So You Want to Screen . . . What About What You May Find: the "Gray-Zone" and Disqualification

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UCLA School of Medicine
Cedars-Sinai/Hospital of the Good Samaritan
October 16, 2015

October 16 - 18
14th EDITION 2015

NO CONFLICT OF INTEREST TO DECLARE

Similarities and Differences Between Bethesda Conference 36 and ESC Recommendations

- Both are written by highly qualified arrhythmia experts with strong opinions
- ESC Guidelines based on Veneto experience between 1979 and 2004: only 55 cardiovascular deaths in screened athletes, 2% disqualified, and none of them had subsequent sudden death.
- ESC Guidelines use ECG screening and BC#36 does not
- Both guidelines are very conservative and risk adverse: based on expert opinion not trial data
- Surprisingly few gray zone areas due to arbitrary exclusions

Gray Areas in 2015: Before the New Bethesda Conference Guidelines are Released

- Long QT – Too Conservative: New data from Mayo Clinic
- Hypertrophic Cardiomyopathy : Too Conservative: data from Yale Registry on ICDs in Athletes
- Represents efforts to liberalize guidelines based on new patient data

36th Bethesda Conference:
Eligibility Recommendations for
Competitive Athletes With Cardiovascular Abnormalities

Barry J. Maron, MD, FACC, *Conference Co-Chair*
Douglas P. Zipes, MD, MACC, *Conference Co-Chair*

Inherited Arrhythmia Syndromes

Recommendations:

1. Regardless of QTc or underlying genotype, all competitive sports, except those in class IA category should be restricted in a patient who has previously experience either: 1) an out-of-hospital cardiac arrest, or 2) a suspected LQTS-precipitated syncopal episode.
2. 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 (LQTS3)
4. LQTS patients with an ICD/pacemaker should not engage in sports with a danger of bodily collision because such trauma may damage the pacemaker system. The presence of an ICD should restrict individuals to class IA activities.

J Am Coll Cardiol 2005;8:1313-1361

Return to play? Athletes with congenital long QT syndrome

Jonathan N Johnson,[1] Michael J Ackerman[1,2,3]

- Ackerman "embraces the tenets of self determination and patient/family autonomy in (his) LQTS/Genetic Heart Rhythm Clinic . . . And respects the athletes and his/her family's right to make a well-informed risk-benefit decision regarding continuation of athletics.
- 7/00 to 11/10, 353 genetically confirmed LQTS between 6 and 40 years old
  - 196/353 were not in athletics
  - 27/157 athletes chose to do discontinue competitive sports
  - 130 chose to continue sports: 60/130 in conflict with BC # 36 and ESC (including 20 with ICDs) Other 67 genotype positive/ phenotype negative, 3 played Type 1A sports

Br J Sports Med 2013;47:28-33

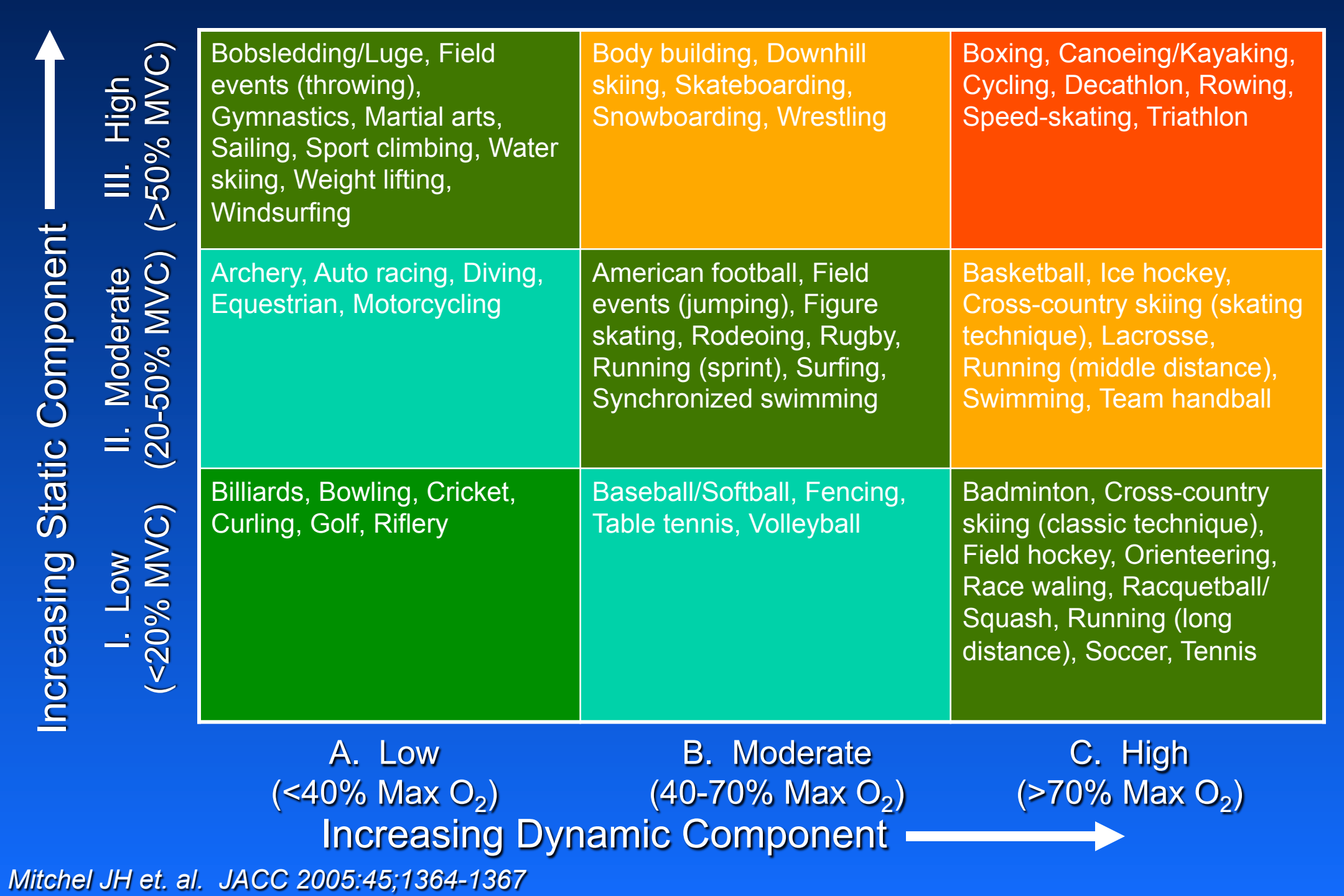
Return to play? Athletes with congenital long QT syndrome

Jonathan N Johnson,[1] Michael J Ackerman[1,2,3]

Demographics of Overall Cohort

	Total Cohort	Athletes	Non-athletes
Number of patients	353	130	223
Age at diagnosis (yrs)	17 ± 11	11 ± 7	20 ± 12
Sex (male/female)	154/199	70/60	84/139
Average QTc (ms)	472 ± 42	471 ± 46	472 ± 39
Genotype			
LQT1	182 (52%)	74 (57%)	108 (48%)
LQT2	130 (37%)	41 (32%)	89 (40%)
LQT3	37 (10%)	11 (8%)	26 (12%)
Multiple	4 (1%)	4 (3%)	0 (0%)
Symptoms	111 (31%)	29 (22%)	82 (37%)
β-blockers	280 (79%)	112 (87%)	168 (75%)
ICD	78 (22%)	20 (15%)	58 (26%)
Follow-up available (yrs)	5.5 ± 3.4	5.1 ± 2.9	5.8 ± 3.7

Br J Sports Med 2013;47:28-33



Mitchel JH et. al. JACC 2005:45;1364-1367

Return to play? Athletes with congenital long QT syndrome

Jonathan N Johnson,[1] Michael J Ackerman[1,2,3]

