Temporal dynamics of the Scale for the Assessment and Rating of Ataxia in autosomal dominant cerebellar ataxias.

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SARA scale, context & aims of the study

SARA (Scale for the Assessment and Rating of Ataxia):

-Clinical score (from 0 to 40)

-Assess the presence and severity of Ataxia

Item 1-Gait (max 8)
Item 2-Stance (max 6)
Item 3-Sitting (max 4)
Item 4-Speech (max 6)
Item 5-Finger Chase (max 4)
Item 6-Finger Nose (max 4)
Item 7-Hand Fast (max 4)
Item 8-Heel Shin (max 4)

Context:

-Upcoming treatments (reliable outcome needed)

Aims:

-Assess the temporal dynamics of SARA:

- At the item level
- Globally

Methods

Participants:

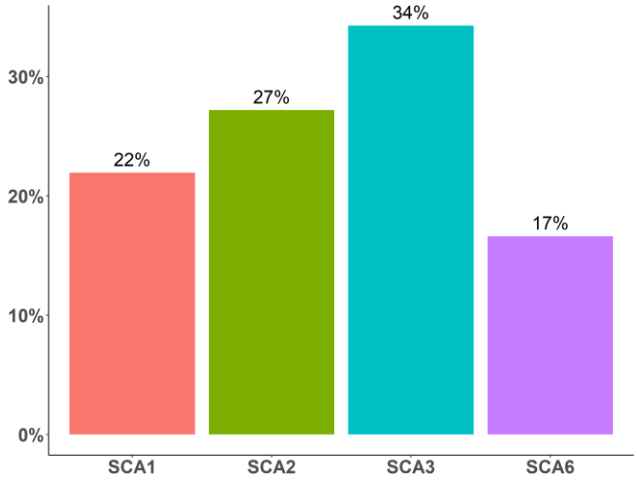
-Grouping of 4 cohorts (EUROSCA, RISCA, CRC-SCA, SPATAX)
-1210 participants (SCA1, SCA2, SCA3, SCA6)
-4092 visits
-3697 patients-years

Statistical tool:

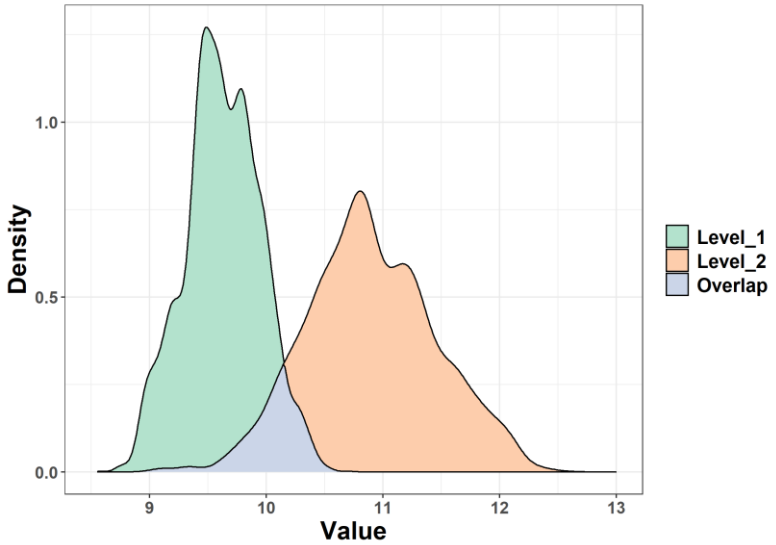
-Bayesian mixed effect model, (Leaspy)

