

Supplementary Online Content

Logroscino G, Piccininni M, Graff C, et al; FRONTIERS group. Incidence of syndromes associated with frontotemporal lobar degeneration in 9 European countries. *JAMA Neurol*. Published online January 30, 2023. doi:10.1001/jamaneurol.2022.5128

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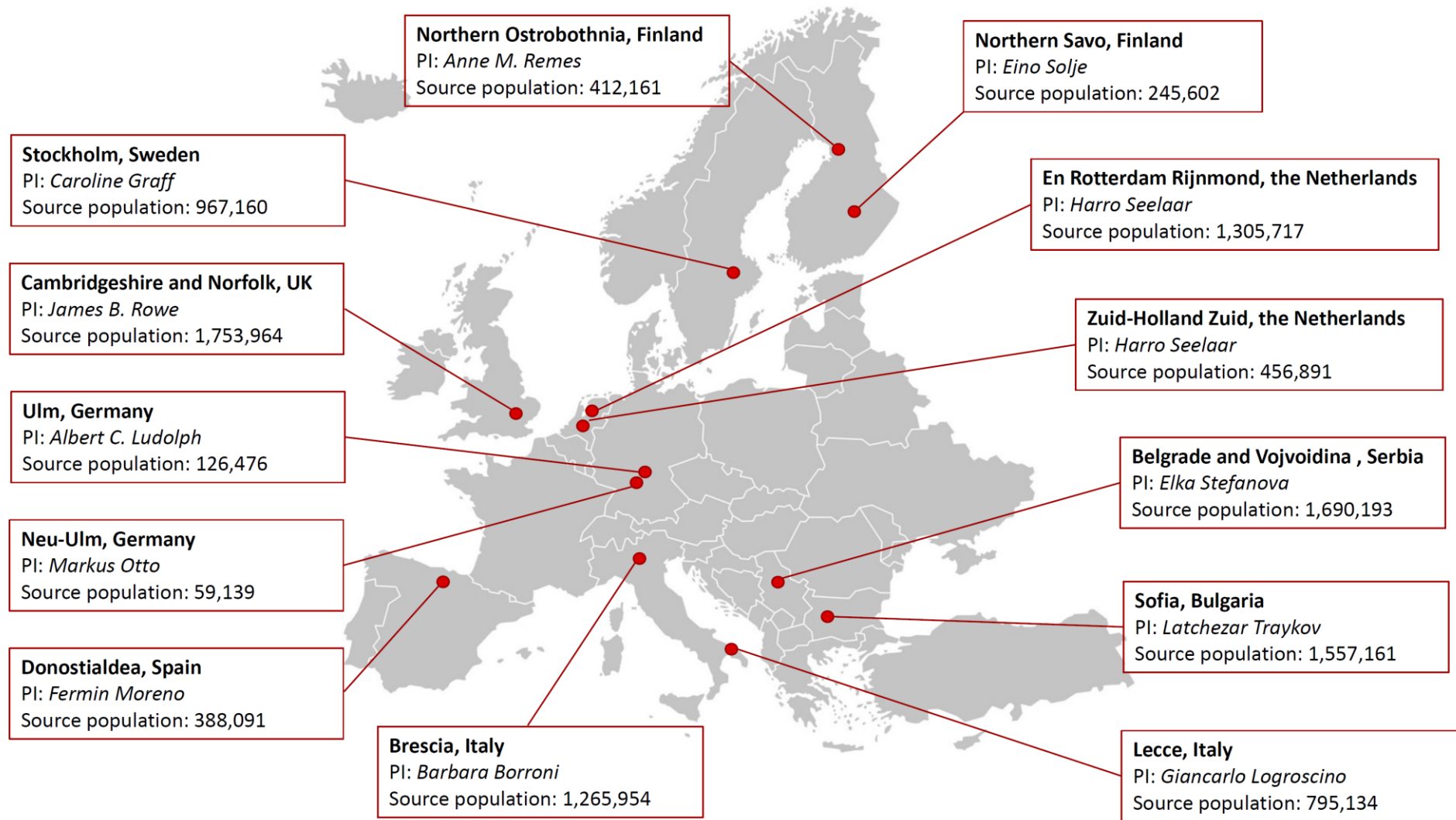
This supplementary material has been provided by the authors to give readers additional information about their work.

