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Offering help and support to affected families

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The CRY International Medical Conference on 9th October 2015 was the 10th since the meeting was first established in 2006. A packed auditorium was again presented with cutting-edge research from renowned experts, before a trademark case studies session featuring current CRY Research Fellows presented challenges in diagnosis and management.

Former CRY Fellow Dr Ahmed Merghani presented first at the conference; his 2015 research on the potential dangers of chronic extreme exercise for veteran athletes had already stimulated debate in the press on both sides of the Atlantic. Following was Italian expert Professor Domenico Corrado, who focussed his presentation on the implications of scarring on the left ventricle of the heart. Such scarring may be associated with life-threatening ventricular arrhythmias, and he warned that it shouldn’t be dismissed as a benign feature of healed myocarditis (inflammatory heart disease).

Professor Mary Sheppard, CRY Consultant Cardiac Pathologist, returned to the conference in 2015 to deliver her unmatched experience in the detection and diagnosis of cardiac disease at post-mortem. CRY’s Consultant Cardiologist Professor Sanjay Sharma then gave a presentation on guidelines for differentiating between hypertrophic cardiomyopathy (HCM) and exercise-related cardiac adaption in athletes. He also made mention of the privileged position the CRY centre’s expertise puts them in, saying, “We are very fortunate at St George’s because we get referred practically any athlete with anything that looks remotely ‘bad’. “ Working at St George’s alongside Professor Sheppard, who receives referrals from 80% of coroners in the UK, CRY doctors have a unique advantage in the field.

Dr Michael Papadakis, who chiefly organises the CRY Conference each year, announced that “CRY has made a significant contribution to the development of [ECG] criteria”, referring the continued refinement of screening guidelines that help save lives whilst driving down the cost of tests. He offered a glimpse of further refined criteria from the CRY Research Group due to be published in 2016.

Another colleague at St George’s, Dr Elijah Behr, delivered a comprehensive assessment of advisable exercise recommendations for both cardiomyopathies (heart muscle diseases) and ion channelopathies (electrical heart diseases), highlighting the many factors which should influence a cardiologist’s assessment of risk: from the source of diagnosis to the specific condition diagnosed; from the dynamic/static nature of the sport undertaken to whether it is recreational or competitive. Professor Mats Börjesson followed with a presentation on exercise recommendations for athletes with hypertension (high blood pressure) or ischemic heart disease.

CRY Director of Screening and Research Dr Steven Cox gave an overview of CRY’s history and the services they now provide, paying tribute to the bereaved families who back the charity so vociferously and the young people in whose memory the achievements have been made. He also explained the critical role that progress in scientific research and clinical practice has in forcing through policy changes, highlighting examples of forward progress from CRY’s 20 years of awareness, support, screening and research.

After lunch, the conference continued with a presentation on HCM obstruction by Dr Antonis Pantazis from the Royal Brompton and Harefield hospitals. Professor Angeliki Asimaki, a research associate at Harvard Medical School in Boston, offered an explanation of her investigation into a drug to combat the arrhythmias brought on by arrhythmogenic right ventricular cardiomyopathy (ARVC). Then University Hospital Birmingham’s Dr Sara Thorne gave a presentation on the risks cardiomyopathies and arrhythmias pose for pregnant women, covering two very different circumstances for doctors to be prepared for: when a condition is already known to the woman before pregnancy; or, more dangerously, when the condition makes itself known during the pregnancy’s course.