Average trajectory:
Time spent at each level

Disease distribution at baseline

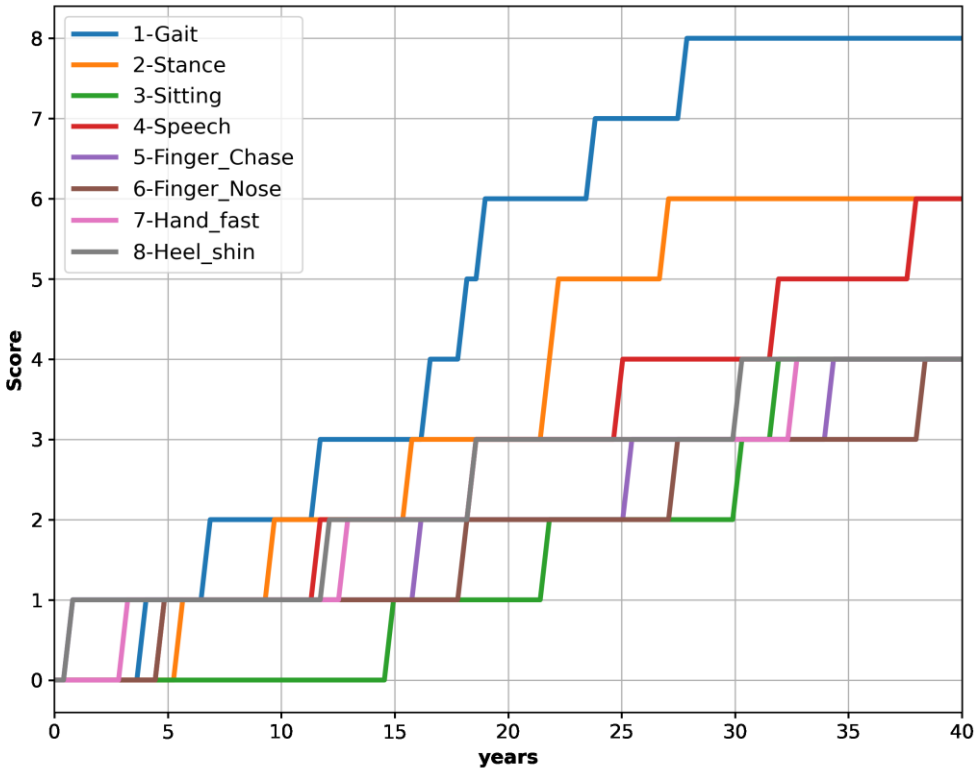


Parameters and comparisons



