Long-term fate of an unselected cohort of congenital long QT syndrome patients diagnosed in childhood

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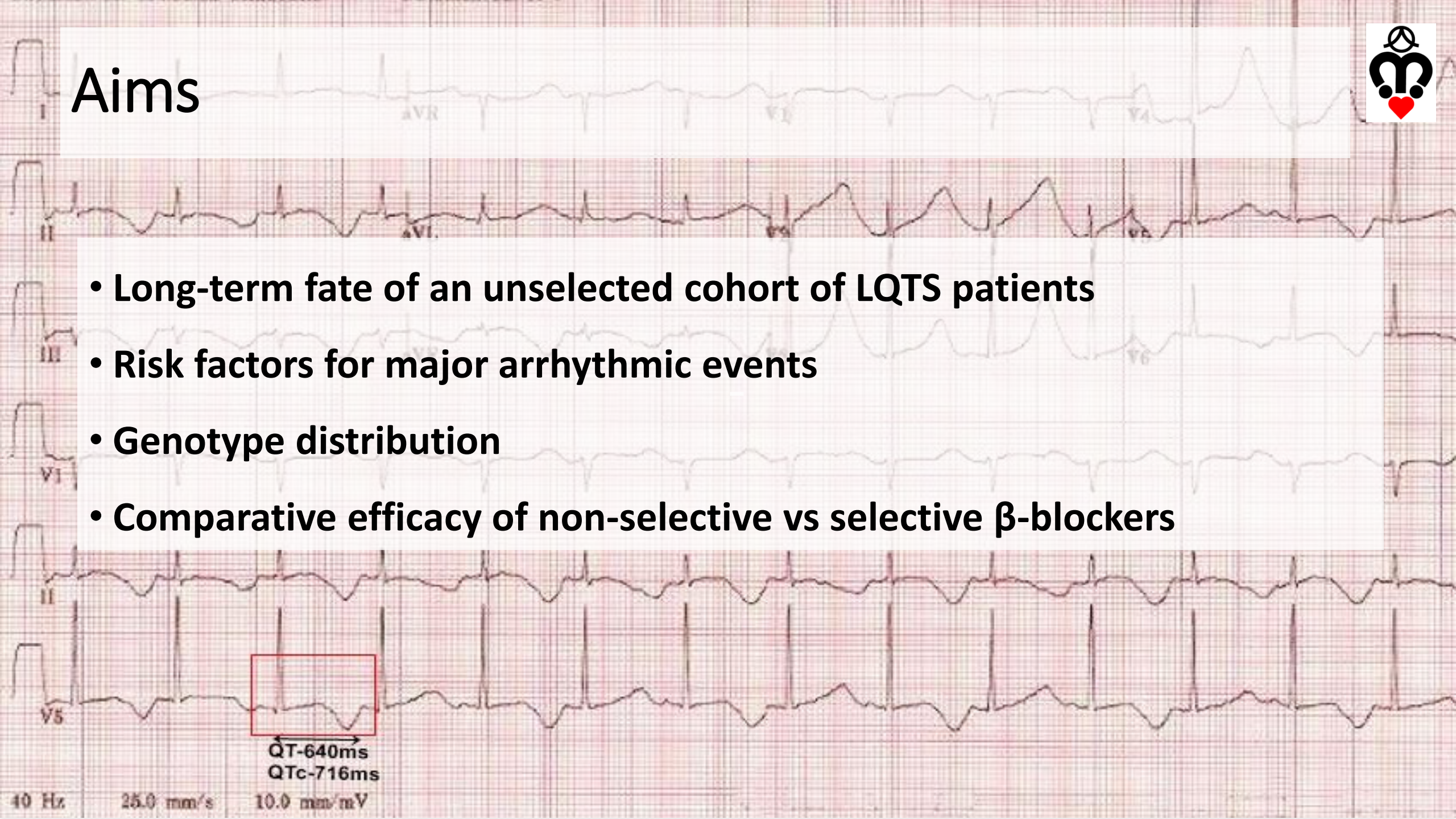
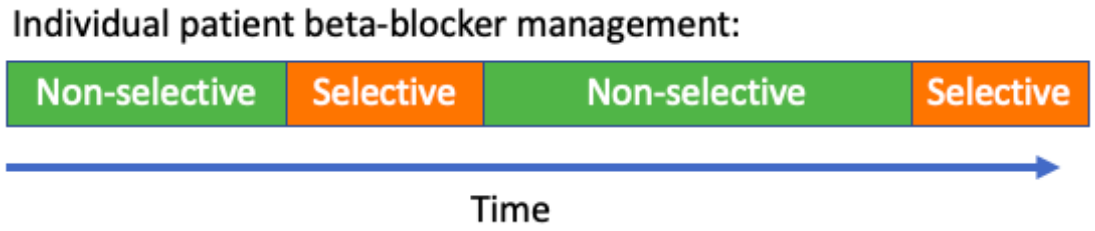
Aims

- Long-term fate of an unselected cohort of LQTS patients
- Risk factors for major arrhythmic events
- Genotype distribution
- Comparative efficacy of non-selective vs selective β-blockers



