Inherited Arrhythmia in Athletes

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COI Disclosure

Hwa jung Kim:

The authors have no financial conflicts of interest to disclose concerning the presentation

Athletes

Individuals who are engaged in exercise training on a regular basis and participate in official sports competition

Elite athletes
: Moderate to vigorous exercise ≥ 10 h/week

Competitive athletes
: ≥ 6 h/week

Recreational athletes
: ≥ 4 h/week

SCD in athletes

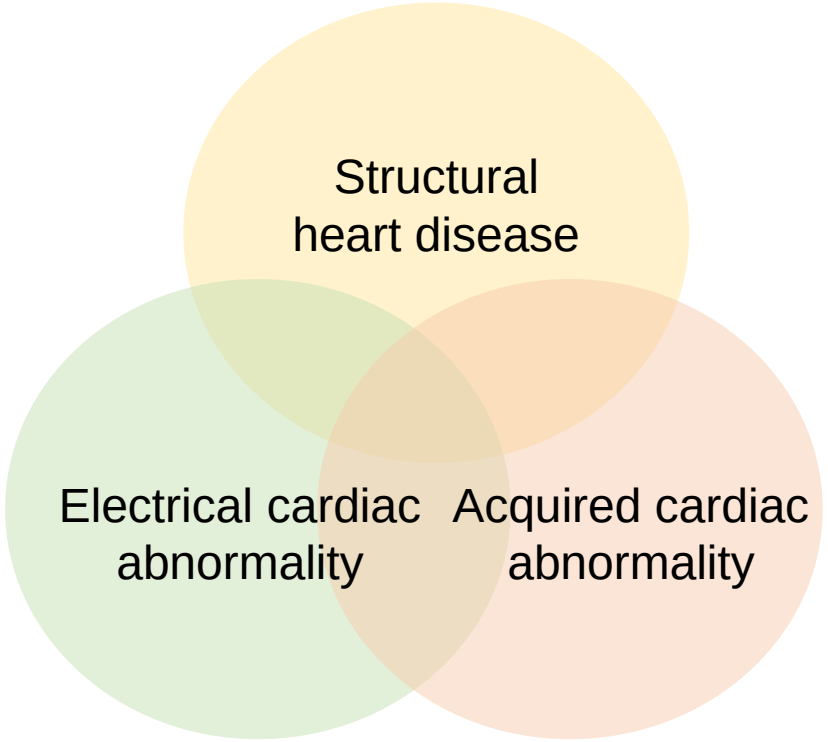
Leading medical cause of death

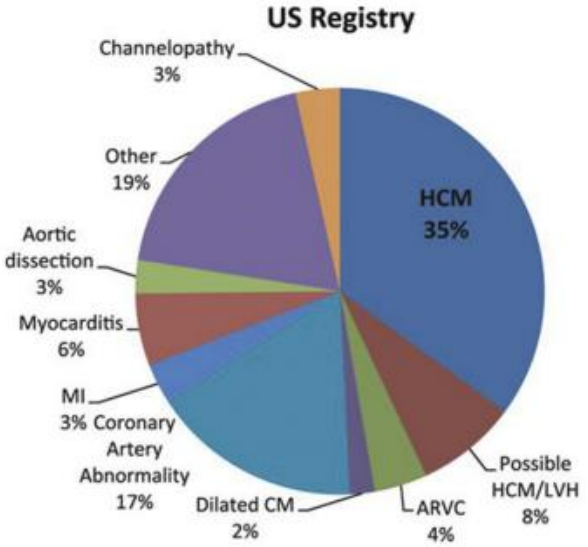
- 1 : 10,000 to 1 : 400,000 per year
- Competitive athletes 2.4 to 4.5 times higher than non- / recreational athletes
- Male athletes 3 to 5 times higher
- Black athletes 3.2 times higher
- 1 : 5,200 per year in basketball players