Results, Item level analysis

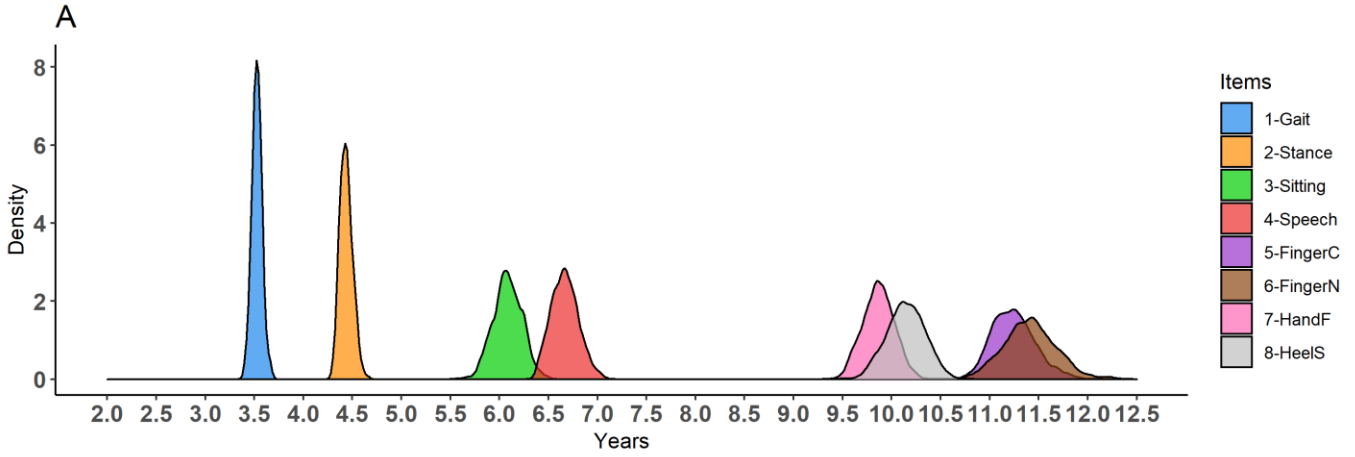
Average progression of each item

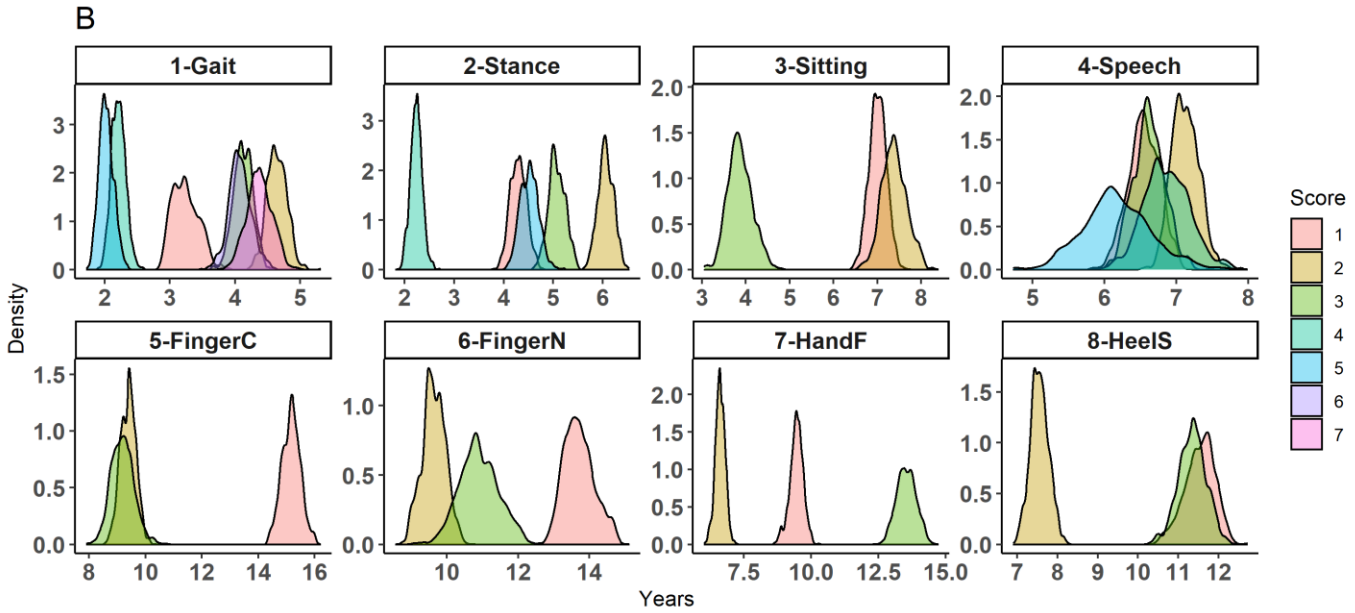


-Different speed of progression.

