**CRY Screening : FAQs**

**What is sudden cardiac death?**

Sudden cardiac death can be simply defined as a sudden death from a cardiac problem within 12 hours of witnessed normal health.

**What is SADS?**

SADS stands for Sudden Arrhythmic Death and this is part of sudden cardiac death. However, SADS is defined as a sudden death whereby subsequent post mortem analysis and a toxicology screen fails to identify any obvious cause.  
In our experience, almost all of these conditions are due to an electrical fault that cannot be picked up at post mortem. So sudden adult death syndrome is part of sudden cardiac death but does not make up all the sudden cardiac deaths because sudden cardiac deaths are due to not just SADS but also the cardiomyopathies, the anomalous coronary arteries, premature coronary artery disease and valvular diseases as well.

**How important is expertise?**

Expertise is extremely important, in fact expertise is the crux of our screening programme – without expertise, one opens themselves up to litigation.  
The conditions that we’re dealing with are rare, one considers that the prevalence of hypertrophic cardiomyopathy is one in 500, the prevalence of ARVC is one in 1000, the prevalence of Brugada syndrome is one in 2000. So most general cardiologists will not have seen many conditions such as hypertrophic cardiomyopathy. It’s also important to be aware that these conditions manifest in many, many different ways, they’re very heterogeneous.

Let’s take hypertrophic cardiomyopathy for example. Some individuals may develop very severe left ventricular wall thickness – which is very easy to recognise by everybody – but in others, there would be no increase in left ventricular wall thickness, the only manifestation of the condition will be an abnormality on the ECG, and many cardiologists are not aware of this. Similarly, arrhythmogenic right ventricular cardiomyopathy may require numerous investigations before it can be diagnosed. Long QT syndrome may fail many cardiologists; they may fail to diagnose it because of the various manifestations and the morphology of the T-wave. So I believe expertise is extremely important.

It’s also important to be aware that people who exercise a lot have to develop an increase in heart size. That increase in heart size may reflect on the ECG and in rare instances, may overlap with findings seen in people with hypertrophic and arrhythmogenic right ventricular cardiomyopathy. Fortunately, experts in sports cardiology have the knowhow of differentiating between physiology – that is, adaptation due to exercise – from pathology; and that expertise is very limited currently in the United Kingdom. The other important issue is about the impact of exercise on the heart. People who participate in a lot of sport will have larger hearts than the general population and this increase in heart size is sometimes reflected on the ECG.

There are some athletes who harbour ECG changes or manifest ECG changes that overlap with those seen in individuals with hypertrophic cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy. In sports cardiology, there are various algorithms that facilitate the differentiation of physiological changes due to exercise, from pathological ones from hypertrophic cardiomyopathy or arrhythmogenic right ventricular cardiomyopathy. Most general cardiologists are not familiar with these algorithms and could make mistakes in this situation. It is also important to be aware that in the context of long QT syndrome for example, the resting ECG may just raise the suspicion of the condition but further specialist tests may be required to show up the syndrome. For this reason, I think expertise is absolutely vital.

The other thing that one needs to consider is the impact of age, gender, size and ethnicity on the ECG. We know that childhood athletes have very different ECGs compared to adult athletes. Black athletes have very different ECGs compared to white athletes. Male athletes have very different ECGs compared to female athletes. Knowledge regarding cardiomyopathy, physiological cardiac adaptation, determinance of ECGs, the phenotypic manifestations of all of these conditions that cause sudden cardiac death is absolutely vital if we’re going to do this properly and reduce the risk of false positives and false negatives which could have very serious consequences.

**Does playing sport cause sudden cardiac death?**

There is a relationship between sudden cardiac death and sport. However it is important to emphasise that sport is good for the heart. Sport reduces the risk of high blood pressure, it improves cholesterol levels and generally, it reduces the risk of someone dying from a cardiac problem. Indeed people who play sport on a regular basis live an average of six years more than people who don’t play any sport at all.  
However, if an individual that plays sport harbours a condition that’s capable of causing sudden cardiac death – such as hypertrophic cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy or a blocked artery – then sport is three times more likely to kill that individual. So there’s a bit of an irony, sport promotes longevity of life and reduces the risk of death from a cardiac problem but increases the risk of sudden death in an individual who already has a cardiac condition.