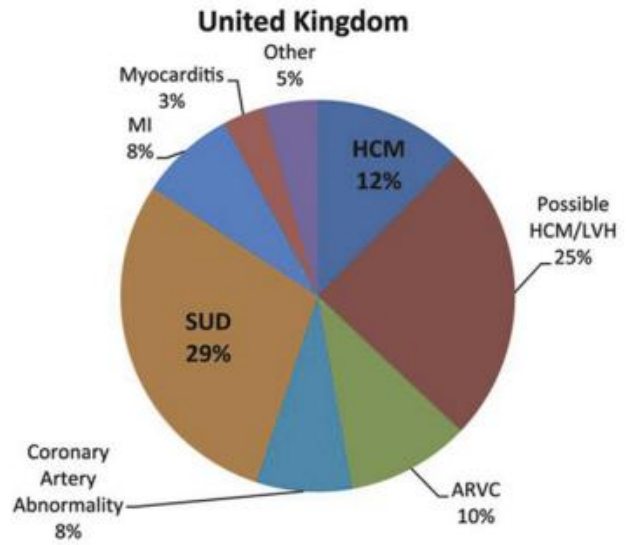
JACC HF January 2018:30 – 40

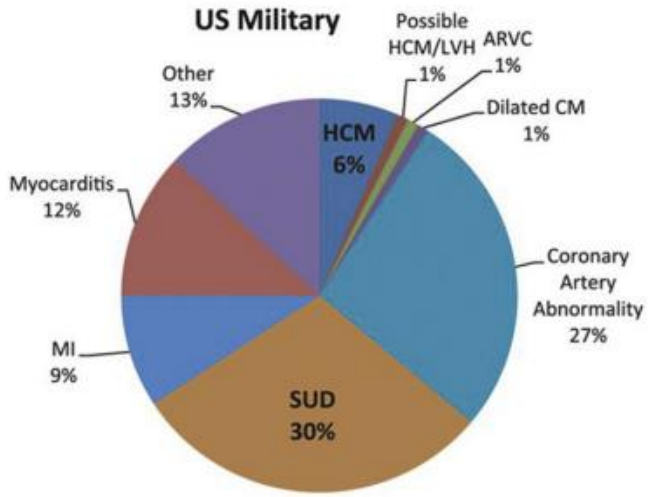
N Engl J Med 2018;379:524-34

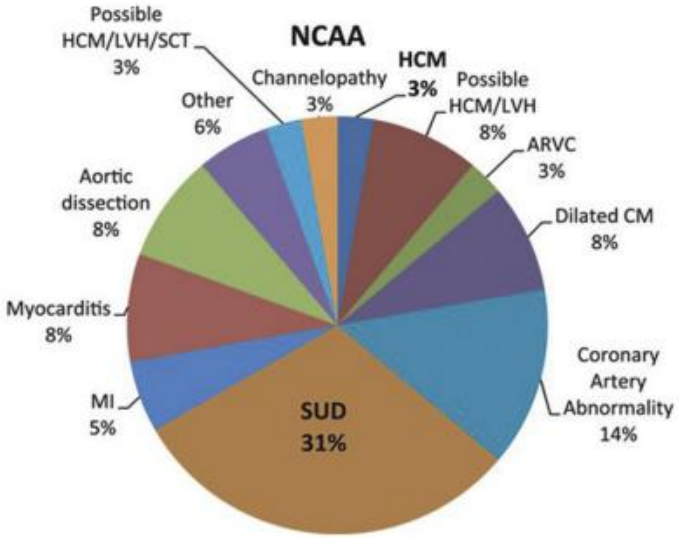
Cause of SCD in athletes











JACC HF January 2018:30 – 40

KHRS 2023

- Primary inherited arrhythmia

