

Athletes Heart Sudden Death Hypertrophic Cardiomyopathy

Stanford University

Not enough studies on genetics and exercise. If you want to be a great athlete, you must pick your parents by getting their times from the NFL compine.

Go to the Stanford website <http://hcm.stanfordhospital.org>
For the program.

Saturday, June 27, 2009

7:00 Breakfast

7:00 Poster Setup

8:00 – Euan Ashley, Welcome & Introduction to Keynote Speaker

8:05 – Barry J. Maron, Measuring and managing risk in HCM

HCM is a global disease, 50 countries and all continents. We have data that shows this condition is prevalent in 1:500 of the general population. That means that over 600,000 people have this condition but most do not know they have it.

Heterogeneity – it expresses in varying segments of the left ventricle. It is possible to have a normal echo and not visualize the wall thickening. Then the MRI shows more specialized exposure of the disease.

Survival with HCM in an unselected cohort of adults – the risk of living is the same as the national standard.

Age distribution in 274 unselected HCM patients. Chart shows that many people live long lives with the disease.

Profiles in prognosis for HCM some have to manage HCM with risk of SD, Afib, other life style issues.

Risk of SCD is very high under the age of 25. The most common cause of death in young athletes, 36% from HCM. Hence, the Bethesda recommendation is that athletes with diagnosis of HCM should be excused from competitive sports.

Strongest risk factors have been built to determine the highest at risk that would need an ICD. MRI is used to also consider possible high event rate.

ICD implantation on various studies. Risk factors (1, 2 or 3) to consider, although with 1 risk factor (unexplained syncope) the important factor for implants.

Virtually always the ICD will work with pts with HCM.

Obstruction seems to be the trigger for heart failure over long periods of time. Case for septal myectomy: the gold standard. In Mayo Clinic study, the myectomy pts showed an increase level of longevity equal to that of the general population (81%) where as, non-myectomy HCM pts have a lower level of longevity (65%)

Septal wall ablations is newer and has less data to determine the benefits.

Clinical pathways of prognosis in HCM shows there are treatment plans and pathways for all pts.

Questions: is it fair to offer a pt w/alcohol ablation an ICD?

Answer: We have to consider these issues on a case by case basis. We can't make a proclamation from the podium for a standard for ICD implantation.

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Question: If you had to say, what are the three most important things to work on this condition over the next five years?

Answer: We would like to find with stratification the ability to offer ICD implantation to the best pts. Like to see more agreement with less emotion the importance of myectomy.

Questions: Sometimes pts tell you what you want to hear. How do you handle that?

Answer: I ask the pt how unhappy are they with their symptoms. Do they want to change these symptoms with a major procedure? The interview of the pt must be very probing and intense to uncover the difference of "normal" for HCM pts to others without HCM.

Question: After a myectomy do you do an implantation of ICD with a pt with one risk factor?

Answer: It depends on the pt and how that compares.

MODERN MANAGEMENT OF HCM – Moderator: Euan Ashley

8:30 – Sharlene Day, Medical therapy & exercise guidelines

The purpose of medical therapy – cure disease (not yet) so we can prolong life & prevent disease progression.

Improve on diastolic dysfunction.

New therapies to reduce hypertrophy and fibrosis. No real data yet, but we are hopeful for the future.

Current target is on therapies for symptom relief. New treatments for prevention of progression of the disease.

What about exercise? These are expert opinion, and guidelines are there for everyone. 36th Bethesda conference determinations for athletes.

Use the charts for determination of the levels of exercises to avoid. Due to benefits of exercise to prevention of SD, it is a good idea for people with HCM to have some habitual exercise in their regiment.

Questions: on exercise & HCM pts, then with AEDs in homes for children that may not have or be ready for ICD implantation.

BREAK – Met Dr. Joseph Marek by the coffee and talked about the screening campaign this fall and the need to join our voices. Contact him after we return from the conference to employ the campaign. We discussed my connection with Dr. Pelliccia – Dr. Marek would like to meet him.

8:45 – Bruce Reitz, Modern myectomy

History of the surgery. The definition of the technique.

Review of the data related to the surgeries and successes.

9:00 – David Lee, Best practice in alcohol septal ablation

World view of Alcohol Ablation – there is a need for more data and studies.

There is not treatment for everything within HCM. About 2/3 of the pts will have septal issues that are targets for this procedure. HCM pts have to have symptoms, have obstruction, high surgical risk (myectomy) to determine the best candidates.

