Supplementary Table e-1. Incidence of the *C9orf72* HRE ALS, ALS/FTD, and FTD in published European Cohorts

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| **Ethnogeographic Origin** | **n** | **ALS** | **ALS/FTD** | **FTD** | **Lab Method** | **Study** |
| London clinic | 296 | fALS 15/64 (23%) | fALS/FTD  14/16 (87.5%) | - | Repeat-primed PCR | Smit et al, 2013 |
| sALS 13/216 (6%) |
| Other UK | 870 | fALS 35/93 (37%) | fALS/FTD  10/13 (77%) | - | Repeat-primed PCR | Smit et al, 2013 |
| sALS 58/737 (7.9%) | sALS/FTD  6/27 (22%) |
| Sweden | 77 | fALS 10/16 (63%) | fALS/FTD 20/25 (80%) | 2/3 (67%) | Repeat-primed PCR | Smit et al, 2013 |
| sALS 3/21 (14%) | sALS/FTD  1/12 (8.3%) |
| Netherlands | 27 | fALS 10/27 (37%) | - | - | Repeat-primed PCR | Smit et al, 2013 |
| Belgium | 22 | fALS 16/19 (84%) | fALS/FTD 3/3 (100%) | - | Repeat-primed PCR | Smit et al, 2013 |
| Italy | 55 | fALS 3/10 (30%) | fALS/FTD 1/10 (10%) | - | Repeat-primed PCR | Smit et al, 2013 |
| sALS 1/5 (20%) | sALS/FTD 5/30 (17%) |
| Finnish | 476 | fALS 52/112 (46.4%) | - | fFTD 13/27 (48.1%) | Repeat-primed PCR | Majounie et al, 2012c |
| sALS 61/289 (21.1%) | sFTD 9/48 (18.8%) |
| Swedish | 7 | - | - | fFTD 1/1 (100%) | Repeat-primed PCR | Majounie et al, 2012c |
| sFTD 0/6 (0%) |
| English | 1,727 | fALS 45/98 (45.9%) | - | fFTD 28/170 (16.5%) | Repeat-primed PCR | Majounie et al, 2012c |
| sALS 62/916 (6.8%) | sFTD 31/543 (5.7%) |
| Irish | 1 | fALS 1/1 (100%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| German | 519 | fALS 15/69 (21.7%) | - | fFTD 4/29 (13.8%) | Repeat-primed PCR | Majounie et al, 2012c |
| sALS 22/421 (5.2%) |
| Dutch | 340 | - | - | fFTD 30/116 (25.9%) | Repeat-primed PCR | Majounie et al, 2012c |
| sFTD 5/224 (2.2%) |
| French | 200 | - | - | fFTD 22/50 (44%) | Repeat-primed PCR | Majounie et al, 2012c |
| sFTD 14/150 (9.3%) |
| Italian | 555 | fALS 34/90 (37.8%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| sALS 19/465 (4.1%) |
| Sardinian | 165 | fALS 11/19 (57.9%) | - | fFTD 1/7 (14.3%) | Repeat-primed PCR | Majounie et al, 2012c |
| sALS 10/129 (7.8%) | sFTD 0/10 (0%) |
| Moldovan | 3 | sALS 0/3 (0%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| European | 659 | - | fALS/FTD 11/23 (47.8%) | fFTD 21/251 (8.4%) | Repeat-primed PCR and short tandem repeat fragment length assay | Van der Zee et al, 2013 |
| sALS/FTD 5/27 (18.5%) | sFTD 13/358 (3.6%) |
| Belgian | 360 | - | fALS/FTD 6/7 (85.7%) | fFTD 12/101 (11.9%) | Repeat-primed PCR and short tandem repeat fragment length assay | Van der Zee et al, 2013 |
| sALS/FTD 1/16 (6.25%) | 9/236 (3.8%) |

Supplementary Table e-2. Incidence of the *C9orf72* HRE ALS, ALS/FTD, and FTD in published Non-European Cohorts

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| **Ethnogeographic Origin** | **n** | **ALS** | **ALS/FTD** | **FTD** | **Lab Method** | **Study** |
| USA White | 1,053 | fALS 59/163 (36.2%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| sALS 48/890 (5.4%) |
| USA Hispanic | 72 | sALS 6/72 (8.3%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| USA Black | 49 | sALS 2/49 (4.1%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| Native American | 3 | sALS 0/3 (0%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| Middle Eastern | 3 | fALS 0/2 (0%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| sALS 0/1 (0%) |
| Israeli | 14 | fALS 3/14 (21.4%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| Indian | 62 | sALS 0/31 (0%) | - | sFTD 0/31 (0%) | Repeat-primed PCR | Majounie et al, 2012c |
| Asian | 271 | fALS 1/20 (5%) | - | fFTD 2/3 (66.7%) | Repeat-primed PCR | Majounie et al, 2012c |
| sALS 0/238 (0%) | sFTD 0/10 (0%) |
| Pacific Islander/Guam | 90 | sALS 0/90 (0%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| Australian | 263 | sALS 14/263 (5.3%) | - | - | Repeat-primed PCR | Majounie et al, 2012c |
| Taiwanese | 124 | fALS 4/22 (18.2%) | - | - | PCR | Tsai et al, 2012 |
| sALS 2/102 (2%) |
| South African | 140 | sALS 10/140 (7%) | - | - | Repeat-primed PCR | Nel et al, 2019 |
| Korean | 254 | fALS 0/8 (0%) | - | - | 2-step PCR | Jang et al, 2013 |
| sALS 0/246 (0%) |
| Japanese | 168 | fALS 2/58 (3.4%) | - | - | Repeat-primed PCR and genotyping-PCR | Konno et al, 2013 |
| sALS 0/110 (0%) |
| Sardinian | 375 | fALS 33/100 (33%) | - | - | PCR | Borghero et al, 2014 |
| sALS 18/275 (6.5%) |
| Brazilian | 402 | fALS 8/68 (11.8%) | - | fFTD 1/14 (7.1%) | PCR | Cintra et al, 2018 |
| sALS 11/309 (3.6%) | sFTD 0/11 (0%) |
| Russian | 208 | fALS 0/9 (0%) | - | - | Fragment length analysis and repeat-primed PCR | Lysogorskaia et al, 2015 |
| sALS 5/199 (2.5%) |
| Spain | 936 | fALS 42/155 (27.1%) | - | - |  | Masso et al, 2014 |
| sALS 25/781 (3.2%) |
| Chinese | 344 | fALS 0/20 (0%) | - | - | Repeat-primed PCR | Zou et al, 2013 |
| sALS 0/324 (0%) |