 (3.51) | 2,667,556 (5.24) | 3,312,000 (6.51) | 603,667 (1.19) |

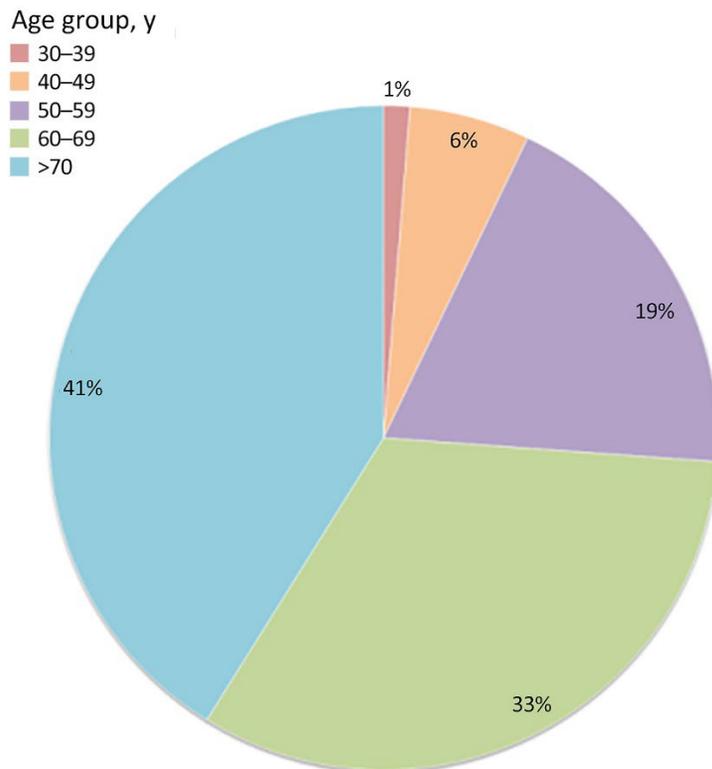
*Pop, population.

†Total number of patients during 2011–2019.

‡The average population per year and city, province, and country.



Appendix Figure 1. Schematic map of the human prion protein (PrP) with germline mutations of the prion protein gene (*PRNP*) found in familial Creutzfeldt-Jakob disease patients, South Korea, 2001–2019. Parentheses indicate the number of patients carrying each germline mutation. OPR, octapeptide repeat region; SP, signal peptide.



Appendix Figure 2. Distribution of Creutzfeldt-Jakob disease cases by age group, South Korea, 2001–2019.