-Non linear progression

A: Average time for a one-point increase

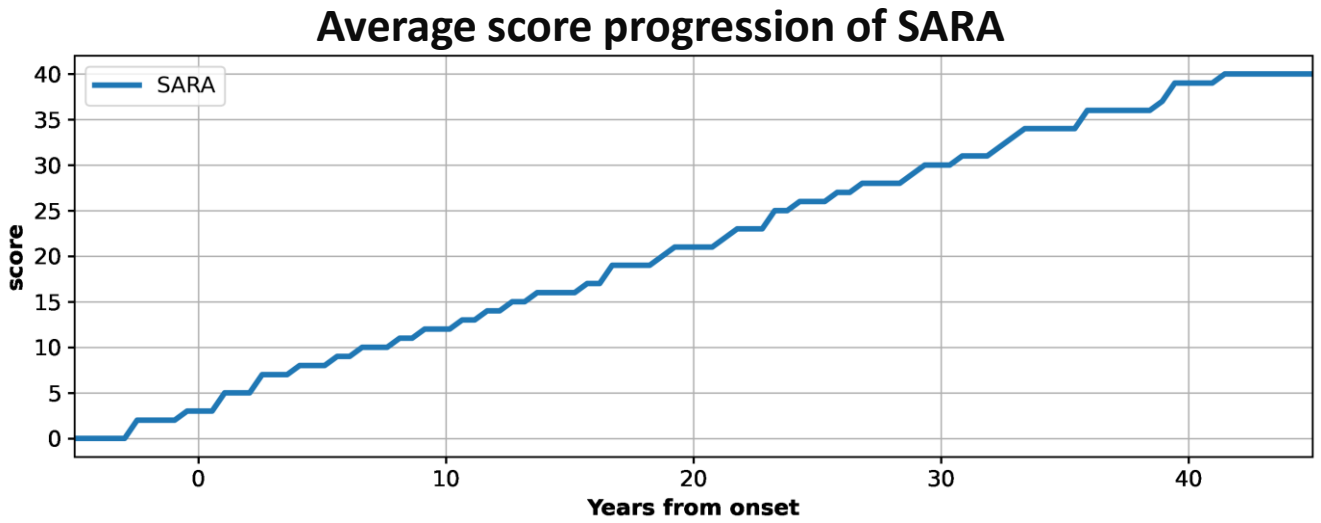




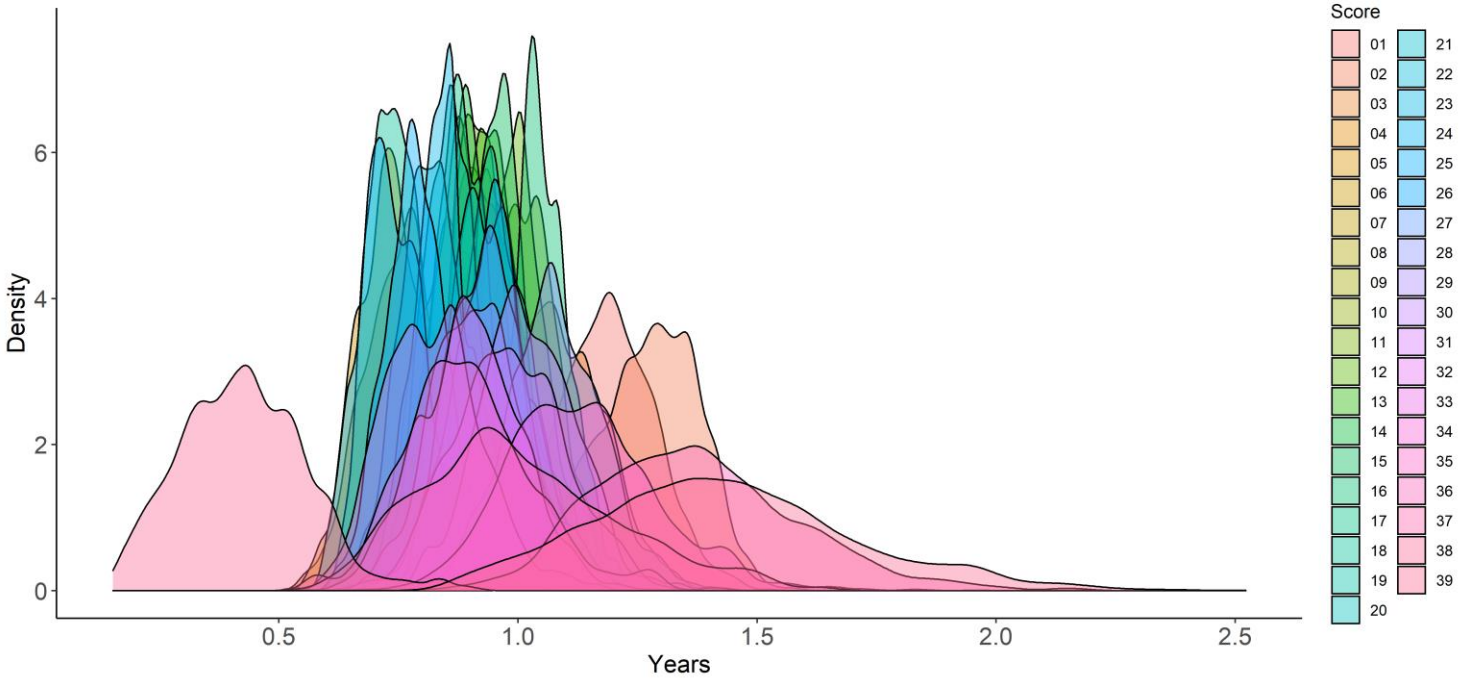
B: Time spent at each level of each item