Increasing Static Component

	A Low	B Moderate	C High
III High	25 5 M / 20 F Age at Dx 14 ± 9 yrs QTc 485 ± 52 ms	4 3 M / 1 F Age at Dx 9 ± 6 yrs QTc 455 ± 23 ms	0
II Moderate	0	14 10 M / 4 F Age at Dx 10 ± 5 yrs QTc 470 ± 40 ms	34 26 M / 8 F Age at Dx 10 ± 5 yrs QTc 458 ± 35 ms
I Low	3 3 M / 0 F Age at Dx 11 ± 4 yrs QTc 444 ± 49 ms	22 3 M / 1 F Age at Dx 10 ± 5 yrs QTc 472 ± 34 ms	28 13 M / 15 F Age at Dx 13 ± 8 yrs QTc 477 ± 60 ms
# of patients			

Increasing Dynamic Component

Br J Sports Med 2013;47:28-33

Return to play? Athletes with congenital long QT syndrome

Jonathan N Johnson,[1] Michael J Ackerman[1,2,3]

- No deaths regardless of athletic status
- No more LQT-triggered cardiac events among athletes compared to the 223 patients not participating in competitive sports
- 1 patient – a 9 year old with LQT1, QTC of 490 msec and a H/O VF resuscitated, received two appropriate VF terminating ICD shocks. Each occurred with admittance of non-compliance with beta-blocker medication

Br J Sports Med 2013;47:28-33

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Hypertrophic Cardiomyopathy

Recommendations:

1. Athletes with a probable or unequivocal clinical diagnosis of HCM should be excluded from most competitive sports, with the possible exception of those of low intensity (class IA). This recommendation is independent of age, gender, and phenotypic appearance.

J Am Coll Cardiol 2005;8:1313-1361

2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy: Executive Summary
A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines
Developed in Collaboration With the American Association for Thoracic Surgery, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

Class III: Harm

1. ICD placement as a routine strategy in patients with HCM without an indication of increased risk is potentially harmful. *(Level of Evidence: C)*
2. ICD placement as a strategy to permit patients with HCM to participate in competitive athletics is potentially harmful. *(Level of Evidence: C)*
3. ICD placement in patients who have an identified HCM genotype in the absence of clinical manifestations of HCM is potentially harmful. *(Level of Evidence: C)*

W. K.– 20-Year-Old Center on Pepperdine University Basketball Team

- In prior good health
- 11/26/02 while running the court in practice, sat down and fainted, hitting his head: no palps or premonitory sx
- No FH of cardiac disease: F 53, M 52 (MD) in good health; sister 27 A&W
- Echo: 12/02 HCM, septum 17-20 mm, posterior wall 18-21 mm, LV mass 389 gms, No SAM, nl LV motion LA 3.9 cm
- Holter: Rare multiform VPCs
- MRI: LVH: EF 66%
- ICD implanted

Subsequent course

- Turned down for athletic scholarship at Pepperdine & UCLA despite pt & family signed waivers, supportive letters from EPs re: safety of playing with ICD
- Ultimately played Division I at Texas Tech for two years without complication: now a basketball coach at UC Riverside
- Took approval of Dr Ken Schein (Provost of UT) to achieve this outcome

THE WALL STREET JOURNAL.

Go-To Guy

Why Heart Trouble Doesn't Sideline Some Athletes

Dr. Cannon Helps Players Touched by Their Schools; Defibrillators and Testing

Entering the Line of Football

Doctors worry a defibrillator won't withstand the rigors of competition.

Postulated Risks of Sports

- Death
- Inability of the ICD to defibrillate due to the influence of the effects of vigorous exercise
- Injury due to syncopal arrhythmia or shock
- Damage to the ICD or lead system

ICD
SPORTS
REGISTRY

Do you have an **Implantable Cardioverter–Defibrillator (ICD)?**

Do you participate in **SPORTS?**

ICD SPORTS REGISTRY

Principle investigators

Rachel Lampert, MD
Yale University

Brian Olshansky, MD
University of Iowa

Christine Lawless, MD
Ohio State University

Elizabeth Saarel, MD
University of Utah

David Cannom, MD
Los Angeles Cardiology Associates
Past President, Heart Rhythm Society

Steering Committee

Hugh Calkins, MD
Johns Hopkins

Mark Estes, MD
New England Medical Ctr

Mark Link, MD
New England Medical Ctr

Barry Maron, MD
Minneapolis Heart Institute

Frank Marcus, MD
University of Arizona

James Perry, MD
Yale University
Past President, Pediatric
Electrophysiology Society

Melvin Scheinman, MD
UC San Francisco

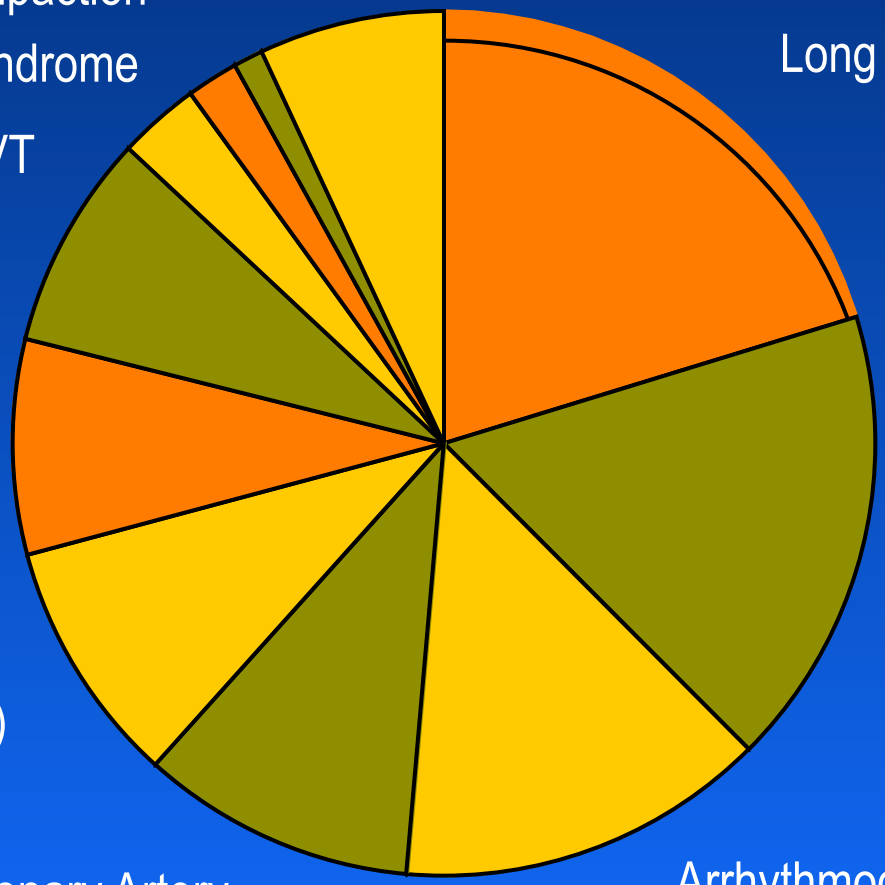
Bruce Wilkoff, MD
Cleveland Clinic Foundation

