EXERCISE AND SPORTS for PATIENTS with SADS CONDITIONS

Rachel Lampert, MD
Yale University School of Medicine
September 30, 2016
How much exercise can I do? How hard? What type?
How do I get started exercising more?

<table>
<thead>
<tr>
<th>Competitive</th>
<th>Recreational</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="image1" alt="Basketball" /> <img src="image2" alt="Track" /> <img src="image3" alt="Swimming" /></td>
<td><img src="image4" alt="Swimming" /> <img src="image5" alt="Cycling" /> <img src="image6" alt="Walking" /></td>
</tr>
</tbody>
</table>
Asymptomatic patients with baseline QT prolongation (QTc of 470 ms or more in males, 480 ms or more in females) should be restricted to class IA sports. The restriction limiting participation to class IA activities may be liberalized for the asymptomatic patient with genetically proven type 3 LQTS (LQT3).

36th Bethesda Conference: Eligibility recommendations for competitive athletes with cardiovascular abnormalities
Maron, Zipes, et al, JACC 2005
# Classification of Sports

**Figure 2.** Classification of sports. This classification is based on peak static and dynamic components achieved during exercise. It is noted, however, that higher values may be reached during training. The increasing dynamic component is defined in terms of the oxygen uptake (MaxO₂) achieved and results in an increasing cardiac output. The increasing static component is related to the voluntary contraction (MVC) reached and results in an increasing blood pressure load. The lowest total cardiovascular demands (e.g., low blood pressure) are shown in green and the highest in red. Blue, yellow, and orange depict low moderate, moderate, and high demands. *Danger of bodily collision. †Increased risk if syncope occurs.*

<table>
<thead>
<tr>
<th>Increasing Dynamic Component</th>
<th>Increasing Static Component</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Low (&lt;40% Max O₂)</td>
<td>I. Low (&lt;20% MVC)</td>
</tr>
<tr>
<td>B. Moderate (40-70% Max O₂)</td>
<td>II. Moderate (20-50% MVC)</td>
</tr>
<tr>
<td>C. High (&gt;70% Max O₂)</td>
<td>III. High (&gt;50% MVC)</td>
</tr>
</tbody>
</table>

- **Bobsledding/Luge:**
- **Field events (throwing):**
- **Gymnastics:**
- **Martial arts:**
- **Sailing:**
- **Sport climbing:**
- **Water skiing:**
- **Weight lifting:**
- **Windsurfing:**

- **Body building:**
- **Downhill skiing:**
- **Skateboarding:**
- **Snowboarding:**
- **Wrestling:**

- **Boxing:**
- **Canoeing/Kayaking:**
- **Cycling:**
- **Decathlon:**
- **Rowing:**
- **Speed-skating:**
- **Triathlon:**

- **American football:**
- **Field events (jumping):**
- **Figure skating:**
- **Rodeoing:**
- **Rugby:**
- **Running (sprint):**
- **Surfing:**
- **Synchronized swimming:**

- **Basketball:**
- **Ice hockey:**
- **Cross-country skiing:**
- **Skating technique:**
- **Lacrosse:**
- **Running (middle distance):**
- **Swimming:**
- **Team handball:**

- **Billiards:**
- **Bowling:**
- **Cricket:**
- **Curling:**
- **Golf:**
- **Rifflery:**

- **Baseball/Softball:**
- **Fencing:**
- **Table tennis:**
- **Volleyball:**

- **Badminton:**
- **Cross-country skiing (classic technique):**
- **Field hockey:**
- **Orienteering:**
- **Race walking:**
- **Racquetball/Squash:**
- **Running (long distance):**
- **Soccer:**
- **Tennis:**
Don’t forget the benefit side of the equation for participation in organized sports.
Adolescents with Implantable Cardioverter Defibrillators: A Patient and Parent Perspective

