

Cardiac Risk in the Young (CRY)

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The story of a young boy's cardiac disease and the parental response initiated the charity organisation CRY, today a primary force regarding prevention of sudden cardiac death in the young

[CRY](#) works with cardiologists, family doctors, families and the bereaved to promote cardiac health of young people. CRY's screening programme gives the opportunity to identify any young person who may be at risk, regardless of sporting ability, level or sport participation at all. The organisation offers support to all people affected by cardiac conditions that can cause young sudden cardiac death. CRY (a thought-provoking abbreviation) offers a unique bereavement support programme and supports expert fast-track pathology and fast-track cardiological referral after a young sudden death, when symptoms present or when a family member has been identified with an inherited cardiac condition.

A number of worldwide events that came together in the early 1990s led to the development of CRY. Read this interview with Dr. Steven Cox, Deputy Chief Executive and Director of Screening, CRY.

"Steven's story

In 1990, aged 16, I relocated to the US to take up a University tennis scholarship. I attended the leading US junior college for tennis. In 1992 I was required to have a full medical with a battery of different health checks (including an ECG) as part of the scholarship agreement. The ECG was identified as abnormal so I was referred for further tests. This was not taken seriously within the team as I was one of the fittest athletes and no one had heard of young sports people being affected by heart conditions. At the meeting with the cardiologist he apologised for the inconvenience and said "this will just take a few minutes, the nurse who took your ECG put the leads on the wrong way round" - then, "Oh, she didn't put the leads on the wrong way round". After many tests over the following month the cardiologist enthusiastically approached me in the lobby, asked a few seemingly meaningless questions and then

said "I've finally found out what is wrong, you have Arrythmogenic Right Ventricular Cardiomyopathy (ARVC). " I smiled and said "great, so what does that mean?" to which he replied, "you'll never be able to exercise or sweat again."

The cardiologist didn't appear to understand the impact such a statement could have on an 18 year old who for 10 years had been aspiring to be a professional tennis player. I was not allowed to step foot on the University tennis courts for legal reasons and my identity of being a great athlete was now in question. Thankfully an ex tennis player and cardiologist at the hospital heard about my story and took me under his wing to explain what had happened and why sport and certain lifestyle decisions were dangerous. The difference this doctor made to my life was transformational.

While I was still in the US, Alison, my mother, researched who was the leading specialist in this area. Professor Bill McKenna was based at St George's Hospital, 30 minutes from where we lived, and when I returned I had my first appointment. She asked Prof. McKenna to write up ARVC in layman's terms, which became the first patient information for ARVC. After a battery of further tests the doctors were still not happy for me to return to sport. At the same time John Marshall aged 16 died suddenly of ARVC. John was one of England's most promising young footballers who had just signed for Everton. John's and my story lead to a media storm and families reading the stories in the papers started to contact my mother saying I had been lucky because their child had died suddenly for no apparent reason.

Foundation of CRY

Alison had recently finishing her training to become a counselor for children who were terminally ill and found herself supporting the families that contacted her. A year later, in 1993, she held the first screening in the UK for the Lawn Tennis players at the National Championships. Alison collated and printed information written by "experts" so that she could understand ARVC, WPW, Long QT and other difficult diagnoses, and these acted also as a resource for bereaved families. She started fundraising for an echo machine that could be used at the first "family" clinic for young sudden cardiac death in St George's Hospital and at the weekends be used for

screenings around the country on request. In 1995, she officially founded and launched the charitable organisation Cardiac Risk in the Young (CRY) to support bereaved families and offer a screening service to proactively identify young people <35 at risk of sudden death.

A few years later, educated in psychology, I began to work at CRY. I evaluated the psychological impact of cardiac screening and looked at the psychological impact of living with hypertrophic cardiomyopathy and later completed a PhD.

Young Sudden Cardiac Death – the CRY experience

Through working closely with Professor Sanjay Sharma, CRY's consultant Cardiologist, the screening programme has evolved and grown exponentially and in 2012 we will test well over 10,000 young people. The majority of the people that CRY tests, will have just an ECG and consultation with a specialist doctor. Some however, need further tests on the basis of family history, symptom questionnaire and/or an abnormal ECG. The majority of the service is funded by bereaved families, research grants and sponsorship.

One of the most important aspects of the CRY programme is the role of the specialist doctor at the screening event. By meeting with each person as they are tested, reviewing their ECG and discussing their symptoms, as well as overseeing their echos when required, we are able to reassure the vast majority of people on the day of the testing. We are also able to minimise the number of people referred for further tests.

From our experience, 1 in every 300 of the young people that CRY tests will be identified with a potentially life-threatening condition. 1 in 100 will have a less serious condition that can be monitored and corrected through surgery if required. What the long term prognosis of each of these people would have been if not identified is an ongoing debate.