Douglas Zipes, MD
Indiana University

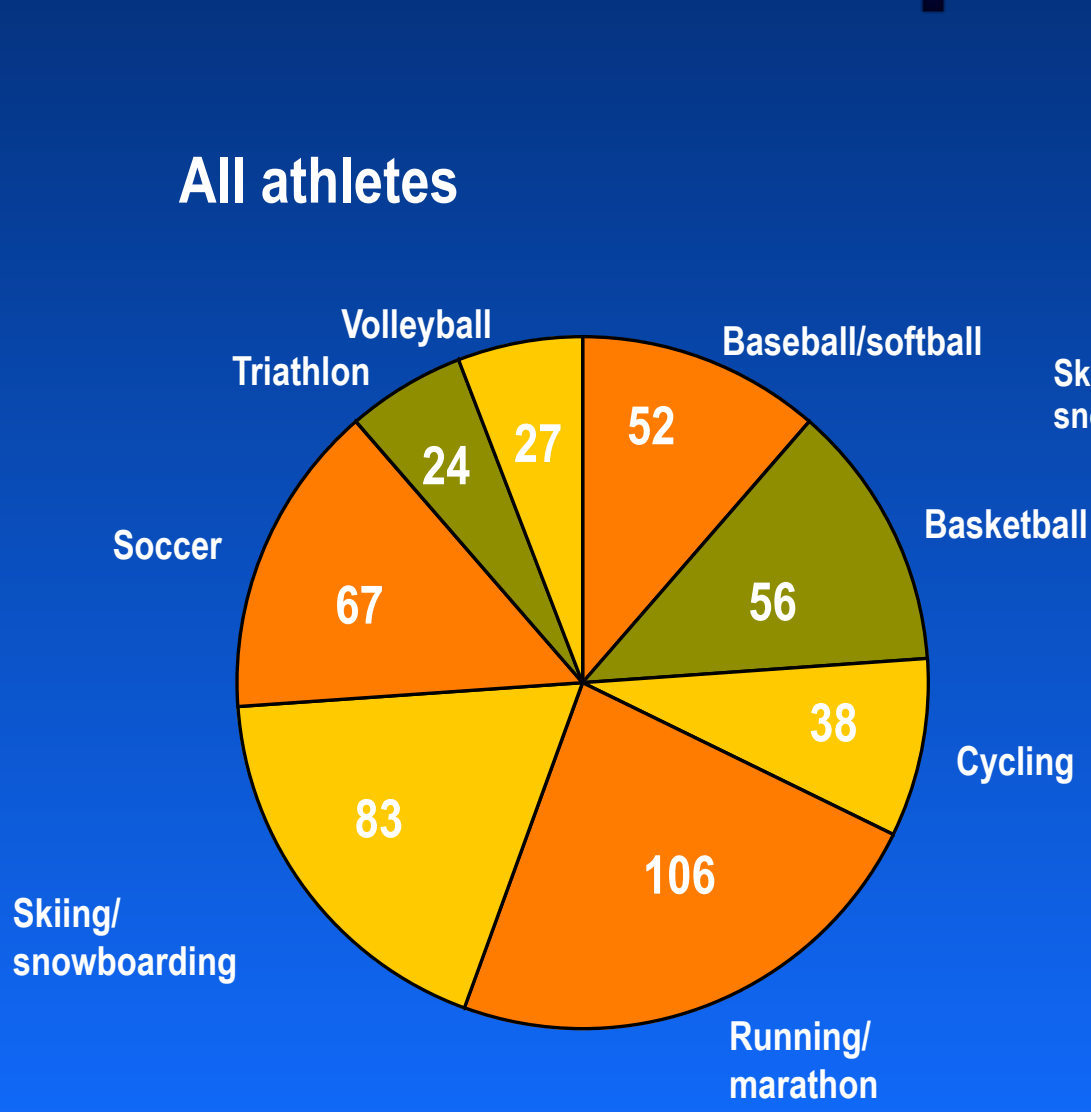
Patient Population: Yale Study

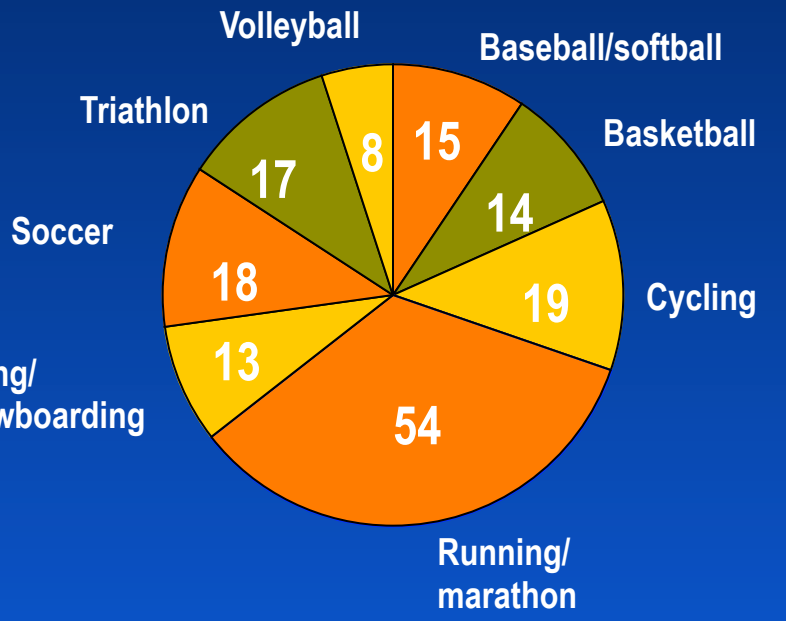
Number	372
Median follow-up, months	31 (21-46)
Age, years	
10-19	89 (24%)
20-39	136 (37%)
40-60	147 (39%)
Male gender	249 (67%)
Caucasian race	349 (94%)
Time since initial ICD implantation, months	27 (12-59)
Ejection fraction, %	60 (50-66)
Taking beta-blocking agents	229 (62%)
ICD indication	
primary prevention	155 (41%)
secondary prevention (102 SCD -- 42 SCD w/ sports)	217 (59%)