I don’t believe that sport per se actually causes a cardiac problem that will pre-dispose to sudden death.

**Why get tested? I am told there is nothing you can do for me if I am identified.**

The antagonist could argue that screening for cardiovascular disease is a waste of time because once a disorder is identified, nothing can be done to prevent sudden death. I need to wipe these types of statements out, completely abolish them because there’s a lot that can be done otherwise there’d be no point screening.  
We can actually prevent deaths and this has been shown internationally, certainly in athletes where a 25 year screening programme performed in the Veneto region of Italy has shown that death rates were reduced by 90% over a 25 year period in individuals who had been screened. In fact, death rates now in athletes in Italy are less common than in non athletes, even though athletes put themselves through such gruelling exercise. That’s because most conditions that cause sudden death have been sifted out through screening, whereas the non athletes aren’t screened. So, death rates in the non athlete group in Italy are higher than in the athlete group.

There’s a lot that can be done and this can range from lifestyle modification, drug treatments and internal cardioverter defibrillators. In some cases, we can do radio frequency ablation with electrophysiological studies and some may be amenable to cardiac surgery.

**What test will I have and how long will it take? What will they be looking for?**

Screening for cardiac diseases can be a complex situation but we are trying to adopt a cost effective screening programme and this screening programme comprises of a health questionnaire which enquires specifically about the cardinal symptoms of cardiac diseases such as chest pain, breathlessness, dizziness, palpitations or black outs and also importantly enquires about family history of cardiac conditions, because most conditions that cause sudden death in young people are hereditary – by that I mean they are genetically transmitted from one person to another so the family history's very important.  
That is usually followed by a 12 lead ECG which is an electrical tracing of the heart that is designed to look for electrical faults of the heart and to raise suspicion of the cardiomyopathies. So the whole process doesn't take long and in many situations, the individual can be reassured on the spot, a small number of people, I would say 4%, need further tests. Of these 4%, 0.2% have a serious problem, about 1% have a minor problem and the other 2.8% can be reassured.

**Who will be reading my test?**

At the CRY screening programme, all ECGs are read by a cardiac research fellow. ECGs deemed remotely abnormal are read by me. So, everybody with a remotely abnormal  
ECG has the ECG read by a junior cardiologist and a very senior cardiologist and based upon this, we make decisions regarding further investigations.

**Do these tests identify all people at risk?**

The current screening programme, which involves an ECG and a health questionnaire, will not identify all conditions that cause sudden cardiac death. There are conditions such as anomalous coronary arteries and premature coronary artery disease that will be missed by our screening programme. Having said that, conditions like the cardiomyopathies and the ion channel disorders and Wolff-Parkinson-White will be identified and so will Marfan syndrome. So most conditions will be picked up but it is true to say that some will be missed.

**What are the chances of having something wrong?**

There are several things that can cause cardiac problems in young people. These can range from very serious conditions such as long QT syndrome or hypertrophic cardiomyopathy or minor conditions such as mitral valve prolapse. By that I mean a floppy mitral valve or a small atrial septal defect, by that I mean a very small hole in the heart.  
Some conditions need a lot of treatment or input medically, some conditions don't need any, just caution in terms of certain lifestyle issues. But if we took into account all the conditions that could potentially cause either fatality or even morbidity in the future, then I would say about one in 200 would be identified with something. Some of these could be mild things such as just congenital abnormalities of the valves.

**Do I need to bring anything with me for the test?**

There is no need to bring anything with you, apart from yourself. This screening programme involves filling out a health questionnaire so it's important to bring details about your General Practitioner, any relevant medical conditions that you may have.  
Any medications that you may be taking, you need to be aware of what they are or bring them with you and that's about it because the rest of it is an ECG which just involves simple leads being placed on the chest wall for about two minutes, there's nothing to the screening programme at all.