For more about the CRY Conference visit www.c-r-y.org.uk/cry-international-conference

# Presentations

**Inflammatory topics in sports cardiology**

“The veteran heart; Is old really gold?” Dr Ahmed Merghani

“A view from the outside in; Epicardial scarring in the elite athlete” Professor Domenico Corrado

“A view from inside out; Post-mortem scarring in the athlete” Professor Mary Sheppard

**Assessment of the athlete; what to do now?**

“Differentiation of athlete’s heart from cardiomyopathy” Professor Sanjay Sharma

“New CRYteria: International guidelines for the athlete’s ECG” Dr Michael Papadakis

“Exercise recommendations in the athlete with cardiomyopathy/channelopathy” Dr Elijah Behr

“Exercise recommendations in the athlete with hypertension/ischemic heart disease” Professor Mats Börjesson

“Cardiac Risk in the Young; 20 years down the line” Dr Steve Cox

**Cardiomyopathies; a new dawn**

“Obstruction in HCM; Diagnosis, management and treatment options” Dr Antonis Pantazis

“Arrhythmogenic right ventricular cardiomyopathy: Past, present and future” Professor Angeliki Asimaki

“Cardiomyopathy in pregnancy; A growing problem” Dr Sara Thorne
European Conferences

**EuroPRevent Conference**

The first major European Society of Cardiology (ESC) conference of the year for CRY is EuroPRevent, which in 2015 was based in Lisbon, Portugal, and ran from May 14th–16th. EuroPRevent is organised by the ESC’s arm for cardiovascular prevention and rehabilitation (EACPR), the category under which CRY’s research into inherited heart disease and sports cardiology falls. It was a very special conference for CRY Consultant Cardiologist Professor Sanjay Sharma, who was granted the honour of being presented with the first Viviane Conraads Outstanding Achievement Award for Established Researchers at the event in recognition of his incredible contributions to medicine in this field.

**BCS Conference**

The summer conferences continued with the British Cardiovascular Society (BCS) Conference, the UK’s biggest cardiology meeting, from June 8th–10th in Manchester. Amongst the most significant research exhibited was former CRY Fellow Dr Abbas Zaidi’s study tackling the challenges of diagnosing ARVC (arrhythmogenic right ventricular cardiomyopathy) in athletes, a paper published later that month with an editorial in the Journal of the American College of Cardiology, and as described by Professor Sharma: “As of August 2015, the best study in sports cardiology this year.”

**St George’s Training Course**

An international EACPR sports cardiology training course on August 28th at St George’s, University of London – which houses the CRY Centre for Inherited Cardiovascular Conditions and Sports Cardiology – was organised by Professor Sharma and CRY myheart Cardiologist Dr Michael Papadakis in collaboration with St George’s and CRY. The day featured presentations on detecting pathology in athletes by renowned experts including Italian cardiologist Professor Domenico Corrado and Swedish cardiologist Professor Mats Börjesson.

**ESC Congress**

The ESC Congress 2015 in London from 29th August-2nd September was a huge opportunity for Professor Sharma and the CRY Research Group to take advantage of the UK location of the annual conference, and the CRY stand attracted a lot of attention over the five-day event. One of Professor Sharma’s responsibilities as Chair of the ESC’s Sports Cardiology Section was to provide a summary of presentation highlights in the field from EuroPRevent in May – a talk which featured current and former CRY Research Fellows exclusively; Dr Harshil Dhutia, Dr Aneil Malhotra, Dr Ahmed Merghani and Dr Abbas Zaidi.
Interview with Dr Zaidi

Edited excerpts from an interview with Dr Abbas Zaidi, who was a CRY Research Fellow from 2010 to 2013, and had the paper “Clinical Differentiation Between Physiological Remodelling and Arrhythmogenic Right Ventricular Cardiomyopathy in Athletes With Marked Electrocardiographic Repolarization Anomalies” published in the Journal of the American College of Cardiology on 30th June 2015.

What was your study about?
The study looked at the changes that occur in the right side of the heart in athletes. Some athletes may have a condition called arrhythmogenic right ventricular cardiomyopathy (ARVC) that is a cause of sudden death in young people. The condition is characterised by enlargement of the right side of the heart, alongside changes on the ECG known as T-wave inversions. The problem is that healthy athletes can also get enlargement of the right side of the heart and can get T-wave inversions on their ECG, so it’s difficult to know if we’re dealing with a healthy athlete or one with a heart problem that can cause sudden death.