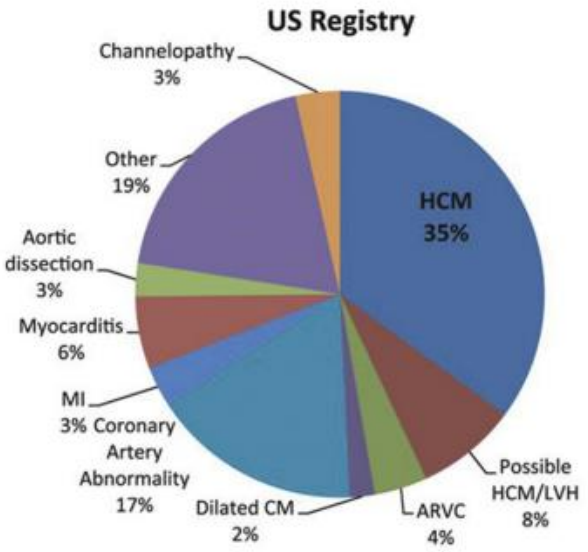
  0 to 5% of all SCD in athletes

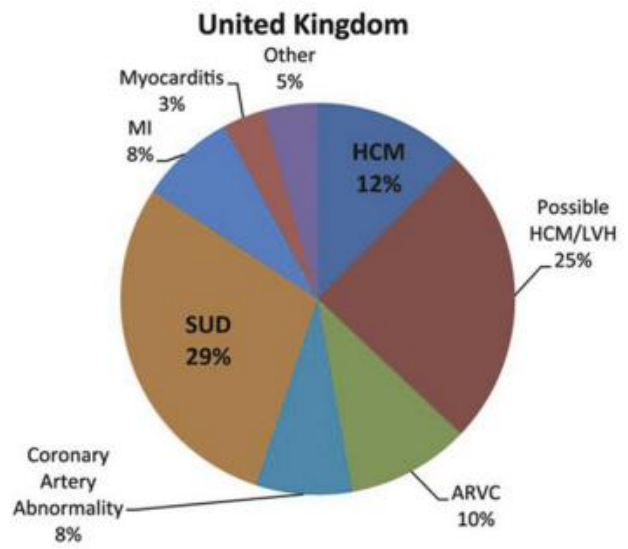
- Sudden Unexplained Death (SUD)

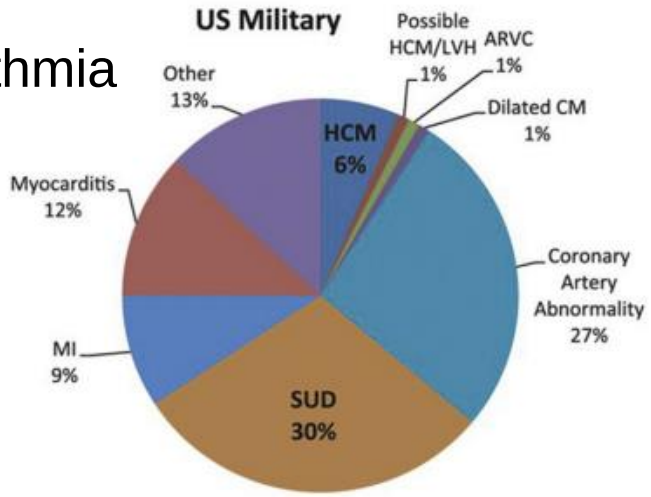
  ≒ Sudden arrhythmic death syndrome

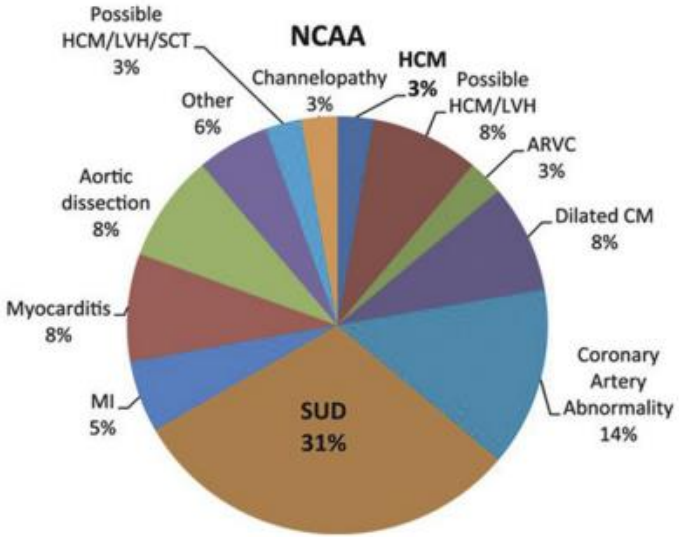
  Morphologically normal heart at autopsy

