Introduction

Sudden cardiac death (SCD) in young people is a tragedy for families and communities alike. It is often due to an underlying genetic arrhythmic condition. In 2008, we identified a significant genetic defect (TMEM43; P358L) in patients and extended relatives from 22 unrelated families with Newfoundland ancestry. We showed that the clinical consequence of harboring the TMEM43 mutation is early SCD (50% of untreated males deceased by 40 years, 80% by 50 years). One study from Ontario using a 2008 cohort assessed the...
incidence of potential SCD in persons aged 2–40y to be 14.8 per 100,000. We hypothesize that NL may have a higher incidence of early SCD in this age category due to TMEM43 and possibly other underlying genetic causes, given the historical genetic isolation of the population.

Methods Following local REB approval (#12.199) we ascertained cases of sudden death from the comprehensive Medical Examiners provincial database. Cases that satisfied the following three inclusion criteria were reviewed: 1) date of death in 2008, 2) age at death aged 2-40y inclusive, and 3) manner of death listed as “natural-cardiac”, “natural-other”, “accidental”, or “undetermined”. Each case was then individually analyzed by the research team to determine likelihood of SCD.

Results All the 2008 NL cases were ascertained, and all (anonymous) data maintained on an SPSS dataset. There were 12 “natural-cardiac” cases, 8 “natural-other”, 48 “accidental”, and 2 “undetermined”, giving a total of 70 'potential' SCD cases in those aged 2–40y. The Ontario study yielded 976 'potential' cases. The 'potential' incidence of sudden death for Ontario of 14.8 per 100,000 compares with 30.1/100,000 in NL (with denominators of 6,602, 680 and 232,210 derived from statistics Canada for the provincial populations between 2–40y respectively). This represents a significant increase of potential SCD in NL (p=0.02).

Conclusion These figures include cases which are likely not SCD’s. However, the methodology used to compare the populations was exactly the same. It appears therefore that NL had more deaths in 2008 in this age cohort than Ontario. The reasons for this difference are yet to be determined.