BELINDA RAHMAN, M.Sc.,* IVAN MACCIOLCCA, M.Sc.,† MARGARET SAHHRAR, M.S.W.,*,†
SULEMAN KAMBERI, B.App.Sc., CDSPECAP,‡,¶ VANESSA CONNELL, C.N.S.,‡
and RONY E. DUNCAN, Ph.D.*,§

“I mean when I was younger and I was told that I couldn’t do sport that really affected me because I was really into sport and you know every kid runs around and plays ‘tiggy and that, so I was like bawling my eyes out when he told me and I hated the doctor” (Matilda, 15, HCM)

“I mean when I was young it probably affected me a bit more because I was sort of like not normal, you know? Most kids could do other stuff and I couldn’t” (Matilda, 15, HCM)

“…you know you see her cry and just say to me, ‘My life is over because this is one thing I like doing and it’s got taken away from me’, and I just felt like I wanted her to play sport because it was something she enjoyed doing” (Maria)
Looked at records all patients seen at Mayo 2000-2010

Approach at Mayo: Give the athlete and their family enough information about prognosis and the guidelines to make an informed decision
Who were these patients?

353 LQTS 1-3

130 chose to continue sports: 67 genotype positive but normal ECG
60 LQTS on ECG
3 playing golf

All were treated with Beta-blockers, left cardiac sympathetic denervation,
And/or ICD (20 had ICDs) as per their doctor

JUST one LQTS-athlete with arrhythmia during sports—9 year old, had ICD after arrest, admitted not taking beta-blockers
103 LQTS patients doing sports
26 competitive
77 recreational

Age 4-21 years
All on beta-blockers

NO arrhythmias during sports
Safety of Sports for Athletes With Implantable Cardioverter-Defibrillators: Results of a Prospective, Multinational Registry

Rachel Lampert, Brian Olshansky, Hein Heidbuchel, Christine Lawless, Elizabeth Saarel, Michael Ackerman, Hugh Calkins, N.A. Mark Estes, Mark S. Link, Barry J. Maron, Frank Marcus, Melvin Scheinman, Bruce L. Wilkoff, Douglas P. Zipes, Charles I. Berul, Alan Cheng, Ian Law, Michele Loomis, Cheryl Barth, Cynthia Brandt, James Dziura, Fangyong Li and David Cannom

Circulation. 2013;127:2021-2030
Who was in the study?

372 people (later up to 440)
Half had prior cardiac arrest or VT, ages 10-60
Sports Participation

All athletes

- Soccer: 67
- Skiing/snowboarding: 83
- Cycling: 106
- Basketball: 38
- Baseball/softball: 56
- Volleyball: 52
- Triathlon: 27
- Running/marathon: 24
- Running: 19
- Baseball/softball: 23
- Basketball: 17
- Volleyball: 11
- Lacrosse: 4
- Football: 2

Competitive Subgroup: Varsity/Junior Varsity/Traveling Teams; n=60
More individuals received shocks during either sports or physical activity vs rest.

No difference sports vs physical activity.
Results: Primary Endpoints

Death or cardiac arrest during or after sports: 0
Injury due to arrhythmia or shock during sports: 0
Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Preamble, Principles, and General Considerations

AHA/ACC Scientific Statement

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 10: The Cardiac Channelopathies

A Scientific Statement From the American Heart Association and American College of Cardiology

Michael J. Ackerman, MD, PhD, FACC, Chair

Douglas P. Zipes, MD, FAHA, MACC*
Richard J. Kovacs, MD, FAHA, FACC*
Barry J. Maron, MD, FACC*
Sports MAY BE CONSIDERED for the athlete with Previously symptomatic LQTS or LQTS on ECG

ADEQUATELY TREATED

beta-blockers

Sympathetic denervation

ICD
APPROPRIATE PRECAUTIONS
Avoiding LQTS Drugs

Stay Hydrated

Avoid hyperthermia from fever or heat stroke
Personal AED (Automated External Defibrillator)

Establish emergency action plan with school or team
What about CPVT??