Cardiac Diagnoses



Patient Population: Sports Participation



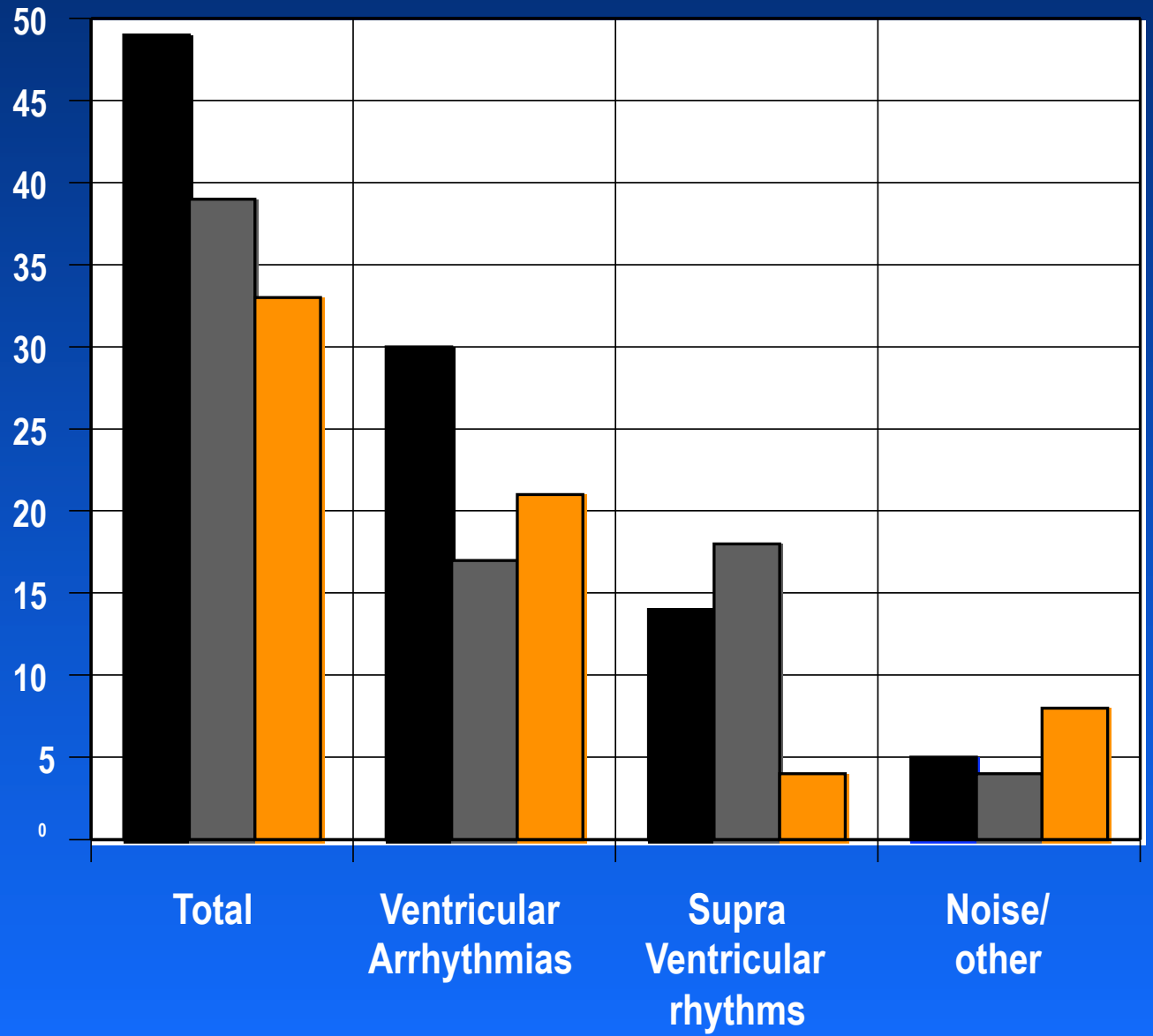


Elite Athletes

Results: Primary Endpoints

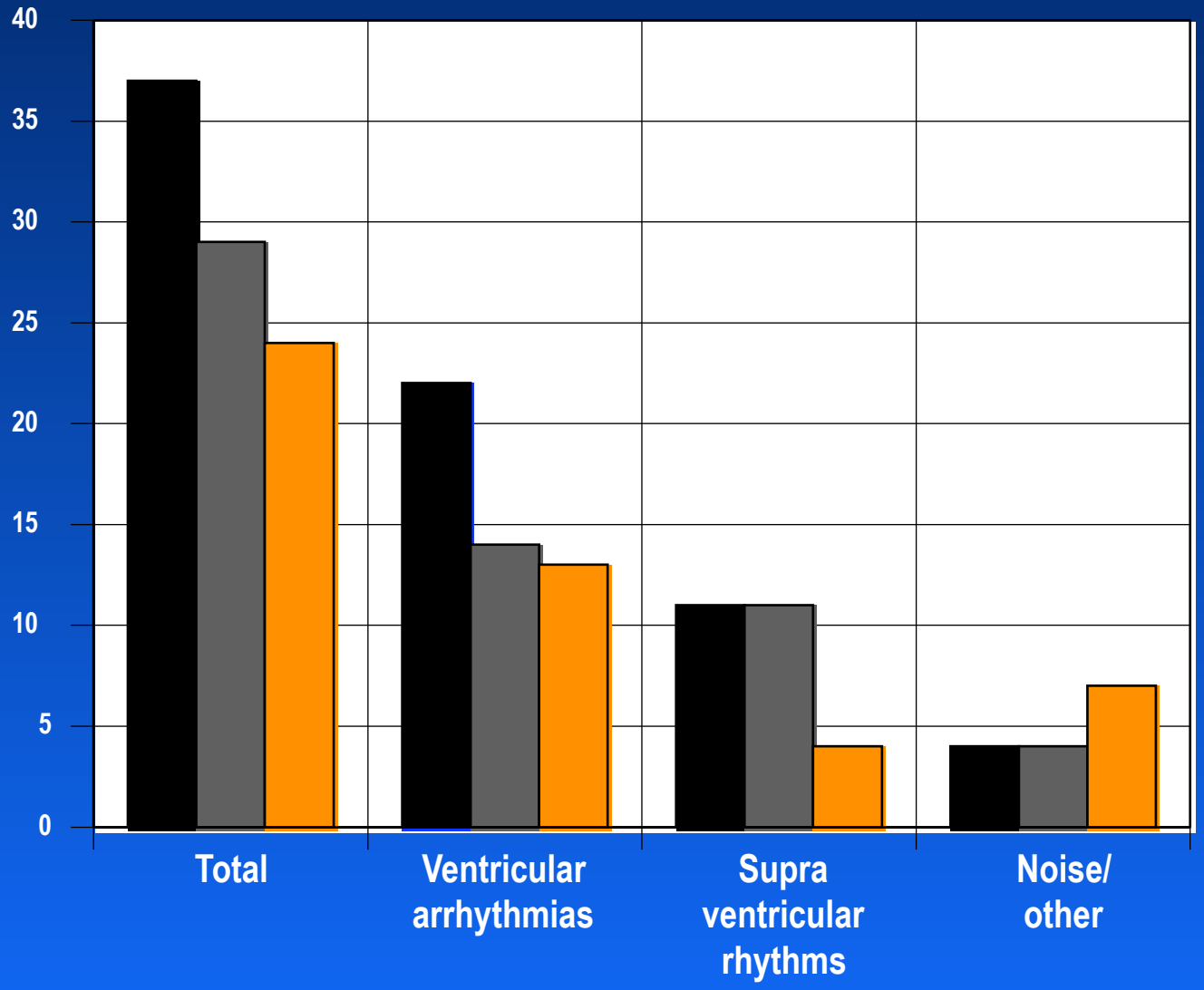
- Tachyarrhythmic death or externally resuscitated tachyarrhythmia during or after sports: 0
- Injury due to arrhythmia or shock during sports: 0
- 95% confidence interval occurrence of endpoints:
  - 1 year (315 athletes): 0-1.2%
  - 2 years (243 athletes): 0-1.5%

Shocks Received (121)



*includes practice, post-competition/practice

Individuals receiving shocks (77 pts)