Results: Global progression of the scale

-Linear progression of SARA scale



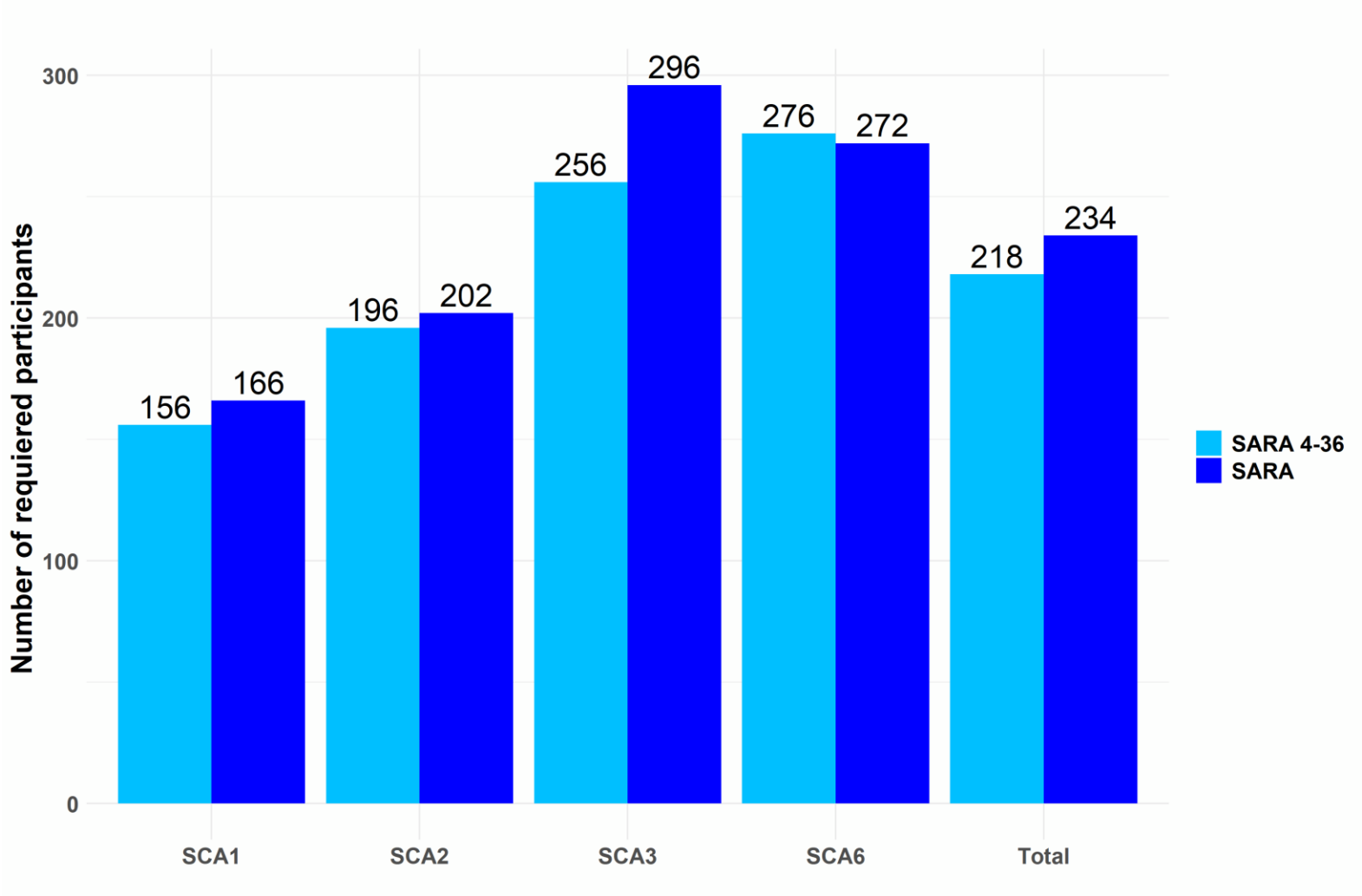
-Best overlap between score 4 and 36



Results: Required sample size for therapeuthic trials

Inclusion criterion SARA between 4 and 36

=> Less participants required to highline the same treatment effect in a trial



α=5%, β=90%, treatment effect=50%, duration=12 mois

Conclusion

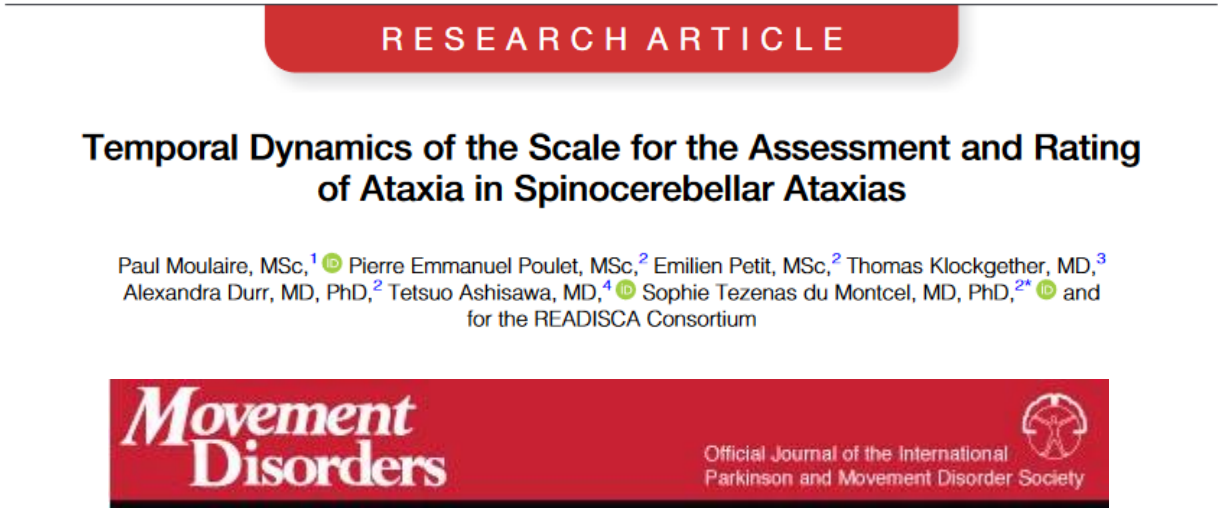
New findings:

-Heterogeneous progression at the item level