For an athlete with previously symptomatic CPVT or an asymptomatic CPVT athlete with exercise-induced premature ventricular contractions in bigeminy, couplets, or nonsustained ventricular tachycardia, participation in competitive sports is not recommended except for class IA sports (Class III; Level of Evidence C). Exceptions to this limitation should be made only after consultation with a CPVT specialist.

Reasons: role of adrenalin in causing the arrhythmia
ICDs do not always work
### Table 5. Events/Individuals Requiring >1 Shock for Termination to Sinus Rhythm

<table>
<thead>
<tr>
<th>Sex</th>
<th>Age, y</th>
<th>Cardiac Diagnosis</th>
<th>Primary Sport</th>
<th>Activity</th>
<th>Activity Type</th>
<th>Shocks, n</th>
</tr>
</thead>
<tbody>
<tr>
<td>M</td>
<td>28</td>
<td>Idiopathic VF</td>
<td>Ultimate Frisbee</td>
<td>Ultimate Frisbee</td>
<td>Competition</td>
<td>5</td>
</tr>
<tr>
<td>F</td>
<td>47</td>
<td>Idiopathic VF</td>
<td>Cycling</td>
<td>Cycling</td>
<td>Practice</td>
<td>4</td>
</tr>
<tr>
<td>M</td>
<td>44</td>
<td>CAD</td>
<td>Running</td>
<td>Running</td>
<td>Practice</td>
<td>2</td>
</tr>
<tr>
<td>M</td>
<td>50</td>
<td>CAD</td>
<td>Cycling</td>
<td>Cycling</td>
<td>Practice</td>
<td>6</td>
</tr>
<tr>
<td>M</td>
<td>57</td>
<td>CAD</td>
<td>Tennis, basketball</td>
<td>Walking</td>
<td>Physical Activity</td>
<td>6</td>
</tr>
<tr>
<td>F</td>
<td>16</td>
<td>CPVT</td>
<td>Lacrosse field hockey</td>
<td>Running</td>
<td>Post-physical activity</td>
<td>3</td>
</tr>
<tr>
<td>M</td>
<td>15</td>
<td>HCM</td>
<td>Baseball</td>
<td>Socializing</td>
<td>Other</td>
<td>2</td>
</tr>
</tbody>
</table>

CAD indicates coronary artery disease; CPVT, catecholaminergic polymorphic ventricular tachycardia; HCM, hypertrophic cardiomyopathy; and VF, ventricular fibrillation.
63 patients with CPVT
24 had been athletes, 21 decided to continue to compete

During follow-up
Athletes: 3 arrhythmic events in 3 of 21 athletes (14%)
Non-athletes: 7 arrhythmic events in 6 of 42 non-athletes (14%)
(no deaths in either group)
Editorialists cautiously optimistic
WHAT ABOUT BRUGADA??

Sports MAY BE CONSIDERED for the athlete with Previously symptomatic BRUGADA ADEQUATELY TREATED

NO symptoms for three months
APPROPRIATE PRECAUTIONS

Avoiding BRUGADA Drugs

Stay Hydrated

Avoid hyperthermia from fever or heat stroke
Personal AED (Automated External Defibrillator)

Establish emergency action plan with school or team
Kids, don’t try this at home

Work closely with your LQTS/SADS doctor to make sure you are exercising safely

* Just a few examples
LifeStyle and Exercise In Genetic Cardiovascular Disease

Aka

LIVE-HCM/ LIVE-LQTS

NIH R01 HL125918-01, PIs Lampert, Ackerman, Day
PURPOSE

The goal of LIVE-HCM/LIVE LQTS is to determine how lifestyle and exercise impact the well-being, physical and emotional, of people with hypertrophic cardiomyopathy and long QT.
WHO CAN ENROLL?