Competition*

Other Physical Activity

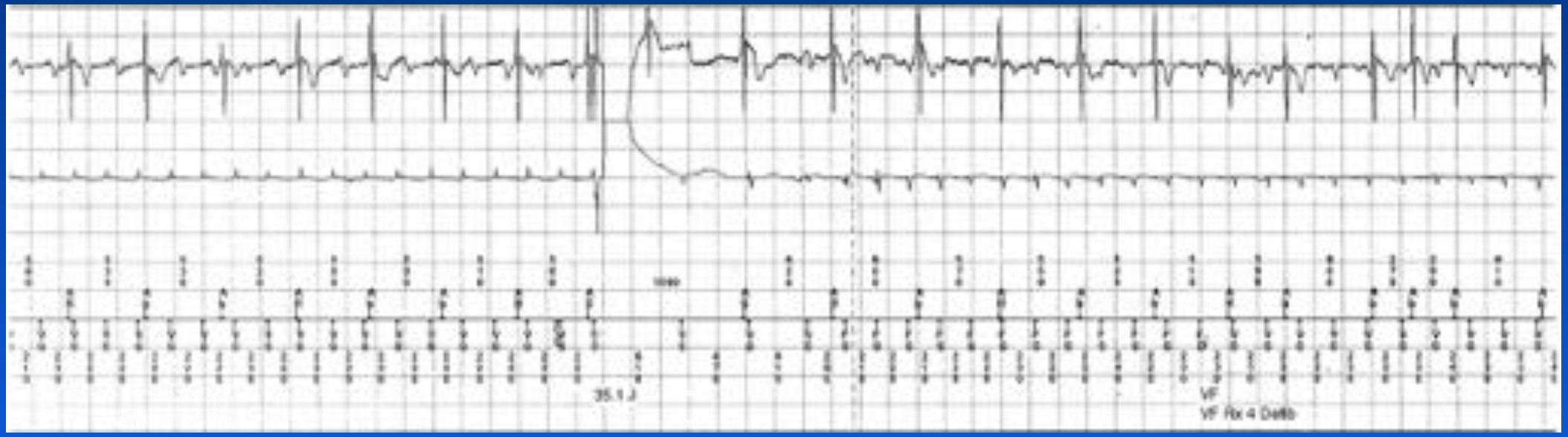
Other/Rest

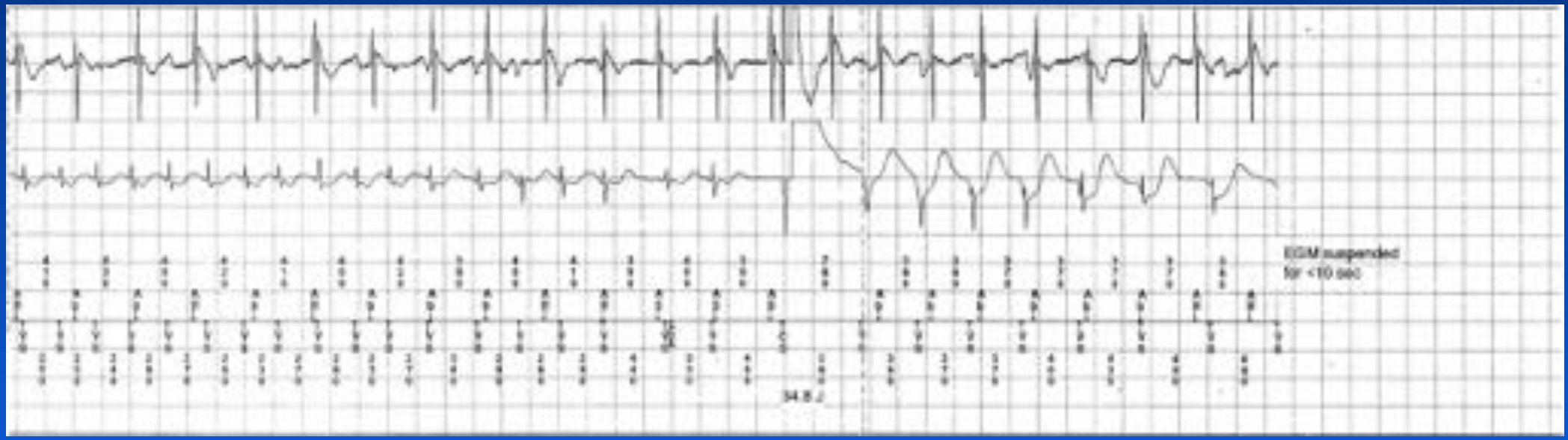
*includes practice, post-competition/practice

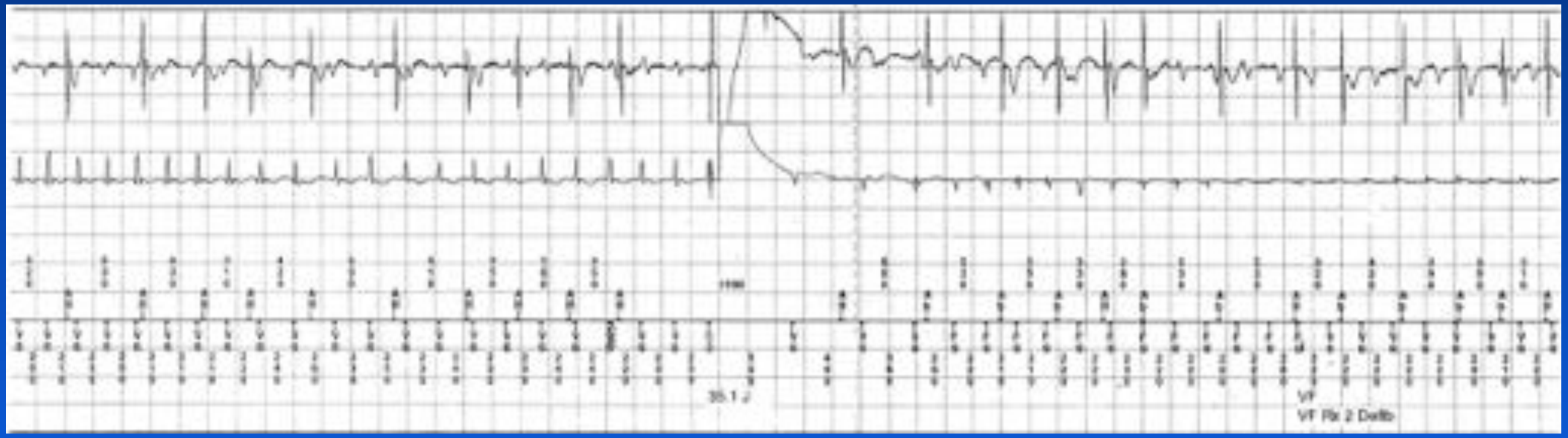
Ventricular arrhythmias requiring multiple shocks for termination

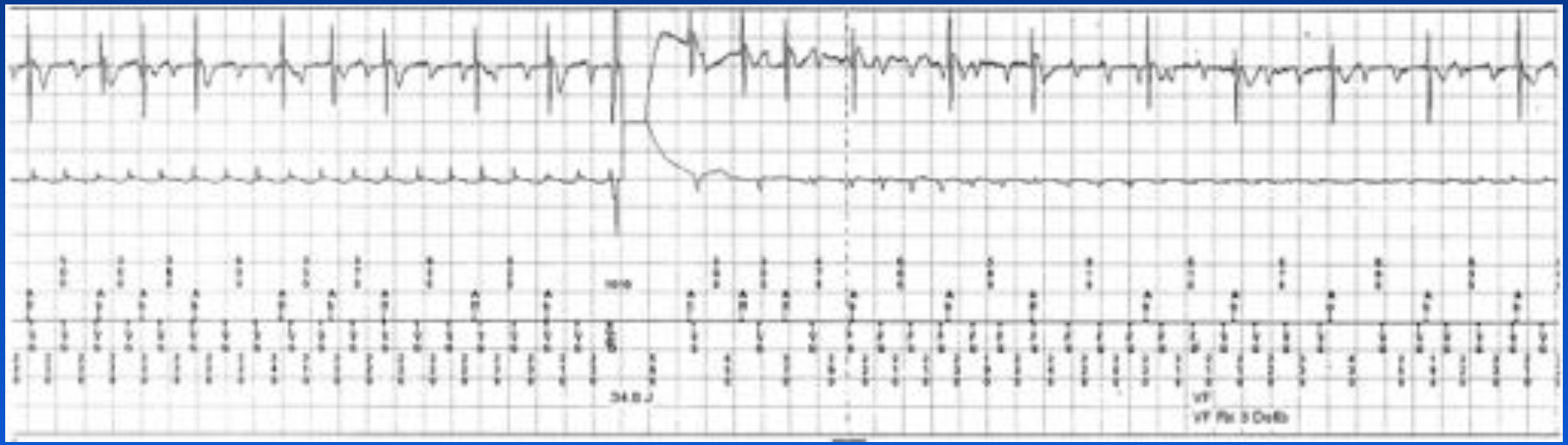
Sex	Age	Cardiac Dx	Primary Sport	Activity	Activity Type	# of shocks
M	28	Idiopathic VF	Ultimate Frisbee	Ultimate Frisbee	Competition	5
F	47	Idiopathic VF	Cycling	Cycling	Practice	4
M	44	CAD	Running	Running	Practice	2
M	50	CAD	Cycling	Cycling	Practice	6
M	57	CAD	Tennis, Basketball	Walking	Physical Activity	6
F	16	CPVT	Lacrosse Field hockey	Running	Post-physical activity	3
				Running	Post-physical activity	4
M	15	HCM	Baseball	Socializing	Other	2

