

## C OUTCOMES OF A NATIONWIDE CARDIAC SCREENING PROGRAMME IN YOUNG INDIVIDUALS

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10.1136/heartjnl-2022-BCS.227

**Background** There is limited information on the impact of screening for conditions associated with sudden cardiac arrest (SCA) and sudden cardiac death (SCD) in young individuals, outside the context of elite sport. We sought to determine the diagnostic yield of cardiac screening in young individuals and the incidence of SCA and SCD amongst individuals who underwent cardiac screening.

**Methods** From 2007 through 2018, 104,369 consecutive individuals, aged 14 to 35 years, underwent voluntary cardiac screening (62% were male, 91% were non-athletes). Initial evaluation consisted of a health questionnaire (HQ), electrocardiogram (ECG) and clinical consultation. Selective on-site echocardiography was available at the discretion of the consulting physician. Clinical outcomes were sourced from the Office for National Statistics-Hospital-Episode-Statistics database and an online HQ.

**Results** During screening 280 (0.27%) individuals were found to have a cardiac condition associated with SCA/SCD and 115 received potentially life-saving treatments. A further 166 (0.16%) with congenital or valvular abnormalities were identified. During a mean follow-up of  $6.2 \pm 2.5$  years, an additional 86 individuals, considered to have normal screening, were found to have a cardiac condition associated with SCA/SCD. After screening there were 86 deaths. Cardiac disorders accounted for 20 deaths (23%), all of which were sudden. In addition, 15 individuals survived a SCA. Primary electrical disorders accounted for 21 (60%) of the 35 cases of SCA/SCD, followed by cardiomyopathies (n=6), and congenital or acquired coronary artery disease (n=5). On the basis of a total of 626,550 person-years (PY), the combined incidence of SCA/SCD was 1:17,901 PY (5.6 per 100,000 PY). Males were at 3.5-fold higher risk than females (1:12,488 PY vs 1:44,067 PY,  $p < 0.01$ ). For prevention of SCA/SCD the programme's sensitivity was 19.4% with a specificity of 97.5%. For identifying conditions associated with SCD, the programme's sensitivity was 76.5% with a specificity was 97.8%.

**Conclusion** Diseases that are associated with SCD were identified in 0.27% of young individuals who underwent cardiac screening. A further 86 (0.08%) conditions were identified after screening. The incidence of SCA and SCD was 5.6 per 100,000 person-years, which is considerable higher than previous estimates in the general population. Most of these events were due to confirmed or presumed primary electrical disorders that had not been detected on screening.

## D A NOVEL INTERNALLY VALIDATED RISK PREDICTION MODEL FOR ADVERSE CARDIAC OUTCOME IN FABRY DISEASE

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10.1136/heartjnl-2022-BCS.228

**Background** The cardiac manifestations of Fabry disease are the leading cause of death, but risk stratification remains inadequate. Identifying patients who are at risk of adverse cardiac outcome may facilitate more evidence-based treatment guidance. Contemporary cardiovascular magnetic resonance imaging (CMR) biomarkers have become widely adopted but their prognostic value remains unclear.

**Objectives** Our objective was to develop, internally validate, and evaluate the performance of, a prognostic model, including contemporary deep phenotyping, which can be used to generate individual risk estimates for adverse cardiac outcome in patients with Fabry disease.

**Methods** Longitudinal prospective cohort study of 200 consecutive patients with Fabry disease undergoing clinical CMR. Median follow-up 1,640 (987 – 2,293) days. Prognostic models were developed using Cox proportional hazards modelling. Outcome was a composite of adverse cardiac events. Model performance was evaluated.

**Results** The highest performing, internally validated, parsimonious multivariable model included age, native myocardial T1 dispersion (standard deviation of per voxel myocardial T1 relaxation times), and indexed left ventricular mass. Median optimism-adjusted c-statistic across 5 imputed model development datasets was 0.77. Model calibration was excellent across the full risk profile. A risk calculator, which provides 5-year estimated risk of adverse cardiac outcome for individual patients, including males and females, was generated.

**Conclusions** This study developed and internally validated a risk prediction model that accurately predicts 5-year risk of adverse cardiac outcome for individual patients with Fabry disease, including males and females, which could easily be integrated into clinical care. External validation is warranted.