People with HCM OR Long QT Syndrome OR
The gene for HCM or LQTS and a family member who has it
Age 8-50 (60) years
With OR without an ICD
Any level exercise—whether you like to run, walk, or sit and read a book.
This study does not endorse competitive sports for LQTS patients

Rather, we want to obtain information on the risks and benefits of exercise at all levels, by following individuals over time at whatever level of exercise they are currently engaged in.
Can enroll directly through coordinating center
All questionnaires, interviews over phone and internet
→ No geographic constraints to participation (US, Canada, England, Australia)
WHAT DOES PARTICIPATION INVOLVE?

MINIMAL TIME COMMITMENT

No testing, no bloodwork, no intervention
Medical Records will be obtained at enrollment,
   ECHO read by Core Lab (Mayo Clinic)

At the time of enrollment:

Questionnaires over the internet (adults) or by phone
   (children under 18)
   Activities
   Quality of life

20-40 minutes, ok to save and come back
Fitbit ZIP

To wear minimum 2 weeks at enrollment 1 week every three months OR as much as you’d like!

Access to your data on the website

Optional wristband accessory

OK to wear your own if you already have one
Three years participation:

Brief internet questionnaire every six months (adults or parents)

(Reminders and a link sent)
STUDY PROGRESS SO FAR:

OVER 700 HCM participants
OVER 800 LQTS participants
ENROLLED
GOAL IS 2000 each

Here’s What Participants Have to Say:

“This study captures the biggest question in my life right now.”
– 32 year old LQTS patient

“This study is helping empower my kids to take control of their diagnosis.”
– Mother of two adolescents with HCM

“I’m so excited to start the study.”
– Pretty much everyone

“I’m having fun with the questions [QOL interview] but I’m really just doing it to get the Fitbit.”
– 8 year old LQTS patient
HOW DO I ENROLL?

Contact the Central Coordinating Center
Yale School of Medicine
866-207-9813
Email live.hcm@yale.edu/live.lqts@yale.edu

Speak with Study Coordinators
Theresa
Cheryl
Sherry
How do I get started exercising?

Talk to your doctor!
COMPETITIVE SPORTS FOR THE ICD PATIENT

“Athletes with conditions that result in cardiac arrest…generally are treated with an ICD and cannot participate in any moderate- or high-intensity competitive sports.

However, athletes with ICDs and who have had no [arrhythmias] for six months may engage in class IA competitive sports.”

36th Bethesda Conference: Eligibility recommendations for competitive athletes with cardiovascular abnormalities
Maron, Zipes, et al, JACC 2005
Postulated risks:

Increased likelihood of ventricular arrhythmias

Inability of ICD to terminate ventricular arrhythmias during sports

Risk of injury due to loss of consciousness due to arrhythmia or shock itself

Damage to ICD system, leads or generator
Safety of Sports Participation in Patients with ICDs: A Survey of Heart Rhythm Society Members

Lead Malfunctions, Definite or Probable

Long-term Fu data:
5 year lead survival 94%
10 year lead survival 85%
Did ICD Shocks Affect Sports Participation?

37 received ICD shocks during sports
- 4 stopped sports completely
- 7 stopped one or some sports

Five patients stopped at least one sport due to shocks received at other times
Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 9: Arrhythmias and Conduction Defects

A Scientific Statement From the American Heart Association and American College of Cardiology

Recommendations

1. ICD indications for competitive athletes should not differ from those applicable to the general population with appropriate diagnoses and clinical profiles (Class I; Level of Evidence C).

2. Recommendations should be based on existing evidence for benefit and risk and should include discussions of potential impact on sport-specific participation and performance (Class I; Level of Evidence C).

3. Participation in sports classified as IA for athletes with an ICD is reasonable if they are free of episodes of ventricular flutter or ventricular fibrillation requiring device therapy for 3 months (Class IIa; Level of Evidence C).