CAD S/P PCI; EF 40%;
Meds Sotalol 80 mg bid
Occurred while walking


35.1 J
VF
VF Rx 4 Delib


EGM suspended
for <10 sec
34 B J


35.1 J
VF
VF Rx 2 Delib


34.8 J
VF
VF Rx 3 Delib

Implications of Registry Data

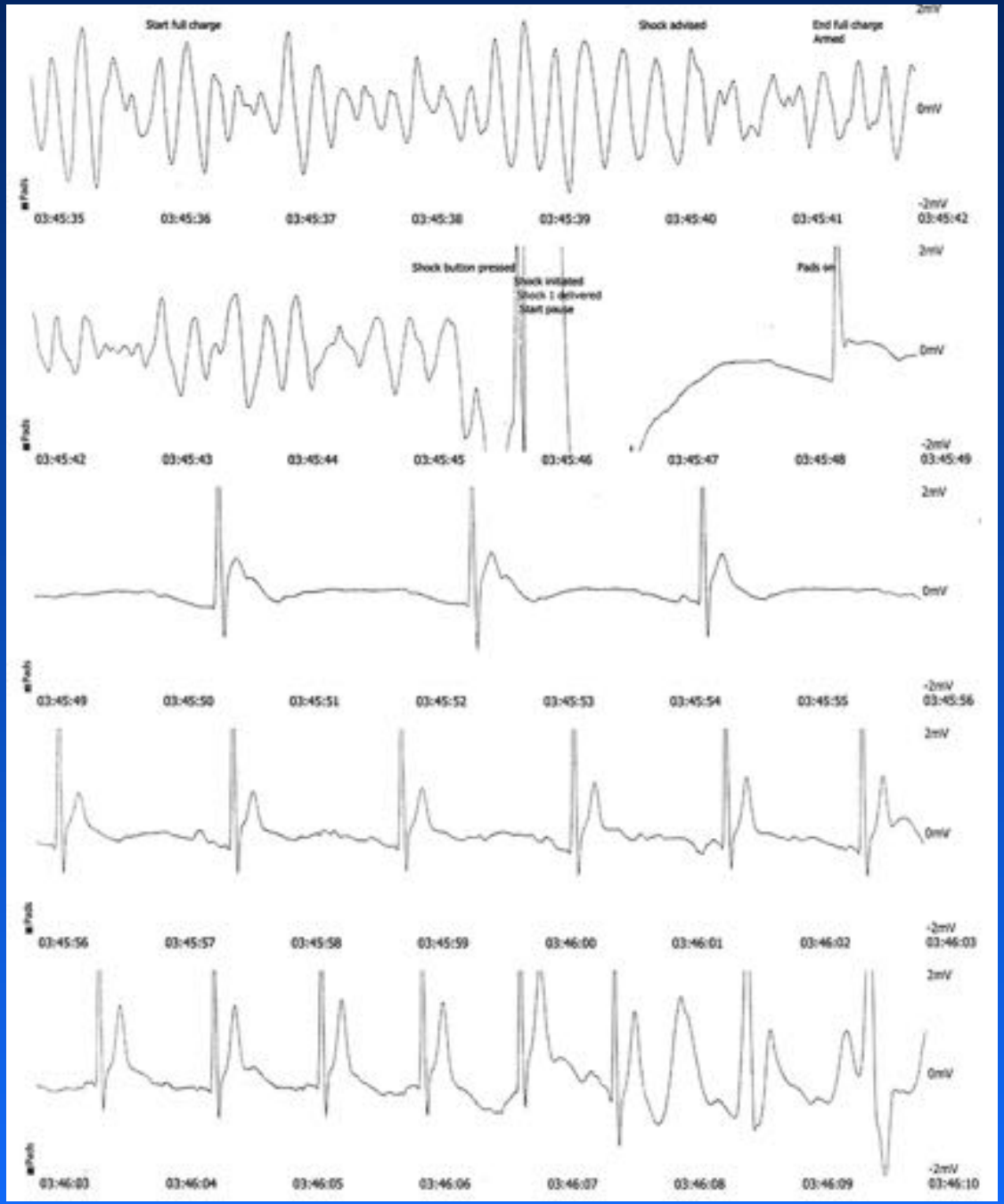
- Many athletes engage in sports with ICD without physical harm
- ICD shocks occured but there were no tachyarrhythmic deaths, resuscitated cardiac arrests or injuries related to sports participation
- Athletic participation for these patients is an issue of quality of life: shocks can decrease Q of L, but so can sports restriction
- Caveats: patients with multiple shocks should restrict exercise until situation stabilized (includes patients with CPVT, idiopathic VF and CAD)

What happens in U.S. When Patients are Disqualified?

- Physician shopping is common

EN: Tennessee BB player 2009

- 21 Nigerian male in previous good health
- Running laps after practice, collapsed, resuscitated by trainers with Phillips Heart Start
- Had been cleared to play BB even after abn EKG noted
- Post arrest: normal CTA and normal HCM genetic testing
- Dx apical HCM and received single chamber ICD
- Played BB at University of New Mexico: turned down by Tennessee and Indiana


Start full charge
Shock advised
End full charge
Armed
2mV
0mV
-2mV
Pads
03:45:35
03:45:36
03:45:37
03:45:38
03:45:39
03:45:40
03:45:41
03:45:42
Shock button pressed
Shock initiated
Shock 1 delivered
Start pause
Pads on
03:45:43
03:45:44
03:45:45
03:45:46
03:45:47
03:45:48
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03:46:00
03:46:01
03:46:02
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03:46:06
03:46:07
03:46:08
03:46:09
03:46:10

EN

Originally Published: May 18, 2010

Negedu moves on after brush with death

Tennessee and Indiana refused to take the risk, but Nigerian welcomed at New Mexico

By Andy Katz
ESPN.com
Archive

Emmanuel Negedu remembers the only thing he ate or drank on Sept. 28, 2009, was a milk shake. He can describe everything he did that morning and early afternoon, highlighting his weight lifting and workout sessions at the University of Tennessee's indoor football practice facility.

But he won't ever remember that moment when his heart stopped during a cardiac arrest, and he collapsed on the field before UT trainer Chad Newman and director of sports medicine Jason McVeigh heroically brought Negedu back to life with CPR and an automatic external defibrillator.

Remarkably, just seven-plus months later and after an internal cardiac defibrillator (ICD) was implanted in his chest, Negedu will play basketball again.

Klenck said he called doctors around the country and within the SEC to gauge public opinion on the subject. Negedu had already gone to see another specialist in Cleveland, seeking other opinions. He also got another opinion in Los Angeles from noted cardiologist Dr. David S. Cannom, the director of cardiology at Good Samaritan Hospital. Cannom had cleared former Pepperdine player Will Kimble to play at UTEP after the Waves wouldn't let Kimble return to the court after collapsing in 2002. Cannom told Kimble that his ICD implant would allow him to play competitive sports again.

[+] Enlarge

Cliff Welch/Icon SMI

Negedu's promising career at Tennessee was cut short after suffering a cardiac arrest last fall.

Matthew Emmons/US Presswire

Negedu hasn't competed in a game since Tennessee's first-round matchup with Oklahoma State in the 2009 NCAA tournament.

Guidelines: what to do?

- Need to maintain high level of expertise on BC and ESC guidelines
- Attempt to take in consideration new data eg Mayo study and Yale study
- Need to allow some level of carefully thought out compromise for patients at known risk who with their family and physician desire to play anyway
- Need to get new BC guidelines published


ID:001469777
29-SEP-2009 01:40:15
UNIV OF TN MED CTR-ER ROUTINE RECORD
23-OCT-1988 (20 yr)
Male Black
Room S759
Loc:3
Vent. rate 62 BPM
PR interval 226 ms
QRS duration 98 ms
QT/QTc 446/452 ms
P-R-T axes 44 69 -6
Sinus rhythm with 1st degree A-V block
ST elevation, consider early repolarization, pericarditis, or injury
ST & T wave abnormality, consider anterolateral ischemia
Abnormal ECG
When compared with ECG of 28-SEP-2009 16:13,
No significant change was found
Confirmed by SCOTT, MD, CHRISTOPHE (108), editor KING, JENNIFER (19) on 9/29/2009 8:43:02 AM
Technician: BW
Test ind: abnormal ekg
Referred by: . No Referring Physician
Confirmed By: CHRISTOPHE SCOTT, MD
I
II
III
aVR
aVL
aVF
V1
V2
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V4
V5
V6
25mm/s 10mm/mV 40Hz 7.1.1 12SL 237 CID: 2
EID:19 EDT: 08:43 29-SEP-2009 ORDER: 308050438 ACCOUNT: 1469777000
Page 1 of

EN

36TH BETHESDA CONFERENCE

Introduction: Eligibility Recommendations for Competitive Athletes With Cardiovascular Abnormalities—General Considerations