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Review the data of the procedures over the past decade (1996 – 2002) from Baylor.

Overview of pt expectations post alcohol ablation.

Question: What do you do for mitral valve repair in ASA procedures?

Answer: That is where better selection of pts for ASE is necessary. Not able to repair mitral valve during this procedure.

9:15 – Paul Wang, Medical and interventional management of atrial fibrillation in HCM.

Discuss AF and treatment options.

Past study from Italy and MN and how this could be reviewed.

Also shows that people with AF that turns up <50 year olds have higher propensity to SD & Stroke.

There are future procedures that will improve the mortality, reduce stroke and offer better options to improve quality of life.

9:30 POSTER SESSION. Break

Lisa Salberg, discussed education forum for family doctors, Coaches & Athletic Trainers on the prevention of Sudden Death in Athletes.

Dr. Chris Davis, Rady Children's Hospital in San Diego. Would like to do more screenings in the area!

SCREENING ATHLETES, Moderator: Victor Froelicher

10:30 – Barry Maron, Screening in the United States

The way things are here in the United States. The idea of Sudden Death in young athletes is emotional and tragic. They used to be private, but now they are in the public, in the papers, television and typically these young people are well known. In a way this helps us.

Sudden Death in Young – chart shows that 70% of the deaths are from Cardiac issues.

US Sudden Death in young competitive athletes seems to be around 75 or so per year. Based on the methodology used here. We don't have a national registry and consistent base of collection of these deaths.

Frequency estimates for Sudden Cardiac Death in Athletes show 1:200,000 for high school/college. This is a low event process.

Differences in the two systems in the US and Italy. Size of countries, number of athletes (12 million to 6 million or less). US does have a screening, through preparticipation physicals. Legal differences (civil/criminal in Italy; about 2000 elite/professional athletes) where as in the US no precedence.

Corrado study – the implication of the study as non-truth. Study is only of the Veneto region of Italy. Comparison of CD Mortality in Young with MN to Veneto.

MN rate 00 – 06 seems to have raised rate of death, is that due to exposure, media attention, increased awareness?

Question: Where did you get your numbers?

Answer: from the premium payout of the MN high school students; all deaths occurring in athletes; databases & media.

Obstacles to the National Mandatory Italian screening Initiative in the US.

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Impracticable debate?

10:45 – Antonio Pelliccia, The Italian experience

Law in Italy – 1982 “citizens of all ages must pass preventive medical evaluation to assess their eligibility for competitive sport participation”

The law protects all kids.

Comparative costs of the screening tests - \$40 but the athletes have to pay for this cost.

Echo is a little more but only for those that are needed to be tested.

Then the athletes becomes the patient.

Other comparisons of costs per country, for Italy to China. The vast majority of the athletes pay their own testing.

The main target of the screening (HCM) have electro cardio-graphic changes that can be found with EKG. This would be useful for the screening as some with normal echos have abnormal EKG and could be at risk of SD during sports.

The Corrado data came from the autopsy reports / study that was used in the Veneto region.

Questions: The athletes that get turned down to play, do they leave the country and play in other areas? Do you have numbers?

Answers: We don't have that happen very often.

Questions: Is there are high percent of ARVD?

Answer: No, HCM is more widely found than ARVD.

11:00 – Jonathan Drezner, A primary care perspective

How certain am I to tell a parent their child is safe to play? Is this a Global debate or Common Ground?

A few givens – warning symptoms for SCA in children should not be missed; preparticipation evaluations should be guided by a comprehensive personal and family history questionnaire; a longitudinal CV risk assessment is necessary.

Warning symptom & Family history in Children with SCA. PHW family; 19:79 cases (24%) reported syncope and/or unexplained seizure activity that went undetected as CV disease; Syncope 14:79 cases (39%)

Prevention of SCD – AEDs save lives, but ECG could add to the saving of kids

Current US Strategy: no data behind the PPE

Can we do better – ECG screening – Yes or No: Incidence & false positive rate

SCD Incidence – difficult to compare incidence studies with profoundly different methodology; what is the numerator / what is the denominator

Traditional numbers 125 athletes per year

Sudden Death in young recruits (1:9,000)

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Prospective population based study by Atkins – 3.75/10000 (1 in 27,000)

Hard time tying this together to the athletes risk

Italian study SCD Incidence

Van Camp & Maron already show a low rate, but Eckart & Atkins show a high rate as at the beginning of the Italian

Incidence or Prevalence – are we screening for incidence or prevalence

The purpose of screening is the detection of 'risk' of SD

We already screen our kids for less risks why not CV (1:350)

ECG Interpretation – A call for uniform terminology

Italian screening program – Current ECG screening programs that have ECG screenings, but not done uniformly. These programs do need attention and demand for uniform screening approach.