At CRY we are supported by over 2000 families affected by a young sudden cardiac death, going back 25 years. Each year 200 more families will contact us seeking support, increasing our understanding of the scope of these tragedies. This gives us a

unique insight into the incidence of these deaths and every year this picture becomes more complete. The vast majority of these deaths are not in elite athletes which is why CRY has always focused on the community rather than sports participation.

I wanted a better understanding of my statistical probability of sudden death due to ARVC, and how this would be modified based on whether I continued playing competitive sport or not. I had aimed to continue my tennis career if the odds were encouraging. 20 years later, this question is unimportant personally, but it is still central to the debate about the role of cardiac screening in preventing sudden death.

People still seem to think that these deaths are rare affecting only 1 in 50,000 to 1 in 200,000 athletes. However, based on just those families that have contacted CRY and are raising awareness and supporting the charity, we know this is just a fraction of the problem. Furthermore, we also know that the CRY supporters only represent a small proportion of the people who have suffered bereavements in the UK. Many will visit our website accessing everything they need and not get in contact with us.

When CRY started, anecdotal evidence suggested there was at least 1 sudden death in the UK every week from an undiagnosed cardiac condition. A few years later this escalated to 4 deaths a week, then to at least 8 deaths a week and now we know it is at least 12 deaths a week in the UK. CRY is contacted by at least 4 families every week where a child or partner under the age of 35 has just died due to a cardiac condition or the cause of death after expert pathology is still unascertained.

Such deaths are devastating for thousands of people every year in the UK, including parents, siblings, children, grandparents, friends and the local community. It is important when we evaluate the impact of a screening programme, that we take into the consideration the whole picture. This is not just about how much it will cost to reduce the burden of mortality due to the young dead, it is about quantifying the full cost to a family and the community.

The CRY Centre for Inherited Cardiovascular Conditions and Sports Cardiology

The CRY-founded Centre operates under the direction of Professor Sanjay Sharma at St George's Hospital and provides fast track referral after a young sudden death or when a young person is symptomatic.

At this centre the family will have all their tests conducted together on the same day regardless of where they live and often includes tests like provocation and MRI. They also see a specialist and receive a diagnosis at that first appointment, which is usually less than 4 weeks after receiving a referral from their General Practitioner.

CRY's bereavement support

Following a tragedy in a family where a young person has died suddenly, family members often require support. From a medical perspective they need to understand the potential genetic risks that may face other family members if affected. CRY provides this support by the provision of specialist cardiac information written by experts in the field, specifically for families or a non-medical community.

CRY has a selected group of bereavement support volunteers who have undergone a similar tragedy themselves and been specifically trained by CRY, acquiring accredited counselling qualifications to help others cope with their traumatic experience. CRY also offers other opportunities for bereaved families to come together including at National and Regional Bereavement Support Days around the UK.

CRY Centre for Cardiac Pathology

The importance of correct pathology cannot be overstated as it gives families the opportunity to obtain valid answers to the cause of death and to assess the risk posed to other family members. When a cause of death is unascertained and the person is < 35 years, the centre will provide a free fast-track cardiac diagnostic service. The examination and report from the centre will be issued on average within 2 weeks. Prior to this free fast-track service being provided for coroners it took up to 2 years for an expert investigation to be conducted. Consequently few families would allow the whole heart be examined and coroners were reluctant to ask families if

they would permit this. Having the centre has revolutionised the diagnosis and understanding of the incidence of these conditions. CRY funds the staff at the CRY centre based at The Royal Brompton Hospital, London.

Supporting those diagnosed - the myheart network

CRY runs support days for members of the myheart network. These young people (35 and under) have had, or are about to confront, potentially life-saving surgery and get the opportunity to talk to others who have been through a similar experience - including fitting of pacemakers, implantable defibrillators, ICD and ablation surgery. The group was set up after feedback from young people who found that the existing support groups available for ICD recipients, and other cardiac conditions, were unhelpful. It was developed as a support system that increases effective coping and decreases social isolation for young implant recipients, whilst also training young people to be able to offer support to others in a similar situation. They are offered 'Question and Answer' sessions with a specialist cardiologist; group counselling to talk through recent difficult social experiences; opportunities to meet other members; and their own myheart newsletter; opportunity to listen to guest speakers.

CRY in the future

About 80 % of young people that die suddenly have no prior symptoms and proactive screening provides the only opportunity to save these lives. In the future we expect to establish the true prevalence and impact on society of these deaths through further research; identify the most cost- effective way of identifying young people with the cardiac conditions that can result in young sudden cardiac death, and support them to adjust to their condition with a good quality of life after a condition is identified."

A story to learn from

It is thought-provoking that it needed a personal history and a motherly fight and responsibility for her child to create a thorough and high-quality care of the sudden cardiac young victims. In many countries, the authorities have not taken this liability. The CRY story,

also focusing the balance between private initiative and the responsibility of the authorities, has something to tell us.

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The content of this article reflects the personal opinion of the author/s and is not necessarily the official position of the European Society of Cardiology