4. Participation in sports with higher peak static and dynamic components than class IA may be considered if the athlete is free of episodes of ventricular flutter or ventricular fibrillation requiring device therapy for 3 months. The decision regarding athletic participation should be made with consideration of, and counseling of, the athlete regarding the higher likelihood of appropriate and inappropriate shocks and the potential for device-related trauma in high-impact sports (Class IIb; Level of Evidence C).

5. The desire of the athlete to continue athletic competition should not represent the primary indication for use of an ICD (Class III; Level of Evidence C).
Athletes with an ICD should be able to participate in sports every minute of every day in every possible circumstance.
### Table 1: Classification of Sports According to Contact

<table>
<thead>
<tr>
<th>Contact</th>
<th>Limited-Contact</th>
<th>Noncontact</th>
</tr>
</thead>
<tbody>
<tr>
<td>Basketball</td>
<td>Adventure racing</td>
<td>Badminton</td>
</tr>
<tr>
<td>Boxing</td>
<td>Baseball</td>
<td>Bodybuilding</td>
</tr>
<tr>
<td>Cheerleading</td>
<td>Bicycling</td>
<td>Bowling</td>
</tr>
<tr>
<td>Diving</td>
<td>Canoeing or kayaking</td>
<td>Canoeing or kayaking (flat water)</td>
</tr>
<tr>
<td>Extreme sports</td>
<td>Fencing</td>
<td>Crew or rowing</td>
</tr>
<tr>
<td>Field hockey</td>
<td>Field events</td>
<td>Curling</td>
</tr>
<tr>
<td>Football, tackle</td>
<td>High jump</td>
<td>Dance</td>
</tr>
<tr>
<td>Gymnastics</td>
<td>Pole vault</td>
<td>Field events</td>
</tr>
<tr>
<td>Ice hockey</td>
<td>Floor hockey</td>
<td>Discus</td>
</tr>
<tr>
<td>Lacrosse</td>
<td>Football, flag or touch</td>
<td>Javelin</td>
</tr>
<tr>
<td>Martial arts</td>
<td>Handball</td>
<td>Shot-put</td>
</tr>
<tr>
<td>Rodeo</td>
<td>Horseback riding</td>
<td>Golf</td>
</tr>
<tr>
<td>Rugby</td>
<td>Martial arts</td>
<td>Orienteering</td>
</tr>
<tr>
<td>Skiing, downhill</td>
<td>Racquetball</td>
<td>Power lifting</td>
</tr>
<tr>
<td>Ski jumping</td>
<td>Skating</td>
<td>Race walking</td>
</tr>
<tr>
<td>Snowboarding</td>
<td>Ice</td>
<td>Riflery</td>
</tr>
<tr>
<td>Soccer</td>
<td>In-line</td>
<td>Rope jumping</td>
</tr>
<tr>
<td>Team handball</td>
<td>Roller</td>
<td>Running</td>
</tr>
<tr>
<td>Ultimate Frisbee</td>
<td>Skiing</td>
<td>Sailing</td>
</tr>
<tr>
<td>Water polo</td>
<td>Cross-country</td>
<td>Scuba diving</td>
</tr>
<tr>
<td>Wrestling</td>
<td>Water</td>
<td>Swimming</td>
</tr>
<tr>
<td>Skateboarding</td>
<td>Softball</td>
<td>Table tennis</td>
</tr>
<tr>
<td>Squash</td>
<td>Weight lifting</td>
<td>Tennis</td>
</tr>
<tr>
<td>Volleyball</td>
<td>Weight lifting</td>
<td>Track</td>
</tr>
</tbody>
</table>

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**American Academy of Pediatrics**

From Rice, Pediatrics 2008
CAVEATS:
Few aggressive contact sports

? Lead survival
ALL ICDs: ICDs in the ICD Sports Safety Registry

0% subcutaneous

100% transvenous

Theoretic advantages: No friction between clavicle and first rib (swimming, rowing)

Theoretic disadvantages: Lead outside thorax, against ribs (ball sports, contact)
SPECIFIC GENETIC HEART DISEASES