Dr Marek positive rate is only 2%.

What is the most appropriate recommendation for screenings? Where colonoscopy & other screenings are not laws. We need to have recommendations that are uniform and available.

With the greatest impact and less false positive rate we should screen the high school athletes.

Move beyond the debate and improve our approach.

Question: Was the incident of seizures for the kids relative?

Answer: They were more likely sent to neurology and no cardiac review was done.

Question: How did the military autopsy turn out?

Answer:

Question: As a primary care doctor – should we do ECG on kids that faint / syncope?

Answer: Yes, these kids deserve at least one ECG.

11:15 – Vy-van Le, The Stanford experience

SD in athletes is a rare possibly preventative event.

ECG may still detect subtle findings that could detect CV issues.

Protocol – 2007 ECG & 2008 ECG & Echo.

Observational study; study protocol was approved by the Stanford Hospital

Approximately 700 athletes, but about 300 athletes would be coming to the screening annually.

Web based questionnaire; and involvement in the study

Volunteer based volunteers; Schilling machines and software was used to transfer all measurements into a database; interpretation was over-read by senior cardiologists

Volunteer echo screening (Dr. Lang) with the assistance of Philips

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Participation rate > 95% accepted to participate in the ECG screening; about 100 added to the Echo portion.

35% of the population screened with ECG need further evaluation; not all the groups would do follow up.

Conclusion – volunteer base was not optimal for sustainable program; best related to college programs.

11:30 – Matthew Wheeler, Cost effectiveness of screening strategies to prevent SCD

Challenges of screening: scope of problem; efficacy of early intervention; reliability; cost considerations; adoption and implementation

Screening athletes for risk of SCD – even high risk groups for SCD in this otherwise young population will have event rates – 1%/athlete per year

Mixing of intermediate and high risk groups distinguished by screening test from low risk may impair overall

Cost affective ratio = Cost of intervention / years of life saved

Costs: includes any & all intervention costs

Includes costs of subsequent testing & treatment including baseling medical costs

Discounted to value near-term

Model decision algorithm based on no-screening and screening arm based on questions

Inputs: costs; effectiveness; test parameters

Risk rate assumptions

Validated of model

Base Case – costs about \$200,000 to save a life; but with ECG screening it drops to \$76,000 for life saved.

Alternate screening method chart – cost per screening (\$82,000 life saved per year)

Limitations – annual screening is unlikely to be cost effective; screening all school children is unlikely to be cost effective

Significant uncertainty regarding magnitude of risk reduction; test specificity critical to cost-effective screen

Uncertainty over reproducibility and value of repeated testing

Magnitude of effect for exercise benefit may be significantly larger per capital than screening effect

11:45 – Paul Thompson, Balancing the evidence: the challenge of guidelines

There is very little careful scientific evidence to advise athletes

Even the best studies are observational only

Making it difficult to evaluate unexpected outcomes

Guidelines must be conservative as they are not used by experts.

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Guidelines often dictate policy, not individual practice

Guidelines do not consider individual patient preferences

Cost of screening is in the additional testing

The death rate in pre-participation is low; how can this be considered.

LUNCH

Philips connection with Stanford is HUGE. Former Agilent, Hewlett/Packart, now Philips. With the Lucile Packart Children's Hospital connected to Stanford Hospital.

CASE & CONTROVERSIES, Moderator: Mark Estes

13:00 – Victor Froelicher/Antonio Pelliccia/Paul Thompson, Difficult screening ECGs

21 yo Stanford mail football player (6'5", 255 lbs) screening echo showing 13mm septal

AHA has new criteria for normal vs normal in athletes

In the absence of data we have controversy

13:15 – David Lee/Bruce Reitz/Euan Ashley, Mid-cavity obstruction

13:30 – Antonio Pelliccia/Sharlene Day/Gary Balady, Athlete's Heart, Hypertension and HCM

14:00 – Paul Wang/Mark Estes, Channelopathy

14:15 – Carolyn Ho/Colleen Brown/Matthew Wheeler, Applied family genetics

14:30 – Paul Wang/Jonathan Drezner, I have an ICD, can I play now?

14:45 – Closing remarks, Euan Ashley