-Stable and linear temporal dynamic of the SARA scale

-Better properties between SARA score 4 and 36

More informations available:



RESEARCH ARTICLE

Temporal Dynamics of the Scale for the Assessment and Rating of Ataxia in Spinocerebellar Ataxias

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Temporal dynamic as a function of cofactors

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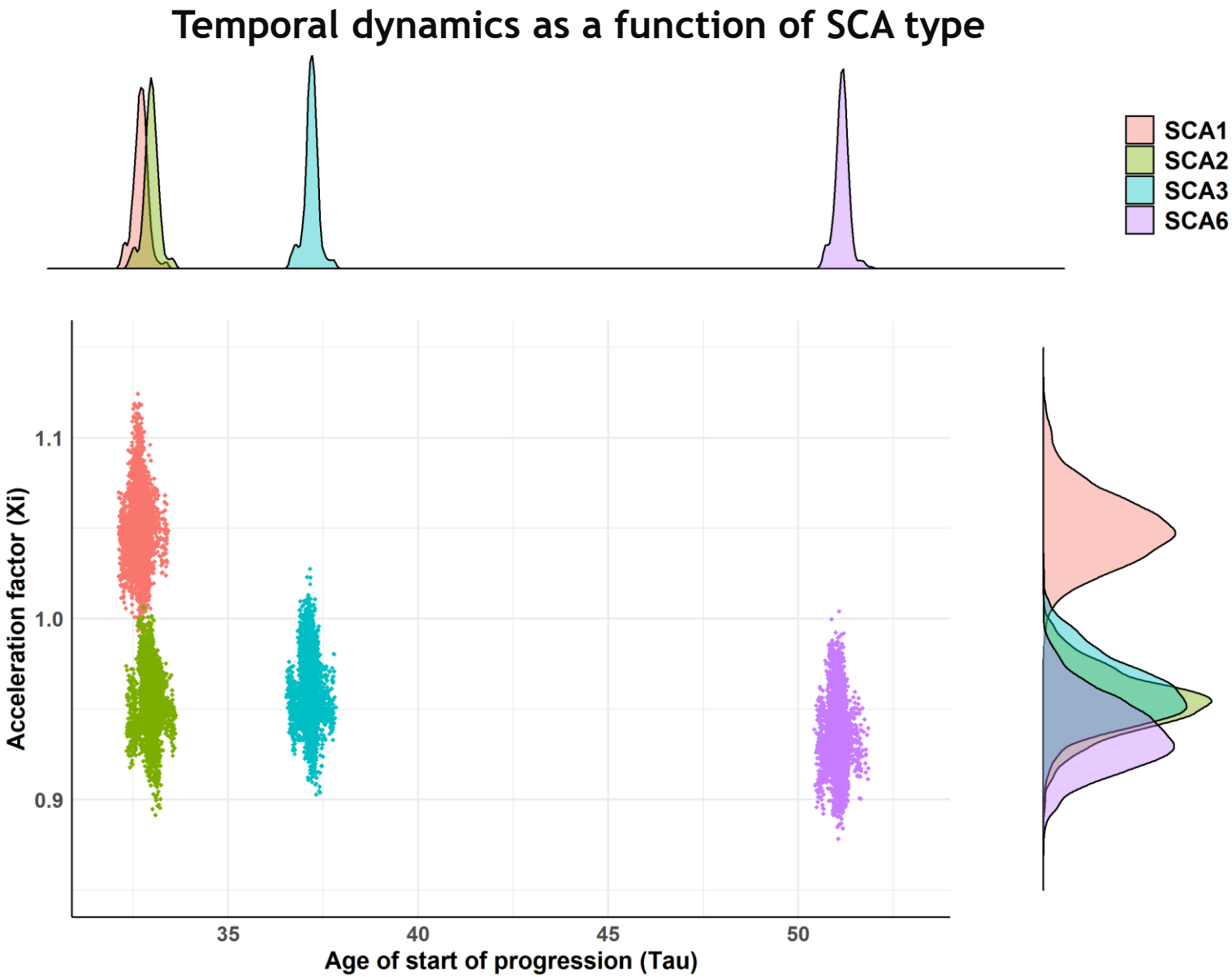
Individual parameters:
- Acceleration factor (Xi)
- Age of start of progression (Tau)