Methods

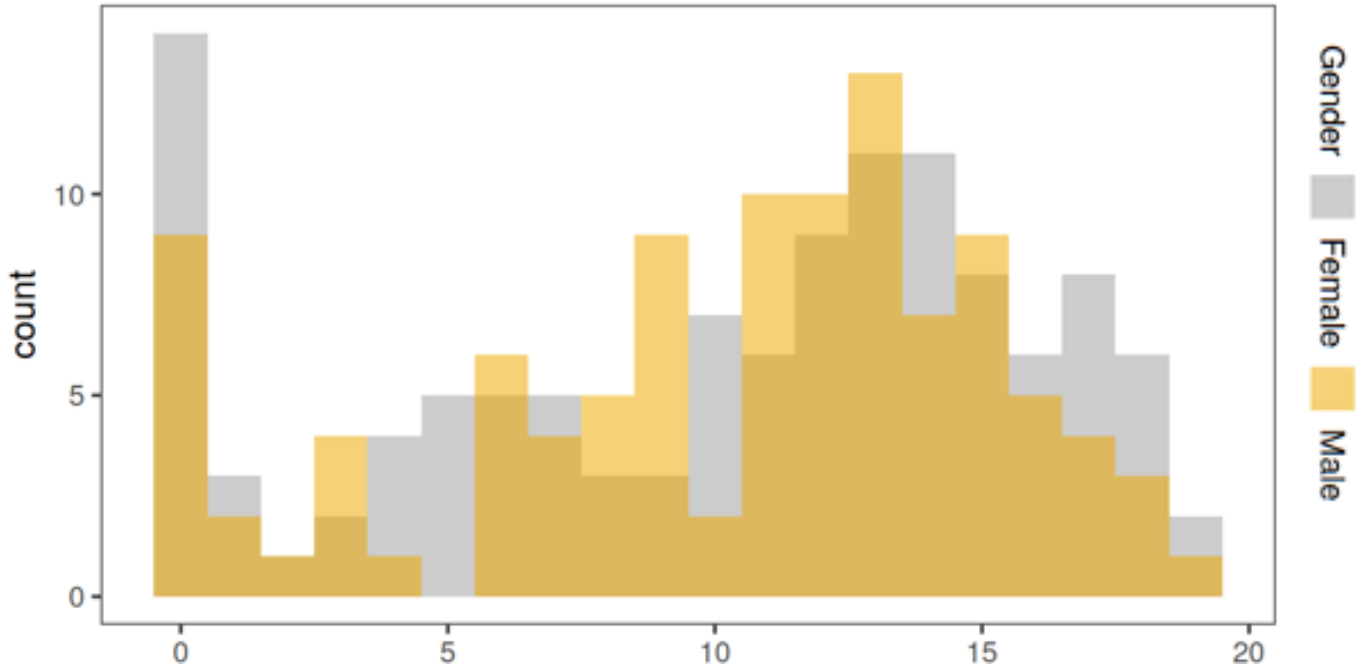
- Retrospective study
  - All patients from the Bohemian region diagnosed with LQTS
- Inclusion criteria
  - Schwartz score >1.5 points and/or likely pathogenic/pathogenic mutation
- Follow up data
  - Hospital/outpatient records
  - Structured patient phone calls
  - National death registry
- Endpoints
  - Death from any cause
  - Major arrhythmic event (MAE)
    - SCD, SCA, VF, VT, appropriate ICD shock
- Statistical analysis:
  - Kaplan-Meier survival analysis (log-rank statistics)
  - Cox proportional hazard model, Poisson regression model
    - prediction of MAE burden



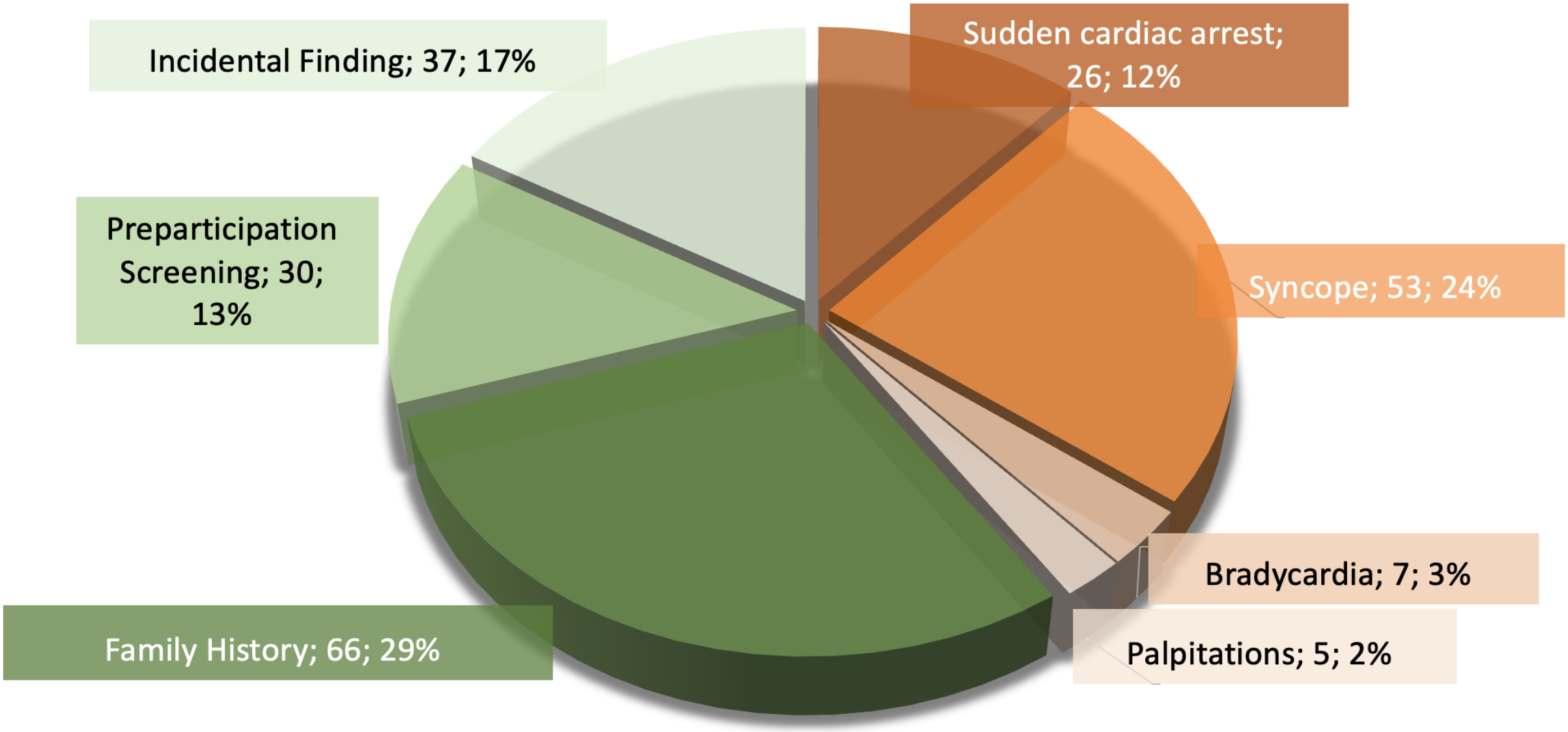
Patients

Number of patients	224
Period	1985 - 2021
Males/females	105/119
Age (median)	11.7 (IQR 6.5 - 14.2) years
Follow-up (median)	8.8 (IQR 2.8 – 16.7) years