eMethods. Details About the Modified Goldman Score and the Random Intercept Poisson Model

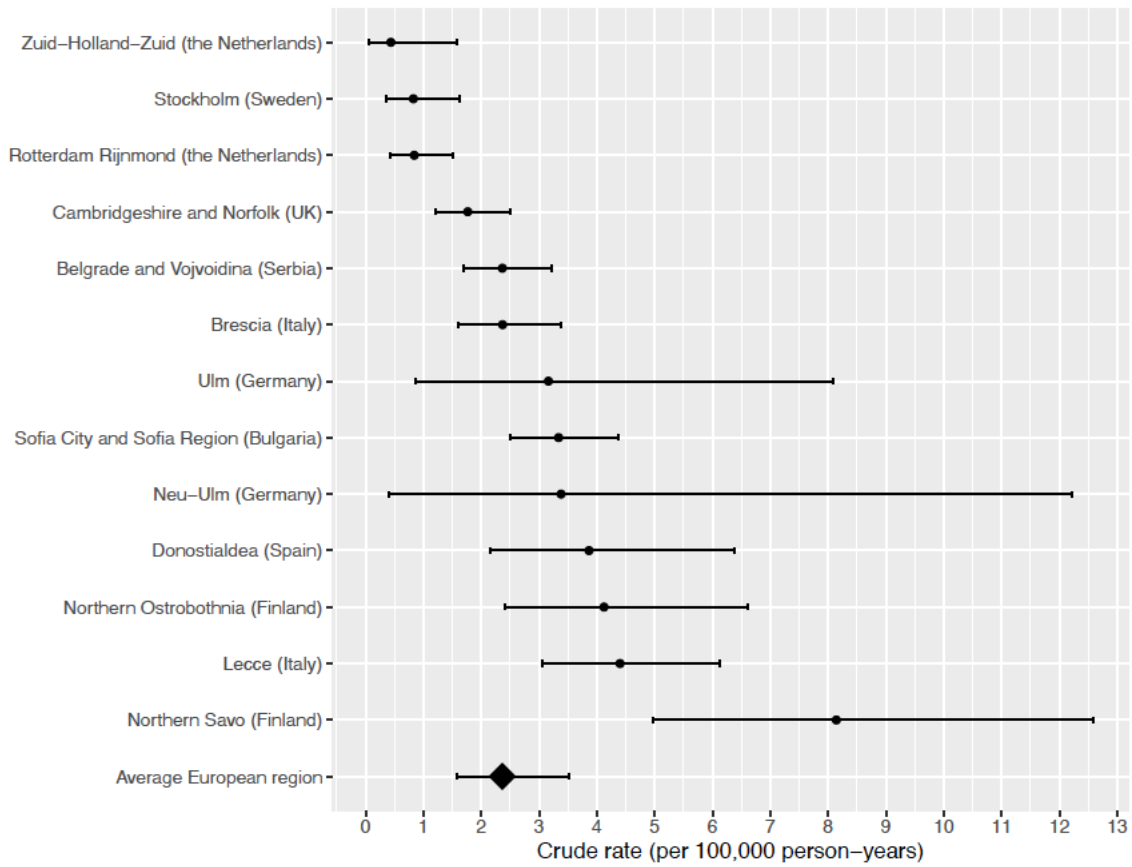
Modified Goldman's score: A modified Goldman's score (GS) of 1 corresponds to a family history consistent with the proband's clinical syndrome with an autosomal dominant inheritance pattern, with at least three people who are affected in two generations and who are linked by a first-degree relative; a GS of 2 indicates familial aggregation of three or more affected relatives but without meeting the criteria for a score of 1; a GS of 3 denotes one other affected relative with a history of young-onset dementia within the family, i.e. < 65; a GS of 3.5 denotes one other affected relative with the onset above 65; a GS of 4 signifies no known family history of neurodegenerative disorders.