Why is it difficult to tell the difference between normal changes in an athlete and conditions like ARVC?
The problem is that quite a large proportion of healthy athletes develop training-induced enlargement of the right side of the heart. We previously showed this in an earlier CRY paper published in the journal Circulation [Zaidi et al., April 2013]. We found that around a half of all healthy athletes develop right-sided heart enlargement that creates a “diagnostic overlap” with ARVC. That doesn’t mean half of all athletes have ARVC – it just means that regular training makes the heart enlarge in a healthy way. In fact only around 1 in 2,000 people have ARVC. But because athletes develop these training-related changes in their hearts, their heart tracing and scans can look like those of people with the condition.

Interview with Dr Dhutia

Edited excerpts from an interview with Dr Harshil Dhutia, who has been a CRY Research Fellow since 2013, and had the paper “The prevalence and significance of a short QT interval in 18,825 low-risk individuals including athletes” published in the Journal of the American College of Cardiology on 30th June 2015.

What kinds of tests did you use to assess the athletes in your study?
We used comprehensive testing using simple clinical investigations that are available in the majority of cardiology departments in the country. Things like ECGs, echocardiograms, Holter monitors, and treadmill tests. We also used some more advanced investigations like cardiac MRI and signal-averaged ECGs.

What did your results show?
The results showed that even when you have an athlete with a large right-sided heart and T-wave inversions on the ECG, simple tests like the ones mentioned above can help you to work out if you’re dealing with someone with ARVC or whether it’s just healthy training-induced change. For example, scarring on the heart on cardiac MRI scanning should never be seen in healthy athletes. Symptoms like unexplained blackouts should always point towards an underlying heart problem. And Holter monitoring and exercise testing were very useful – if you see for example lots of extra beats coming from the heart, or if the patient develops symptoms during exercise, it points strongly towards an underlying problem.

What are the implications of these results?
The research confirms that simple, widely available investigations can help to differentiate between healthy changes in the heart and a serious heart disorder. But the tests need to be carried out in an expert centre, like the CRY Centre for Inherited Cardiovascular Conditions and Sports Cardiology at St George’s Hospital, London. In addition, the tests need to be interpreted by people who have training and experience in dealing with sports cardiology and inherited cardiac disease in young people. That’s why CRY’s work in training young cardiologists in these issues is so important.

Read the interview at www.c-r-y.org.uk/dr-abbas-zaidi-interview
What was your study about?
The study investigated the prevalence and clinical significance of a short QT interval in young individuals including athletes.

What is short QT syndrome?
Short QT syndrome is a condition that can cause a disruption of the heart’s normal rhythm (arrhythmia). In people with this condition, the heart (cardiac) muscle takes less time than usual to recharge between beats. The term “short QT” refers to a specific pattern of heart activity that is detected with an electrocardiogram (ECG), which is used to measure the electrical activity of the heart. In people with this condition, the part of the heartbeat known as the QT interval is abnormally short.

Why are there debates about the prevalence and significance of a short QT interval?
The definition of a short QT interval by various learned expert consensus panels varies greatly from <390 to ≤320ms. Furthermore, the prevalence of a short QT interval by any criteria definition in young and apparently healthy individuals is not known. Whether short QT intervals pose a risk of sudden cardiac death in apparently healthy adolescent and young adult populations has not been established; however, sudden cardiac arrest in this cohort, particularly in athletes, is highly scrutinized.

What did your results show?
The prevalence of a short QT interval varies widely, from 15.8% (<390ms) to 0.1% (≤320ms), depending on which of four current guideline definitions are applied. Gender and ethnicity influence the QT interval. Males are three times more likely to have a short QT interval (≤320ms) than females, and Afro-Caribbean individuals are five times more likely to have a short QT interval than Caucasians.

Even when QT intervals are ≤320ms, they are associated with an excellent medium-term prognosis. This questions the value of a short QT interval as a marker of short QT syndrome in adolescents and young adults in the absence of symptoms or of a family history of sudden arrhythmic death syndrome. Applying the 2013 Seattle criteria for ECG interpretation results in a 50-fold lower rate (8.62% vs 0.17%) of athletes with suspected short QT syndrome compared to the 2010 European Society of Cardiology recommendations.