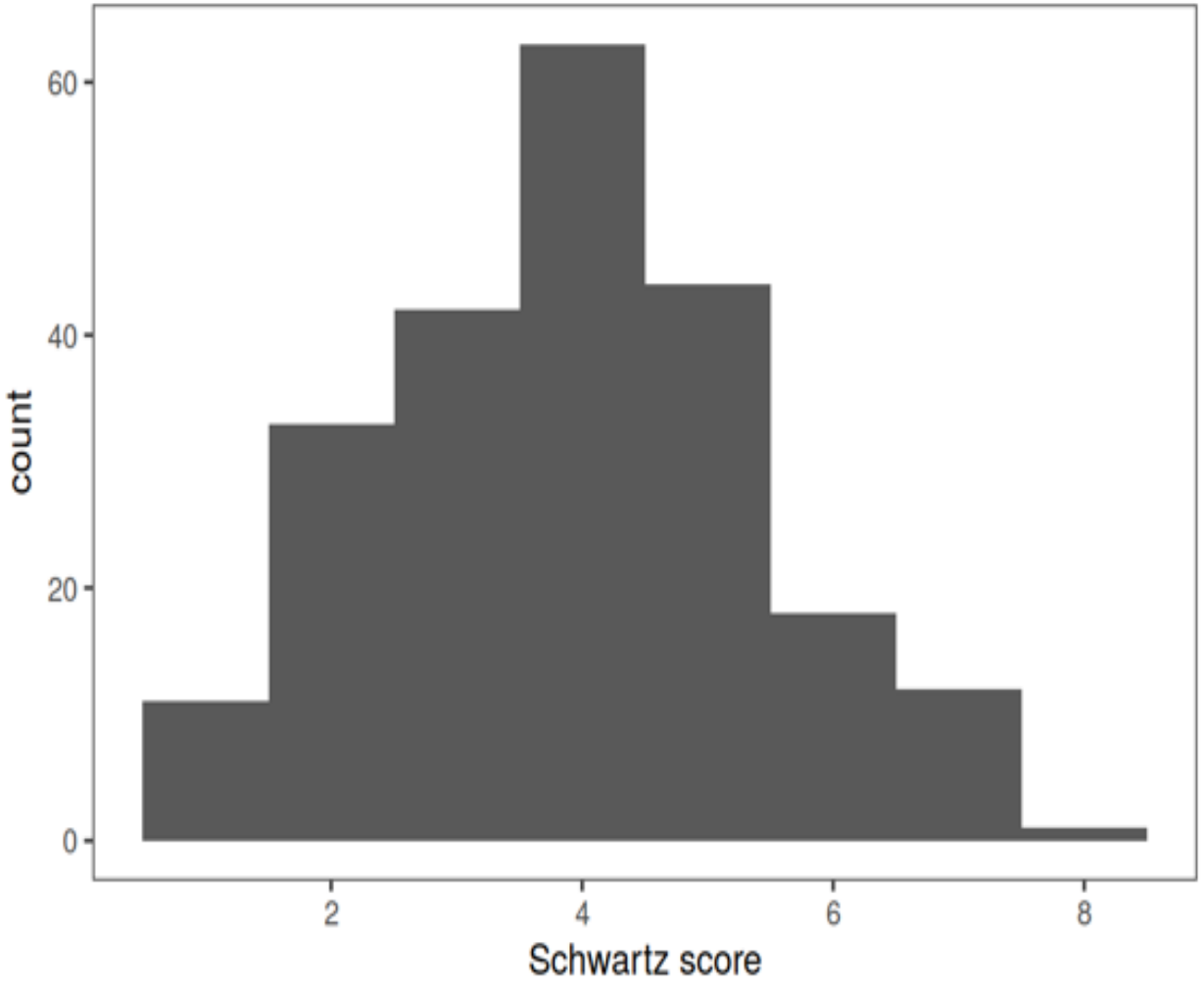
Presentation Age:



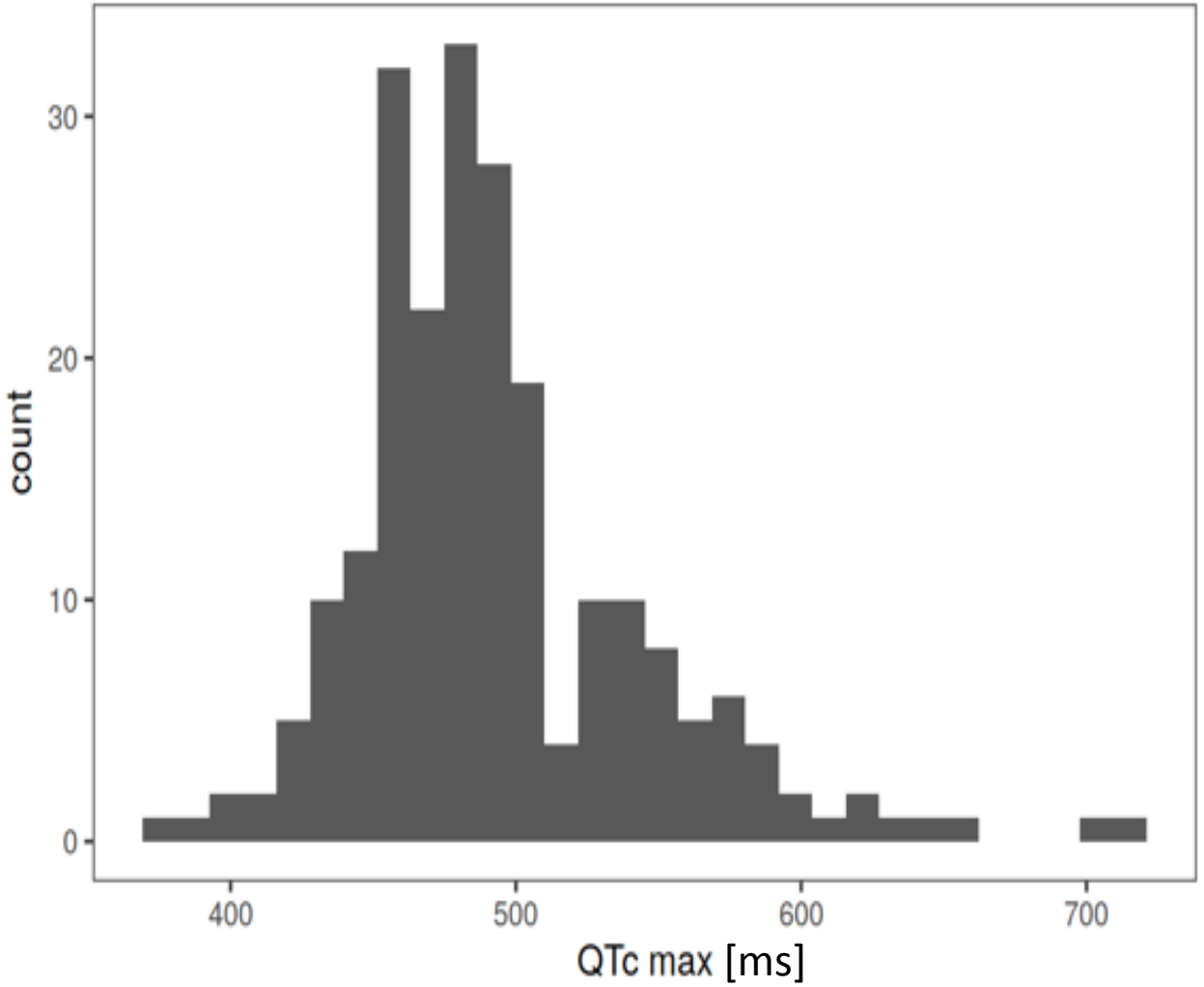
Reasons for presentation



Schwartz Score

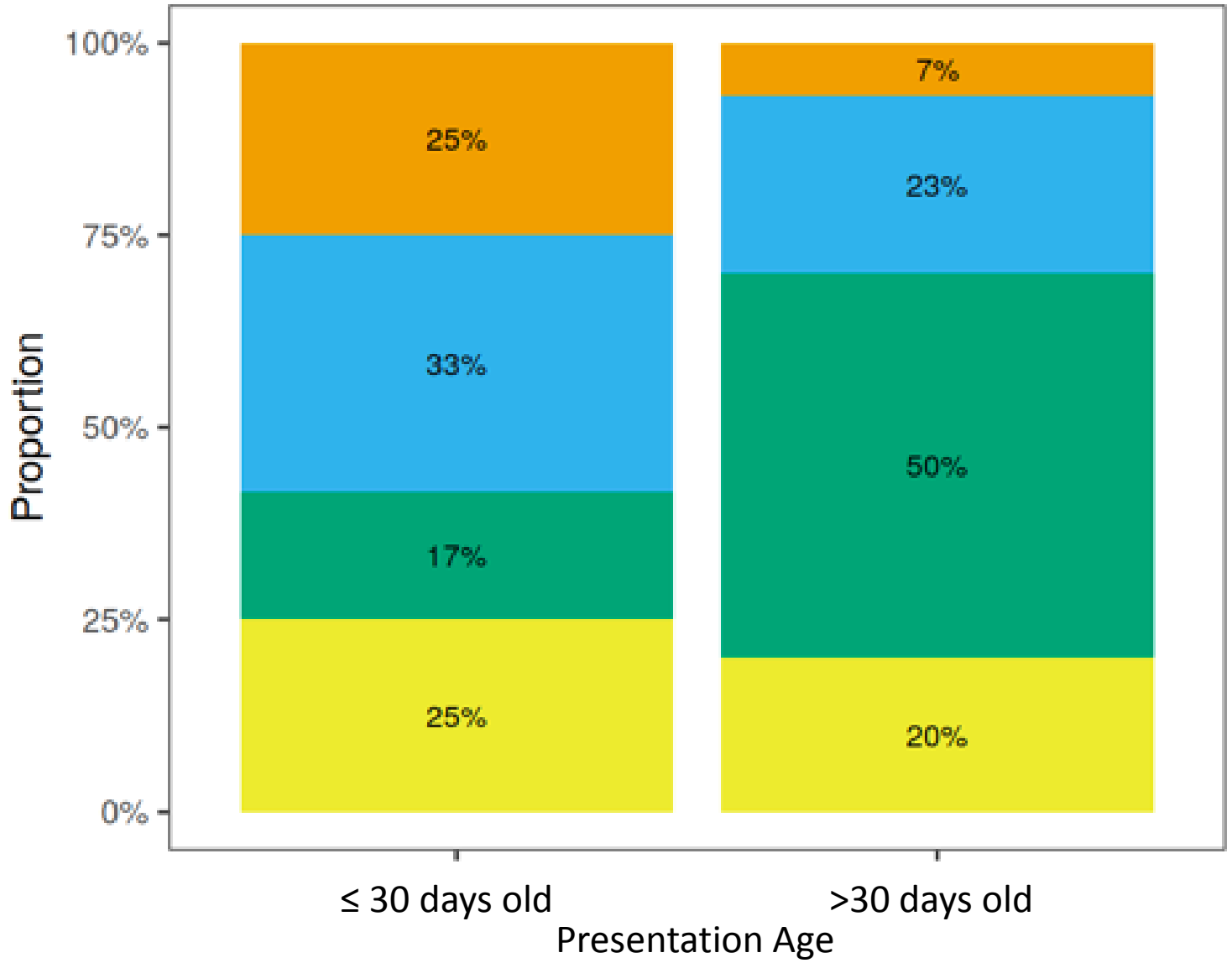