-Confirmatory results:
Different dynamics between SCAs



Temporal dynamic as a function of continent (A) and cohort (B)

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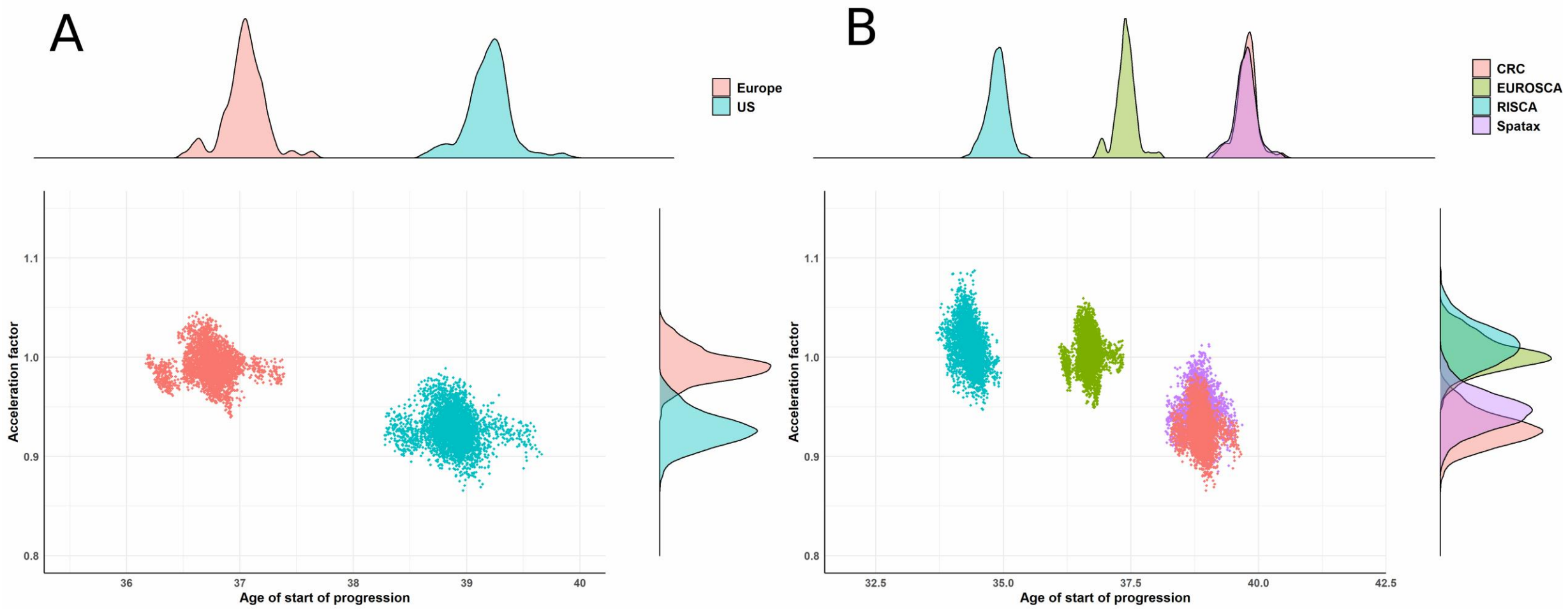

Temporal dynamic as a gender

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