What are the implications of these results?
A short QT interval in isolation has poor medium-term diagnostic and prognostic value, and should not be solely relied on to diagnose short QT syndrome in young individuals. The European Society of Cardiology definition for a short QT interval in athletes (<380ms) is associated with a high false positive rate. The Seattle criteria definition (≤320ms) is more pragmatic for ECG interpretation guidelines in athletes.

Read the interview at www.c-r-y.org.uk/dr-harshil-dhutia-interview
Papers / Articles

- Sharma, S., Papadakis, M. “Interpreting the Athlete’s EKG: Are All Repolarization Anomalies Created Equal?” Circulation, January 2015.
- Gati, S., Sharma, S. “The dilemmas in diagnosing left ventricular non-compaction in athletes.” European Heart Journal: CardioPulse, April 2015.
- Sharma, S., Merghani, A., Mont, L. “Exercise and the heart: the good, the bad, and the ugly.” European Heart Journal, June 2015.
- Zaidi, A., Sharma, S. “Arhythmic right ventricular remodelling in endurance athletes: Pandora’s box or Achilles’ heel?” European Heart Journal, August 2015.

Posters


For a full list of research visit www.c-r-y.org.uk/research/cry/>
Professor Sanjay Sharma, CRY Consultant Cardiologist

The CRY Research Programme is overseen by Professor Sanjay Sharma. Sanjay is Professor of Inherited Cardiovascular Disease and Sports Cardiology at St George’s Hospital, London; Virgin Money London Marathon Medical Director; and was the London 2012 Olympic Cardiologist.

CRY’s Research Fellowship Programme funds doctors for up to three years who choose to specialise in the fields of inherited cardiac diseases, sudden cardiac death, screening and sports cardiology. As of January 2016 there are eight CRY Research Fellows and two visiting Honorary Fellows at St George’s Hospital who divide their time between NHS clinics, CRY screenings and research. As well as the Fellows CRY are currently funding, 17 former Fellows have been trained as specialists by CRY and are now working in the NHS throughout the UK.

Professor Sharma oversees the CRY National Screening Programme in which the CRY Research Fellows play a central role. Every person that CRY tests within the programme is asked to consent to having their data used anonymously for research purposes. This has developed a symbiotic relationship between research and screening where we are able to identify young individuals at risk whilst learning from our experience and publishing these findings.

CRY’s Research Programme

CRY’s research is focussed on cardiac pathology and screening in both the general population and elite athletes. It is our unique expertise in sports cardiology and how athleticism, ethnicity and gender affect the ECG that enables us to conduct screening on a general population level. To test young people you need to be able to interpret the results of every young person. It is CRY’s expertise in sports cardiology that gives us the authority to take forward screening in the UK. Thousands of young people aged 14–35 have been evaluated, resulting in many “firsts” in the scientific literature.

CRY’s Research Group has taken the lead in the UK in identifying the prevalence of cardiac conditions in young people in the general population. Our findings are that screening young, apparently healthy individuals will identify minor cardiac abnormalities in around 1% of people and potentially life-threatening disorders in around 0.3% – one in 300 young people. The false positive rate for CRY’s screening programme is just over 3% – lower than any comparable screening programme.

One of the most important papers in recent years informs the statistic that every week in the UK at least 12 apparently fit and healthy young people die of undiagnosed heart conditions is: Papadakis, M., Sharma, S., Cox, S., Sheppard, M.N., Panoulas, V.F. and Behr, E.R. “The magnitude of sudden cardiac death in the young: A death certificate-based review in England and Wales.” Europace. 2009, Vol.11, No.10, p1353–1358.

CRY’s future research aims include increasing our knowledge of the prevalence and characteristics of inherited heart disorders which can cause YSCD; enhancing the accuracy of diagnostic tools for identifying these conditions; and improving the feasibility of a cardiac screening programme available through the NHS for young people nationwide.

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