PE2067/F: Improve data on young people affected by conditions causing Sudden Cardiac Death

Cardiac Risk in the Young (CRY) written submission, 21 April 2024

Thank you for raising this issue in Scottish Parliament and the comments which were raised on 20th March 2024 in response to the petition.

Our response to the document will be around two concerns; how the incidence of young sudden cardiac death (YSCD) is **calculated** and **understood.**

How the incidence of YSCD is calculated

The focus of this petition is to correctly establish the incidence of Young Sudden Cardiac Death (YSCD) and the most appropriate measures to identify those at risk prior to a cardiac arrest or death. This is of utmost importance because government advice has consistently failed to address the first, and most important, question of how many young people this affects and therefore grossly underestimated the impact these conditions are having on families, communities and wider society. This point is clearly demonstrated in the SPICe briefing, where an FOI in June 2023 reporting 19 myocardial infarctions in 0-29 year olds in 2022 has been referenced. This is an example of how the data informing policy, and being given in response to politician's questions, underrepresents the incidence and the impact of these deaths. It is understood by specialists that there are inaccuracies in the way YSCDs are recorded within the Office of National Statistics (ONS), with the mortality statistics largely derived from documentation on death certificates which may under-report the true incidence of cardiac arrhythmias. This was discussed by Papadakis et al., (2009) (acknowledged in the SPICe response), which evaluated the ONS data and reported cardiac death rates in England and Wales.

The recent paper of the largest SCD cohort with autopsy findings ever reported (N=7,214), between 1994 and 2021, (Sheppard et al, 2023) presents SCD figures based on examinations (macroscopically and microscopically) by two expert cardiac pathologists. When compared to the FOI in June 2023 it highlights the significant disparity between government advice and actual death rates. The paper presents data on referrals to the centre by year, age groups, gender and conditions. 49% of the cases referred to the centre were under the age of 35 (N=3,547) and in 2021 there were 499 hearts were referred to the centre, 634 hearts in 2020. It is important to note this centre does not see all cases of SCD in the UK.

How the incidence of YSCD is understood

Whilst the SPICe briefing has provided an explanation of the statistical expression of "number of deaths per 100,000 people/year" it is unclear why they have used a hypothetical scenario of diabetes to illustrate this. Furthermore, notwithstanding the error in the 3rd bullet point, the way they have presented this does not enable the following question to be answered, ***based on the evidence which is supporting the NSC's policy and understanding of the incidence of Young Sudden Cardiac Death, how many of the young people who are aged 14 today are anticipated to die before their 36th birthday of a heart condition.***

When trying to evaluate the impact of YSCD it is necessary to understand the incidence of YSCD. Many people appear to believe there are only 2 per 100,000 people dying of these conditions. However, they are confusing incidence, incidence rate and cumulative incidence. These are common measures used in Epidemiology. The confusion is when 2 different statistics are conflated, "XX per 100,000 people" with "XX per 100,000 people **per year**". 2 per 100,000 people **per year** is the **incidence rate** (also known as **incidence density rate** or **person-time incidence rate**). However, in the case of young sudden cardiac deaths, the person is at risk of dying over many years, not just one year. In the case of CRY's screening programme it is testing people between the age of 14 and 35, covering a 22 year period of risk. It is therefore important to calculate the **cumulative incidence** (or **incidence proportion**).

A crude approach to extrapolating an estimated incidence rate of 2 deaths per 100,000 people per year, over 22 years of being at risk, would estimate approximately 44 deaths per 100,000 people, 1 in 2,272 people, dying of the conditions between the age of 14 and 36. This is oversimplified on a number of accounts, including incidence rates are likely to differ significantly by age, gender, lifestyle and ethnicity. However, it highlights how a person may perceive the risk of SCD to be very differently if they understood it to be 1 or 2 per 100,000 people, as opposed to 1 in 2,272 people, or more for higher risk groups (e.g. male athletes).

In Italy, the incidence rates of young people involved in organised sport (prior to the implementation of their screening programme) was 3.6 per 100,000 per year (Corrado et al, 2006). In the case of screening elite footballers Malhotra et al, (2018) reported an incident rate of 6.8 per 100,000 per year, post screening. This illustrates that male elite athletes are at significantly higher risk (compared to the general population), and this figure was calculated after cardiac screening had identified 42 high risk cases.

Whilst this analysis has evaluated the risk of dying before the age of 36, it is important to recognise there is a **lifetime risk** from these conditions, with sudden deaths from conditions causing SADS and cardiomyopathies occurring after the age of 35 (Sheppard et al, 2023).

The error, or misunderstanding of the incidence of YSCD, is why we have made repeated requests to meet with the National Screening Committee to clarify this issue, so politicians are correctly briefed. We have also requested for the NSC to transparently publish the pre-screening and post-screening incidence death rates for other conditions which meet the NSC screening criteria.

There are a number of issues and inaccuracies raised in the SPICe briefing which have been addressed previously in our response(s) to the National Screening Committee (for instance much of the research is not limited to "professional" athletes, it is young people involved in organized sport). These can be read on the CRY website.