QTc max



QTc max = maximal QTc at any time

Genotypes

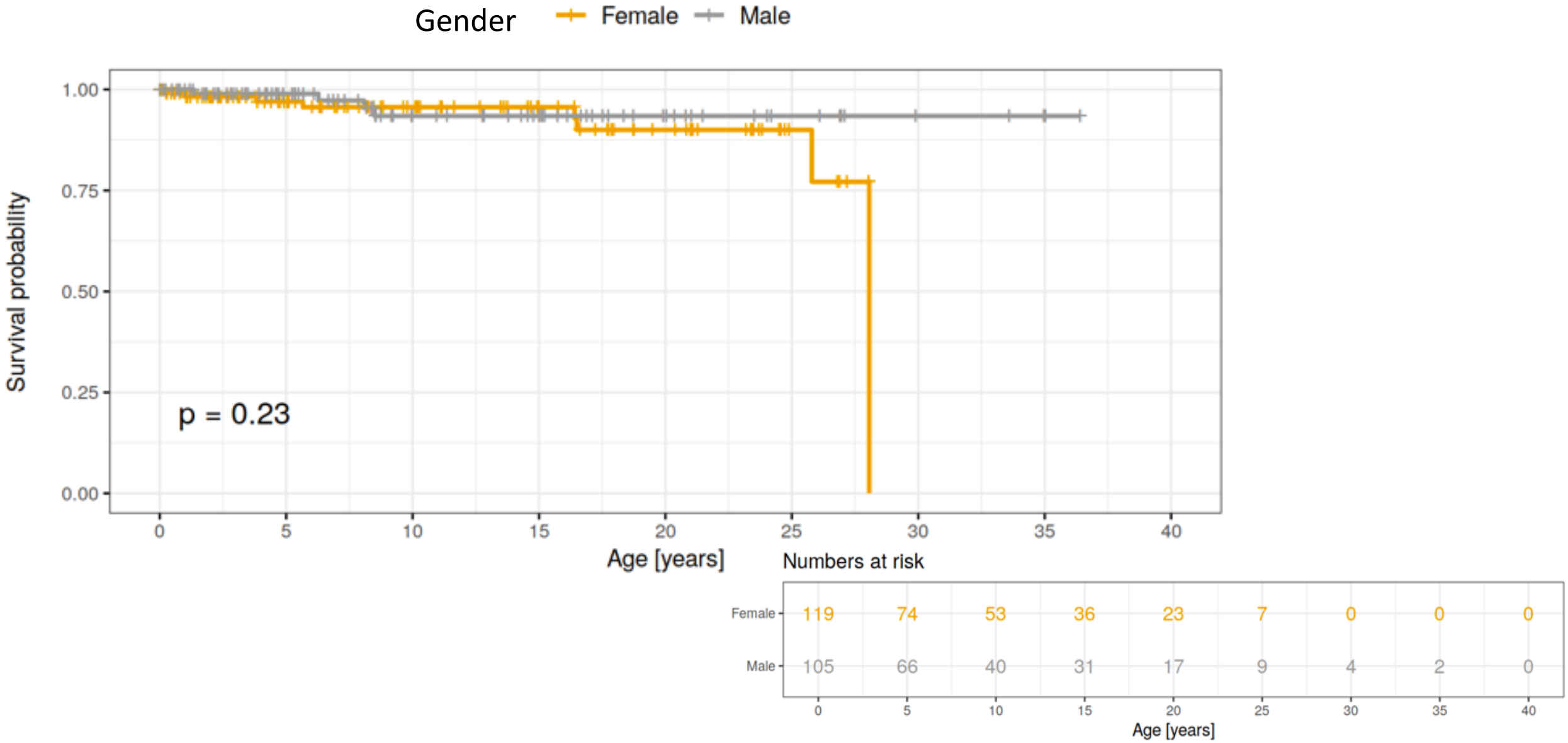


Evaluated in:
159/224 patients (71%)