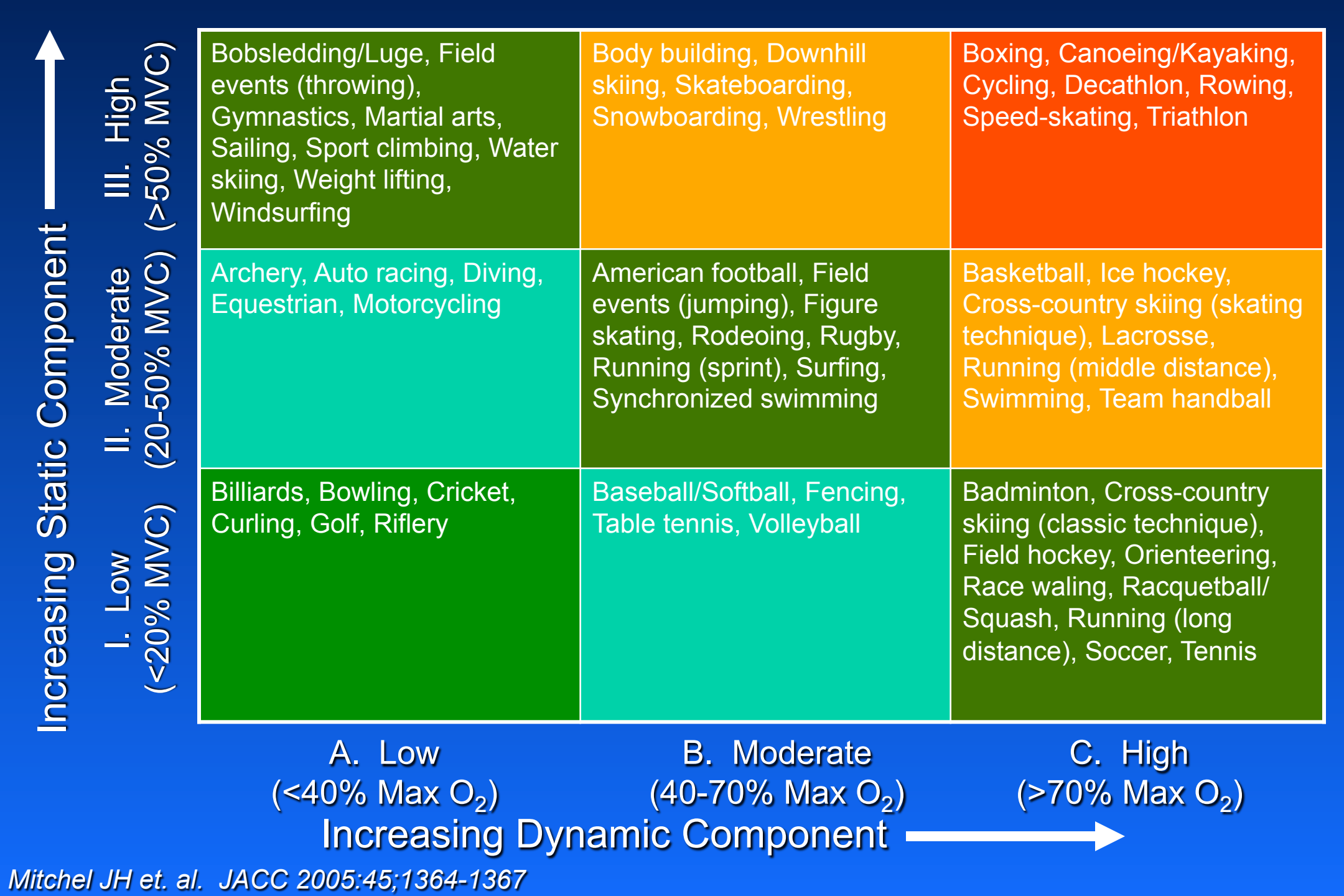
Barry J. Maron, MD, FACC, *Co-Chair*
Douglas P. Zipes, MD, MACC, *Co-Chair*

"Although effective for sudden death prevention in observational studies, the unique physiologic milieu associated with competitive athletic activities, including intravascular volume and electrolyte disturbances, neurohormonal activity, and the potential for myocardial ischemia make the absolute reliability of ICDs in such settings unpredictable."

Maron BJ and Zipes DP. JACC 2005:45;1318-1321

"Although differences of opinion exist and little direct evidence is available, the panel asserts that the presence of an ICD (whether for primary or secondary prevention of sudden death) should disqualify athletes from most competitive sports (with the exception of low-intensity, class IA), including those that potentially involve bodily trauma.

Maron BJ and Zipes DP. JACC 2005:45;1318-1321



Mitchel JH et. al. JACC 2005:45;1364-1367

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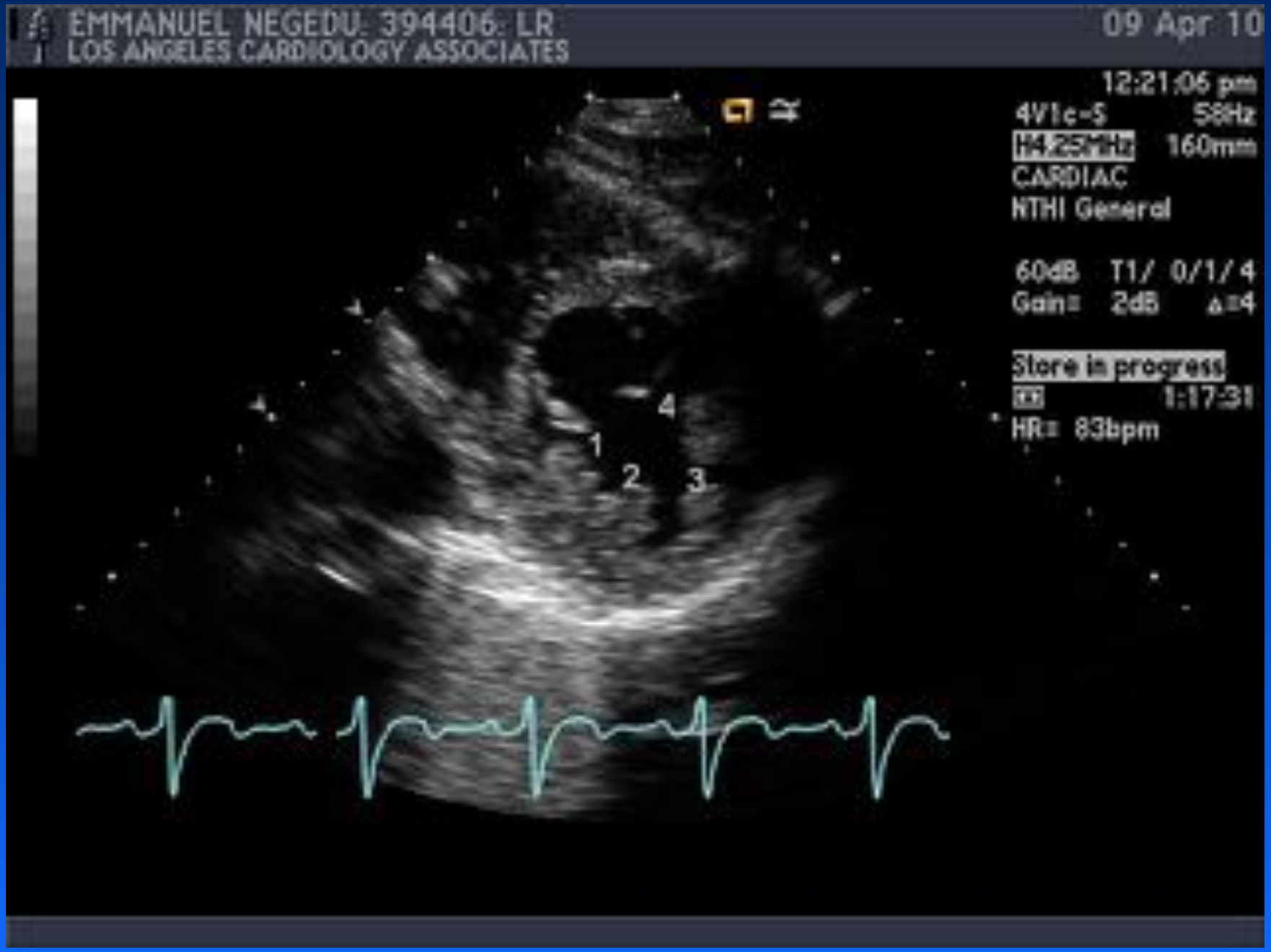
" Indeed, the managing physician with particular knowledge regarding a given athlete's cardiovascular abnormality, psychological response to competition, and other medically relevant factors may choose to adopt somewhat different recommendations in selected individuals."