  Many of these are probably inherited arrhythmia









JACC HF January 2018:30 – 40

Inherited arrhythmia

Long QT syndrome (LQTS)

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)

Brugada syndrome (BrS)

Short QT syndrome (SQTS)

Inherited Cardiomyopathy

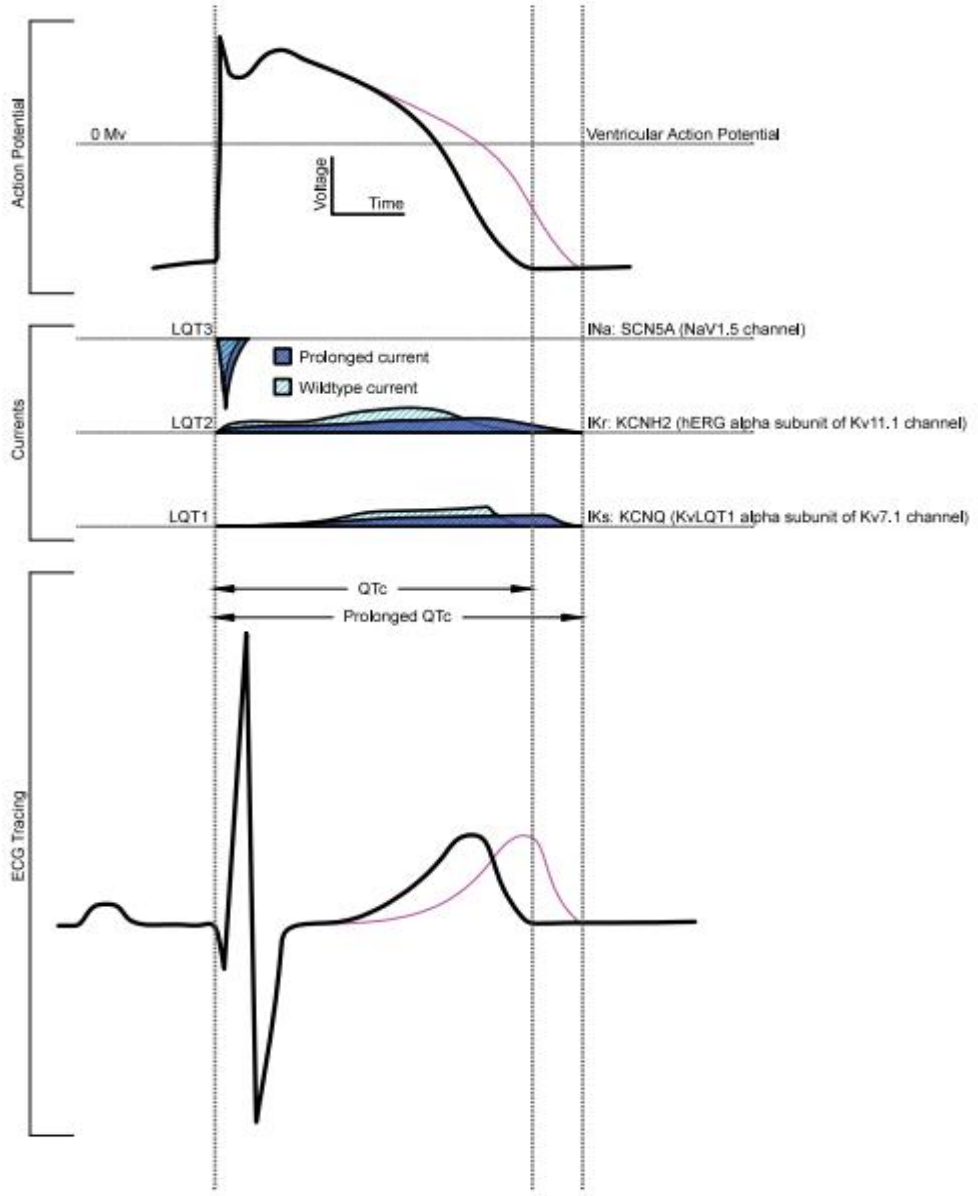
Hypertrophic cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy

LQTS

- 1:2000 in Caucasian, 1:5000 in Korean
- 2 of 2,352 Olympic athletes (2004 to 2014)
- 0 to 5% of all SCD in athletes
- Suspected if QTc ≥ 470ms for male,

  ≥ 480ms for female athletes
- Diagnosed if QTc ≥ 500ms

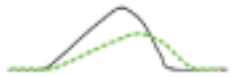


Pacing Clin Electrophysiol34(6):717–723

Br J Sports Med 2017;51:238–243

Clin Sports Med 41 (2022) 369–388

Br J Sports Med 2021;55:1196–1203

	Type 1	Type 2	Type 3
Gene	*KCNQ1*	*KCNH2*	*SCN5a*
Protein	$K_v7.1$ 	$K_v11.1$ 	$Na_v1.5$ 
Effect on current	$I_{Ks}\downarrow$ 	$I_{Kr}\downarrow$ 	$I_{Na,L}\uparrow$ 
Frequency among LQTS	± 35% 	± 30% 	≤ 10% 
Penetrance	± 65% 	± 80% 	± 90% 
Main trigger of events	Exercise *(swimming)* 	Arousal 	Rest 
Age of onset arrhythmias	Childhood 	Puberty 	Puberty 
Gender most at risk	♂	♀	♀