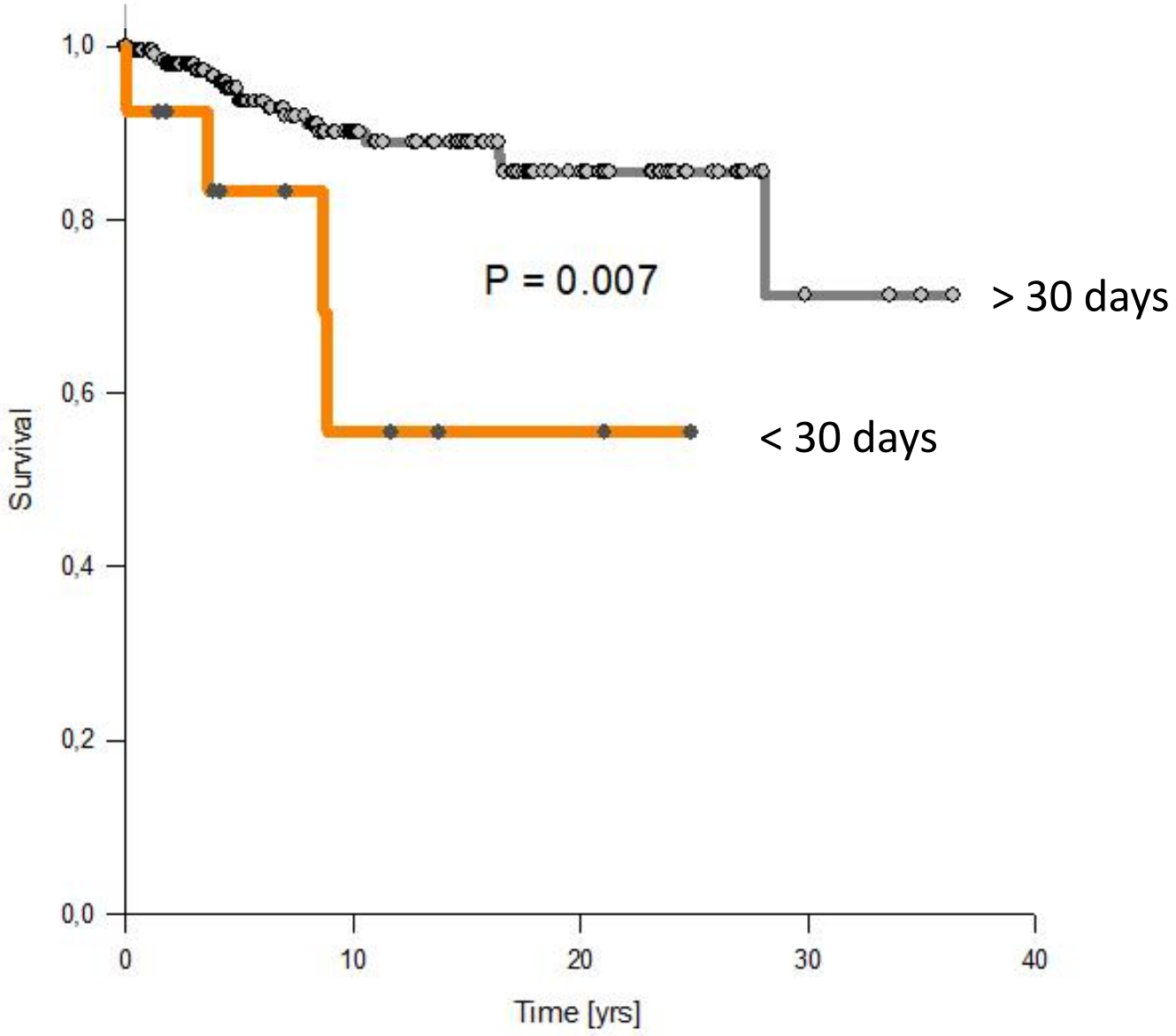
Genotype
- No LP/P mutation found
- LQTS1 (KCNQ1)
- LQTS2(KCNH2)
- LQTS3 (SCN5A)

LQTS 2 and 3 more prevalent in those presenting as newborns (p=0.042)

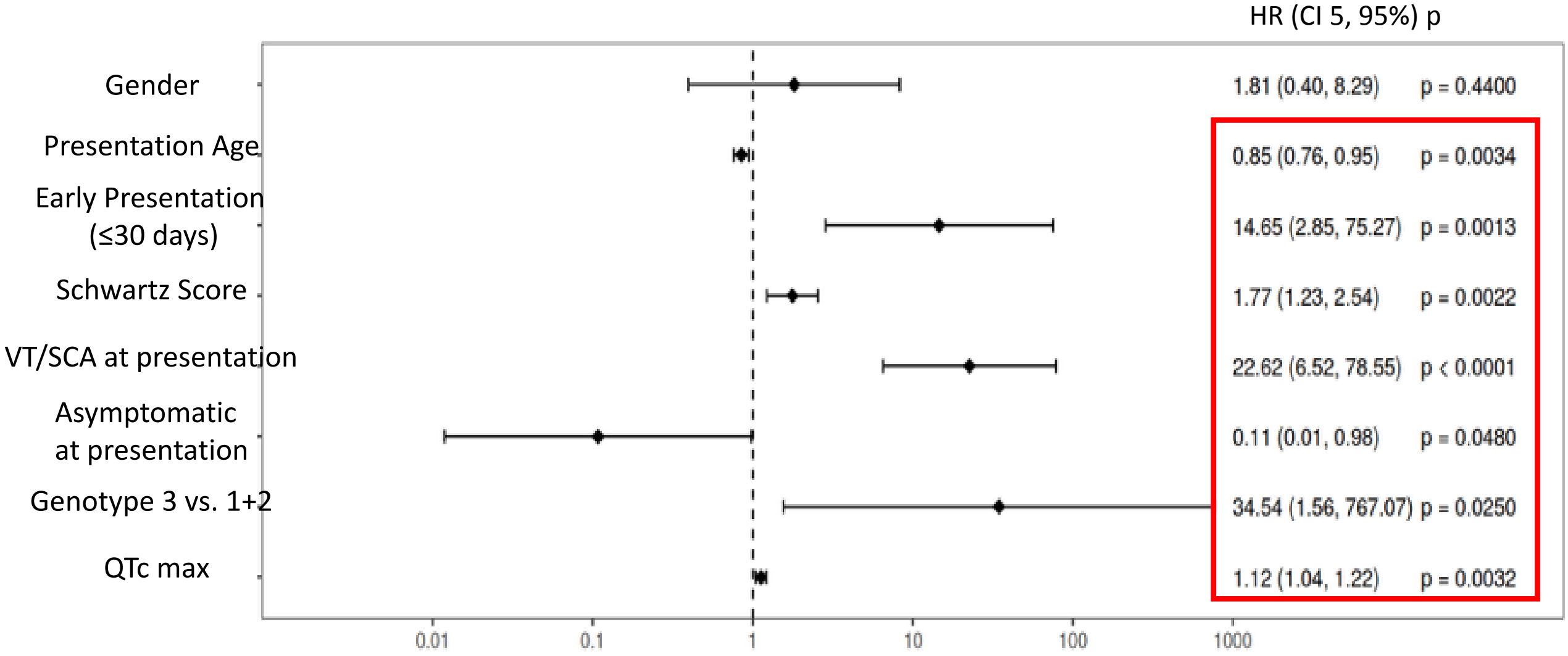
Survival Probability – Death from any cause