1) Long QT Syndrome
2) Brugada Syndrome
3) Hypertrophic Cardiomyopathy (HCM or “HOCM”)
4) Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC or ARVD)
CAVEAT

Cardiomyopathy and Progression of Underlying Disease: Not addressed by the ICD Sports Safety Registry

Earlier onset in endurance athletes
More heart failure in endurance athletes
Higher risk arrhythmias in endurance athletes
Association of competitive and recreational sport participation with cardiac events in patients with arrhythmogenic right ventricular cardiomyopathy: results from the North American multidisciplinary study of arrhythmogenic right ventricular cardiomyopathy

Anne-Christine Ruwadl, Frank Marcus, N.A. Mark Estes III, Mark Link, Scott McNitt, Bronislava Polonsky, Hugh Calkins, Jeffrey A. Towbin, Arthur J. Moss, and Wojciech Zareba

**Special Report**

Treatment of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia
An International Task Force Consensus Statement

Domenico Corrado, MD, PhD,
Francis E. Marchlinski, MD,
Corinne Bronchi, MD,
Christian Schmied, MD,
NA Mark Estes

**Recommendations**

- It is recommended that patients with a definite diagnosis of ARVD/C not participate in competitive and/or endurance sports (Class I).
- Patients with a definite diagnosis of ARVD/C should be restricted from participation in athletic activities, with the possible exception of recreational low-intensity sports (Class IIa).
- Restriction from competitive sports activity may be considered in ARVD/C family members with a negative phenotype, either healthy gene carriers (class IIa) or with unknown genotype (class IIb).

**HOW MUCH CAN I DO??**
Competitive versus recreational
Hours per year—
"highest quartile” >500 hours/year
What is the risk associated with sports participation in HCM patients?

1) Arrhythmias/risk of sudden death:  
For the person with an ICD, the ICD works  
For the person without an ICD, risk is uncertain

2) Progression of disease:  
No data that increases  
Some data in animal model that decreases
GUIDELINES STRATIFY ADVISABLE MODES OF RECREATIONAL EXERCISE FOR PATIENTS WITH HCM ACCORDING TO INTENSITY

There are no data to guide recommendations

**Not Advised (0-1):** Basketball, Running, Soccer, Bodybuilding, Ice Hockey, Racquetball, Skiing, Singles Tennis, Weightlifting, Baseball/Softball, Football, *Rock climbing, *Scuba diving

**Intermediate Risk (2-3) – Assess individually:** Jogging, Hiking, Sailing, Horseback riding, Motorcycling

**Probably Permitted (4-5):** Swimming, Biking, Bowling, Golf, Skating, Snorkeling, Brisk walking, Treadmill/Stationary Bike, Doubles Tennis

**Anaerobic, burst Isometric**
* Risk for trauma related to impaired consciousness

**Aerobic – Moderate**

**Aerobic – low to moderate intensity**
HCM patients are less active than the general population

- Survey administered to adult HCMA patients and compared to national population

- HCM patients were overall less active at work and with recreation, but many are very active recreationally and some doing vigorous or competitive sports

- Purposeful reduction in exercise (in 64%) had negative emotional impact

Study by S Day, University of Michigan, slide courtesy of S Day
WE NEED MORE KNOWLEDGE!

• Existing guidelines are based on little more than anecdotal evidence

• We don’t know if risks are truly higher for someone with HCM to participate in competitive sports or vigorous exercise than if they were less active or sedentary. i.e. We know what makes the headlines, but we don’t know what doesn’t.

• Exercise restrictions can have a negative impact on quality of life and also on general health (obesity, sedentary lifestyle).
How do I get started exercising?

Talk to your doctor!
Exercising training and cardiac rehabilitation in patients with implantable cardioverter defibrillators: a review of current literature focusing on safety, psychological impact of programme participation and the effects of exercise training on the current literature focusing on safety, psychological impact of programme participation and the effects of exercise training on the.