Random intercept Poisson model by age and sex: In order to model the FTLD incidence rate based on age and sex, we fitted a random-intercept Poisson model with the number of cases as the dependent variable and the logarithm of the number of residents as the offset. Sex and the midpoint of the age group interval (105 was used for the last age category) were introduced into the model as independent variables. Age was further centred at 50 and divided by 10 to ensure the convergence of the model, and, in order to account for the non-linearity of the effect, it was modelled as a cubic polynomial. A random intercept was included for each residence area to account for heterogeneity. Goodness of fit was evaluated graphically by comparing the observed rates with the predicted rates by age group, sex and residence area. The model was fitted with the Laplace approximation to the log-likelihood using the `glmer` function in the `lme4` R package.

eFigure 1. FRONTIERS Centres, Principal Investigators, and Source Populations



eFigure 2. FTLD incidence rates in the 13 referral geographical areas.



eTable. FTLD incidence rates in the 13 referral geographical areas by age groups.

Residence Area	Age groups, years				
	0-44	45-64	65-74	75-84	85+
Belgrade and Vojvodina (Serbia)	0.00 (0.00-0.33)	5.59 (3.62-8.26)	6.74 (3.59-11.53)	1.97 (0.24-7.11)	0.00 (0.00-10.06)
Brescia (Italy)	0.00 (0.00-0.48)	2.93 (1.46-5.24)	9.66 (5.14-16.52)	6.17 (2.26-13.42)	0.00 (0.00-7.38)
Cambridgeshire and Norfolk (UK)	0.00 (0.00-0.32)	1.31 (0.48-2.86)	8.44 (4.92-13.51)	6.73 (2.90-13.26)	0.00 (0.00-5.85)
Donostialdea (Spain)	0.00 (0.00 -1.60)	0.86 (0.02-4.81)	18.72 (8.08-36.89)	18.12 (5.88-42.28)	6.65 (0.17-37.08)
Rotterdam Rijnmond (the Netherlands)	0.00 (0.00-0.41)	0.87 (0.18-2.55)	5.32 (2.14-10.96)	1.40 (0.04-7.79)	0.00 (0.00-10.46)
Lecce (Italy)	0.00 (0.00-0.80)	3.05 (1.23-6.28)	14.81 (8.10-24.85)	20.87 (11.41-35.02)	0.00 (0.00-10.13)
Northern Ostrobothnia (Finland)	0.00 (0.00-1.27)	7.14 (2.87-14.72)	13.20 (4.84-28.73)	16.72 (4.56-42.81)	0.00 (0.00-32.16)
Northern Savo (Finland)	0.84 (0.02-4.66)	10.62 (4.27-21.89)	26.24 (12.00-49.82)	16.41 (3.39-47.97)	0.00 (0.00-39.00)
Neu-Ulm (Germany)	0.00 (0.00-9.17)	0.00 (0.00-19.11)	0.00 (0.00-57.03)	46.36 (5.61-167.47)	0.00 (0.00-243.75)
Sofia City and Sofia Region (Bulgaria)	0.11 (0.00-0.63)	7.14 (4.75-10.32)	9.52 (5.44-15.46)	7.99 (3.21-16.47)	0.00 (0.00-9.95)
Stockholm (Sweden)	0.00 (0.00-0.51)	0.86 (0.10-3.11)	6.03 (1.96-14.08)	2.16 (0.05 -12.06)	0.00 (0.00-14.63)
Ulm (Germany)	0.00 (0.00-4.26)	0.00 (0.00-9.28)	17.90 (2.17- 64.67)	21.25 (2.57-76.75)	0.00 (0.00-89.64)
Zuid-Holland-Zuid (the Netherlands)	0.00 (0.00-1.25)	0.00 (0.00-2.36)	3.87 (0.47-13.99)	0.00 (0.00-10.45)	0.00 (0.00-28.03)
Average European region	0.03 (0.01-0.14)	3.27 (2.20-4.87)	9.06 (6.12-13.42)	7.55 (4.89-11.65)	0.35 (0.05-2.50)

All incidence rates are expressed per 100,000 person-years. Age-group specific incidence rates of Frontotemporal Lobar Degeneration in the Average European region are derived from the fixed effects of a random-intercept Poisson model including age group as covariate, and a random intercept for residence area. 95% Confidence Intervals are reported between brackets.