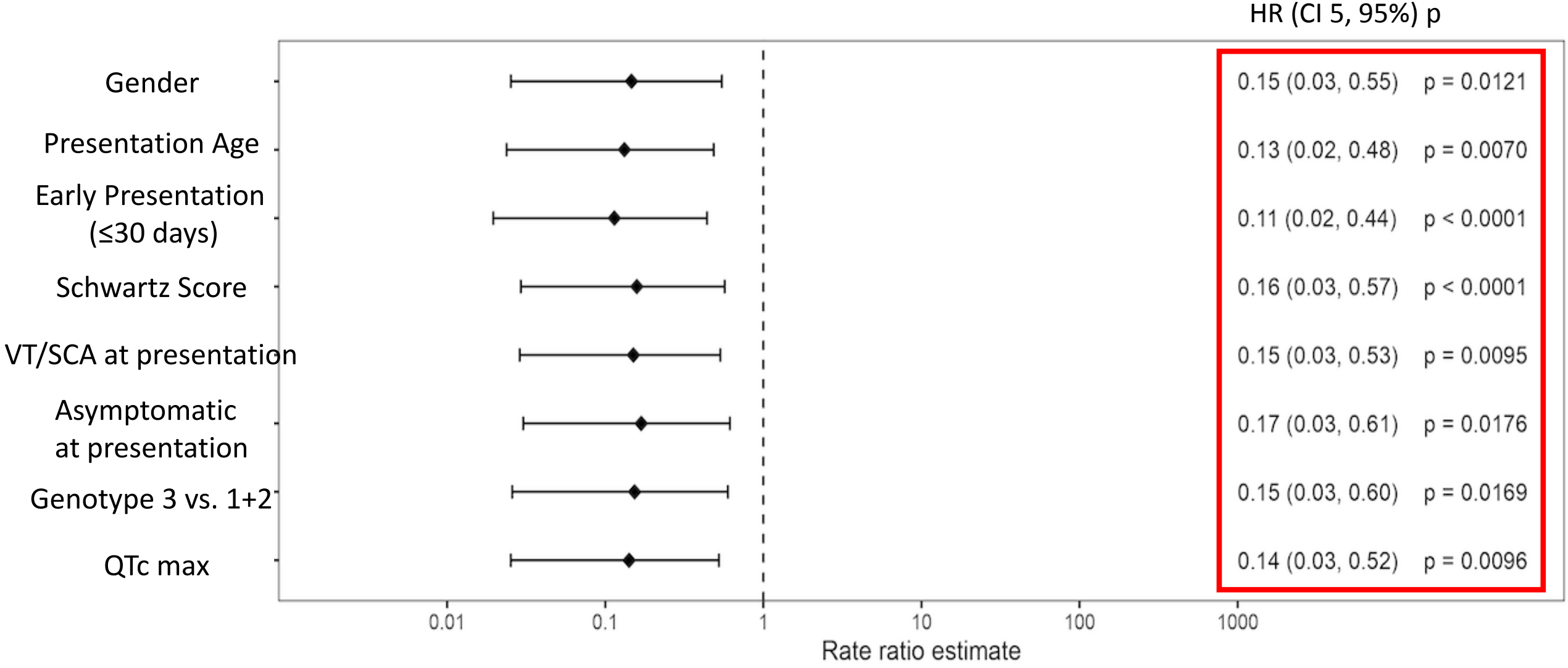
Survival Probability – MAE Presentation age



Hazard Ratio Estimates - MAE



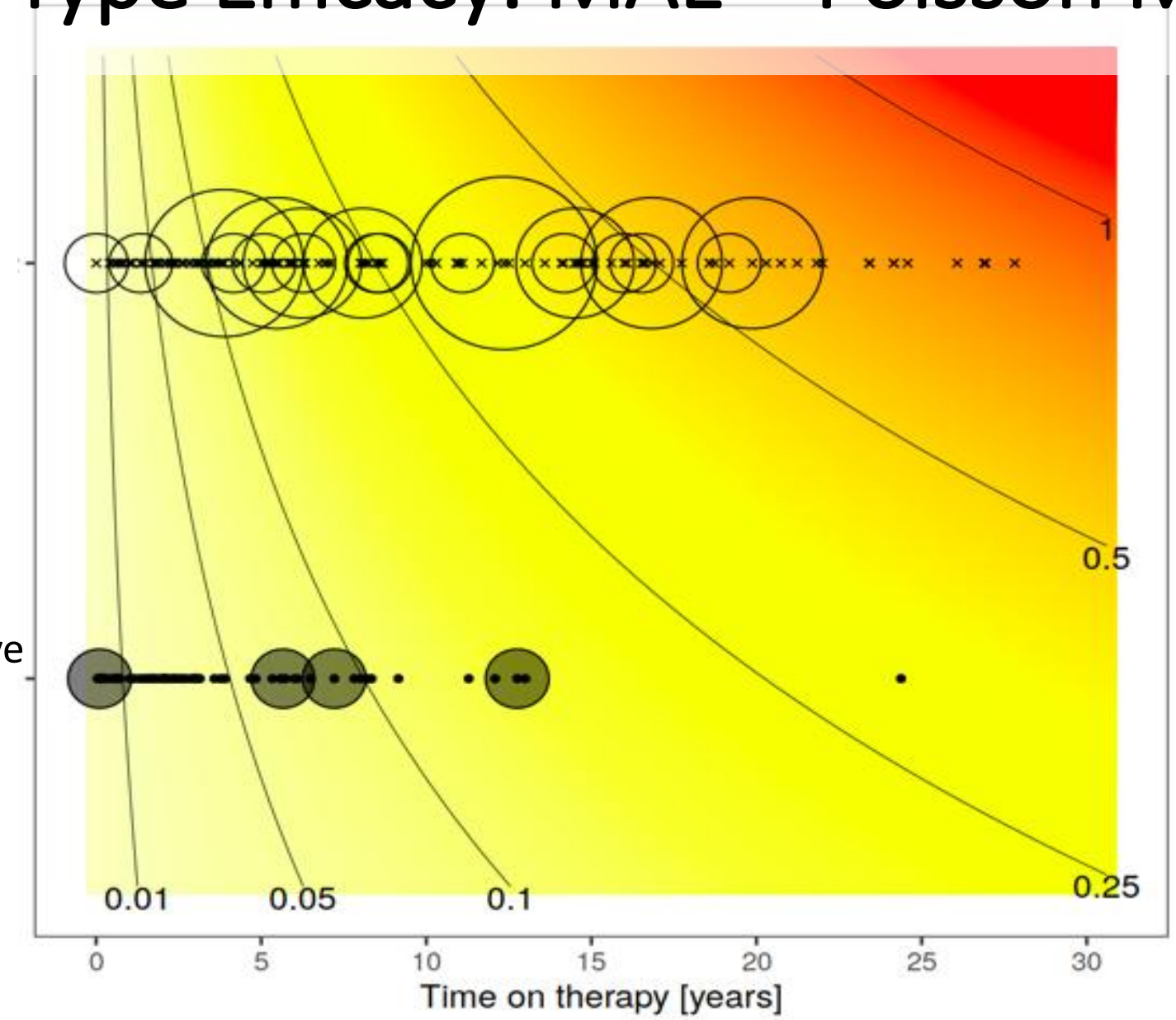
Hazard Ratio Estimates - MAE : Non-selective vs. selective β-blockers