Maron BJ and Zipes DP. JACC 2005:45;1318-1321


EMMANUEL NEGEDU. 394406. LR
LOS ANGELES CARDIOLOGY ASSOCIATES
09 Apr 10
12:21:06 pm
4V1c-S
58Hz
160mm
CARDIAC
NTHI General
60dB T1/ 0/1/ 4
Gain= 2dB
Store in progress
1:17:31
HR= 83bpm
1
2
3
4

Follow-Up

- Median follow-up: 31 mos (21-46 mos)
- 21 did not complete study:
  - 9 lost to follow up (all confirmed alive)
  - 6 withdrew
  - 4 developed worsening cardiac or medical conditions precluding sports
  - 2 died
  - 52 yo cyclist with CAD died at work (desk job) after receiving multiple shocks
  - 34 yo volleyball/softball player with familial cardiomyopathy died during CHF hospitalization

Did ICD Shocks Affect Sports Participation?

- 37 received ICD shocks during sports
  - 4 stopped sports completely
  - 7 stopped one or some sports
- 5 patients stopped at least one sport due to shocks received at other times

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Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

Recommendation:

1. Athletes with probable or definite diagnosis of ARVC should be excluded from most competitive sports, with the possible exception of those of low intensity (class IA)

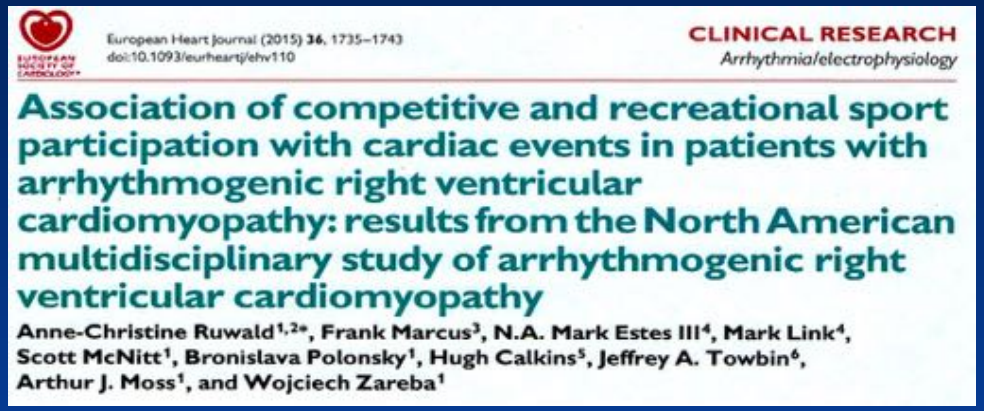
J Am Coll Cardiol 2005;8:1313-1361

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 Ruwald[1,2*], Frank Marcus[3], N.A. Mark Estes III[4], Mark Link[4], Scott McNitt[1], Bronislava Polonsky[1], Hugh Calkins[5], Jeffrey A. Towbin[6], Arthur J. Moss[1], and Wojciech Zareba[1]

- LaGerche showed that endurance exercise is associated with isolated pronounced dilation of the RV and decreased function
- Endurance competitive exercise might promote RV changes compatible with the diagnosis of ARVC with a low frequency of genotype positive individuals
- Dilation of the RV associated with competitive/endurance exercise may lead to excessive myocardial damage and fibrofatty replacement of the RV
- Larger RV volumes were only seen in patients who participated in competitive sports eg. running, biking, basketball, soccer. No risk for high dynamic sports on a recreational level while significant risk on a competitive level

European Heart Journal 2015;36:1735-1743



European Heart Journal (2015) 36, 1735–1743
doi:10.1093/eurheartj/ehv110

CLINICAL RESEARCH
Arrhythmia/electrophysiology

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

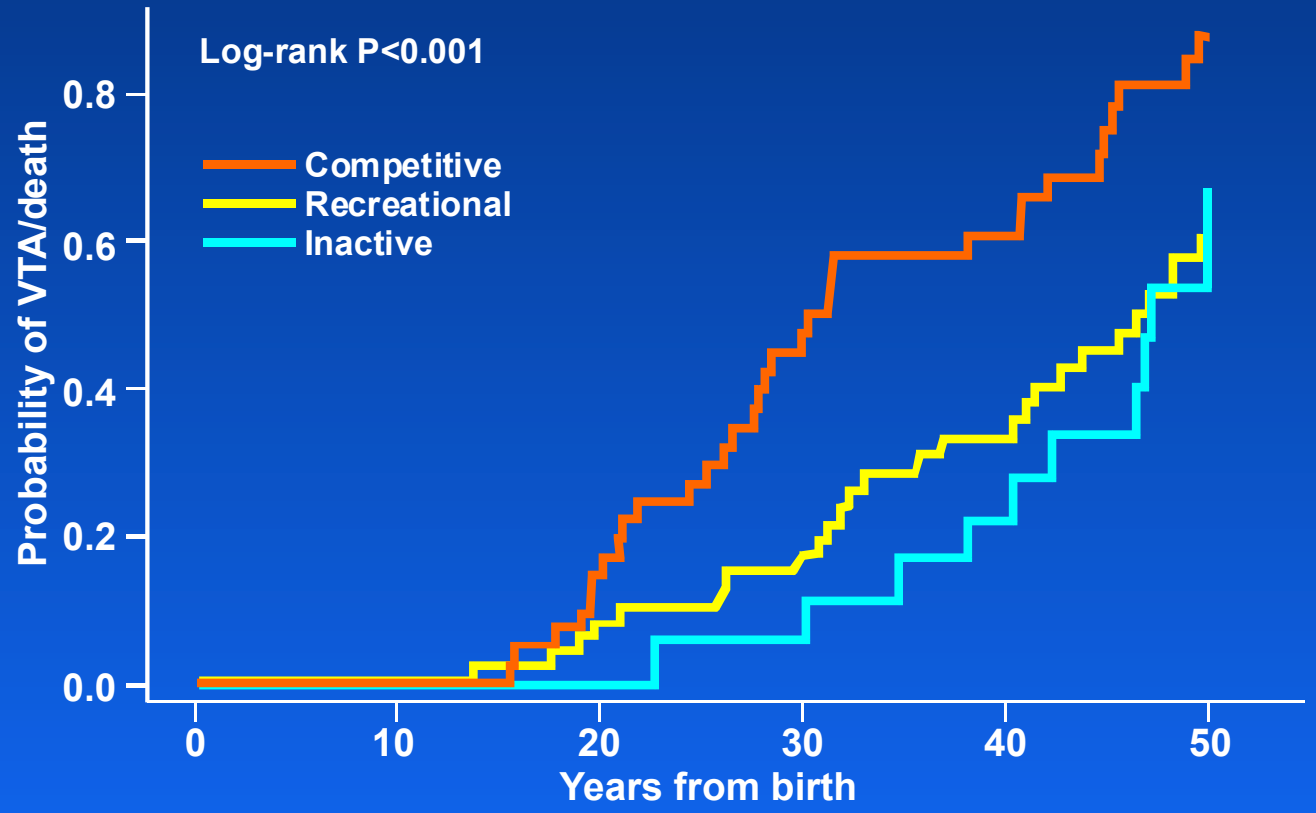
Anne-Christine Ruwald[1,2*], Frank Marcus[3], N.A. Mark Estes III[4], Mark Link[4], Scott McNitt[1], Bronislava Polonsky[1], Hugh Calkins[5], Jeffrey A. Towbin[6], Arthur J. Moss[1], and Wojciech Zareba[1]

- 108 patients with ARVC according to 2010 task force criteria
- Study patients were questioned about exercise level prior to and after ARVC diagnosis with 3 categories of sports competitive (41), recreational (48) and inactive (19)
- Competitive sport was associated with significantly higher risk of SCD compared to recreational sport (HR 1.99) and inactive patients (HR 2.05)
- Symptoms developed at an earlier age in competitive 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 Ruwald[1,2*], Frank Marcus[3], N.A. Mark Estes III[4], Mark Link[4], Scott McNitt[1], Bronislava Polonsky[1], Hugh Calkins[5], Jeffrey A. Towbin[6], Arthur J. Moss[1], and Wojciech Zareba[1]

Probability of VTA/death from Birth by Sports Level



Number of patients at risk					
41	41 (0.00)	35 (0.15)	20 (0.47)	15 (0.61)	4 (0.87)
48	48 (0.00)	43 (0.08)	37 (0.17)	29 (0.33)	15 (0.60)
19	19 (0.00)	19 (0.00)	17 (0.05)	14 (0.22)	5 (0.67)

European Heart Journal 2015;36:1735-1743