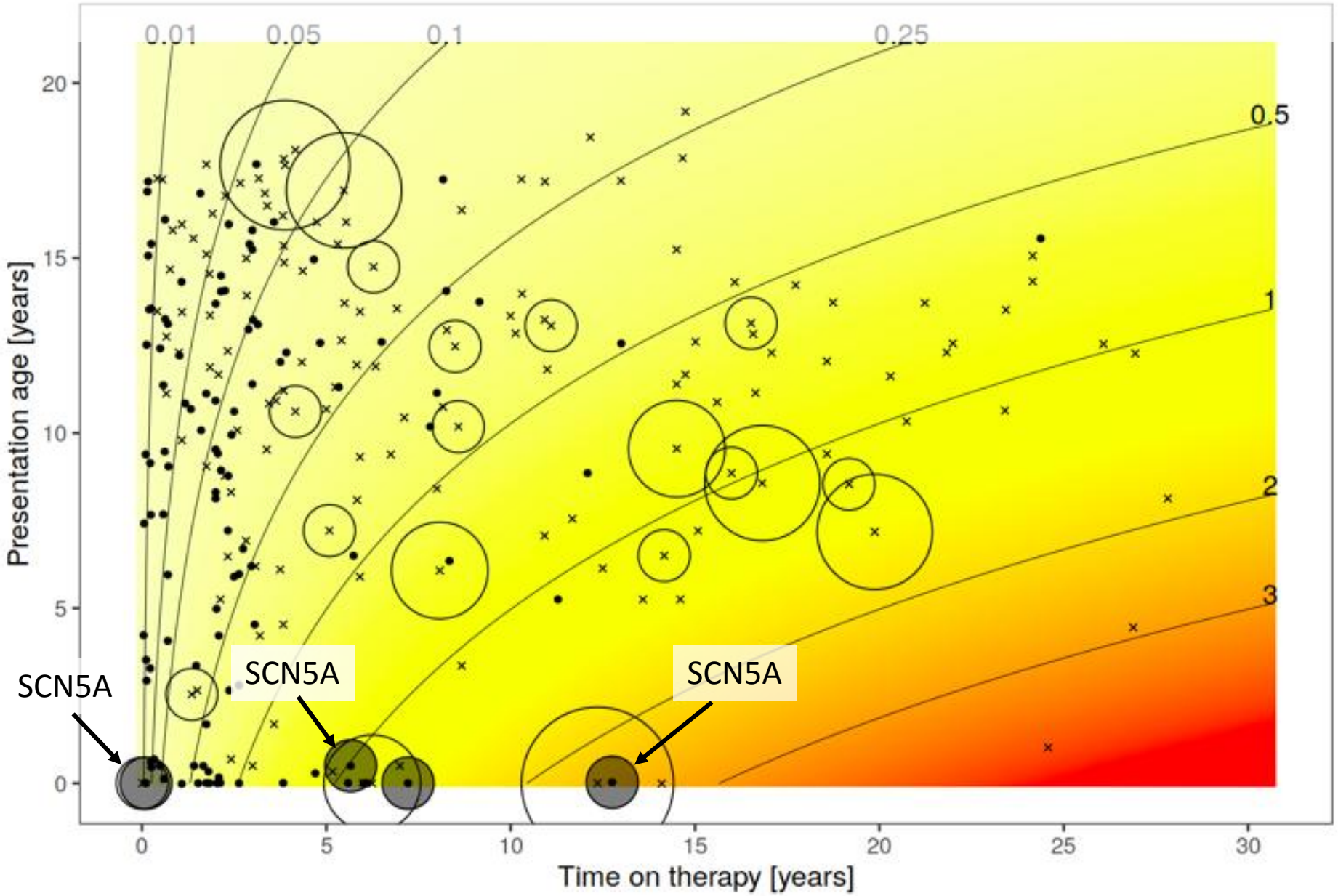
β-blocker Type Efficacy: MAE – Poisson Model

× selective β-blockers

● non-selective β-blockers



Presentation Age: MAE – Poisson Model



× selective β-blockers

● non-selective be β-blockers

Conclusions

- Overall survival probability at 20 years 90%
- LQTS 2 and 3 more prevalent in patients presenting as newborns
- Increased risk of MAE
  - Early presentation
  - Symptoms at presentation
  - High Schwartz score
  - Longer QTc
  - LQTS type 3
- Non-selective β-blockers more effective than selective β-blockers regardless of other variables
  - Decrease MAE burden

Thank you for your attention

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