Heart 2022;108:332–338

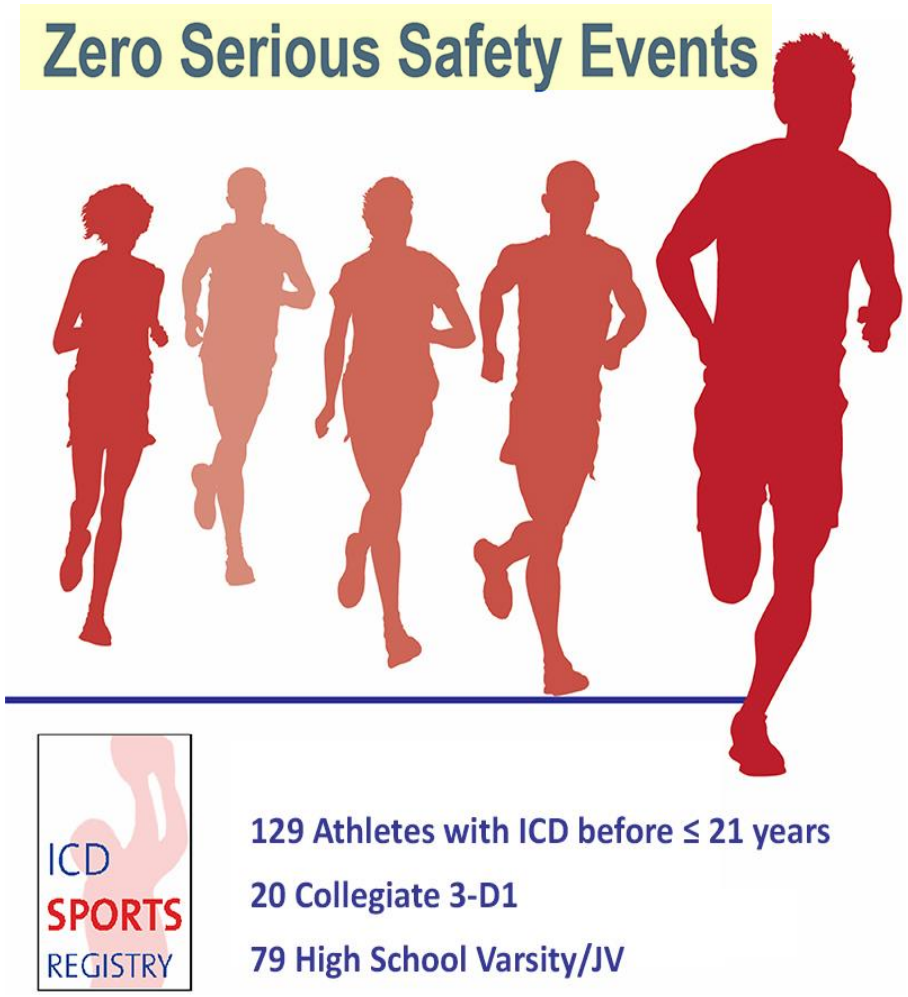
Prior guidelines for athletes

2005 The Bethesda Conference Guidelines

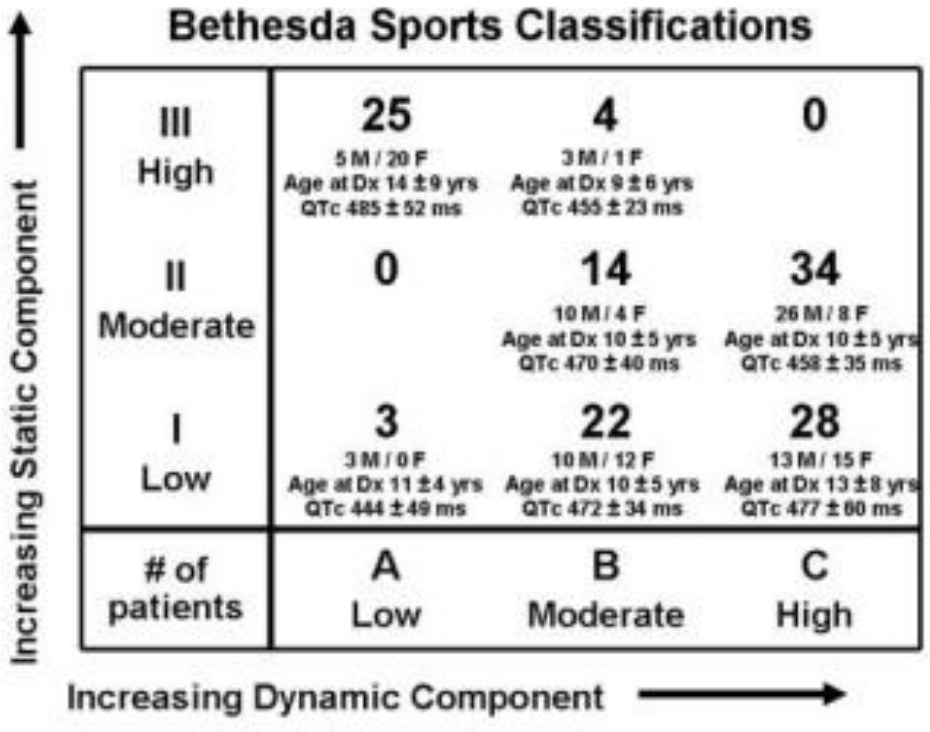
All symptomatic LQTS patients, those with ECG manifestations should be restricted all competitive sports, except class IA category (billiards, bowling, cricket, curling, golf…)

2005 ESC Guidelines

All LQTS patients not participate in any sport even for those without documented major arrhythmic events



LQTS n=49



130 athletes with LQTS

86% with BB, 16% with LCSD, 15% with ICD

60 athletes participated (against recommendation)

No death, 2 appropriate shock

Circulation: Arrhythmia and Electrophysiology. 2018;11:e006305

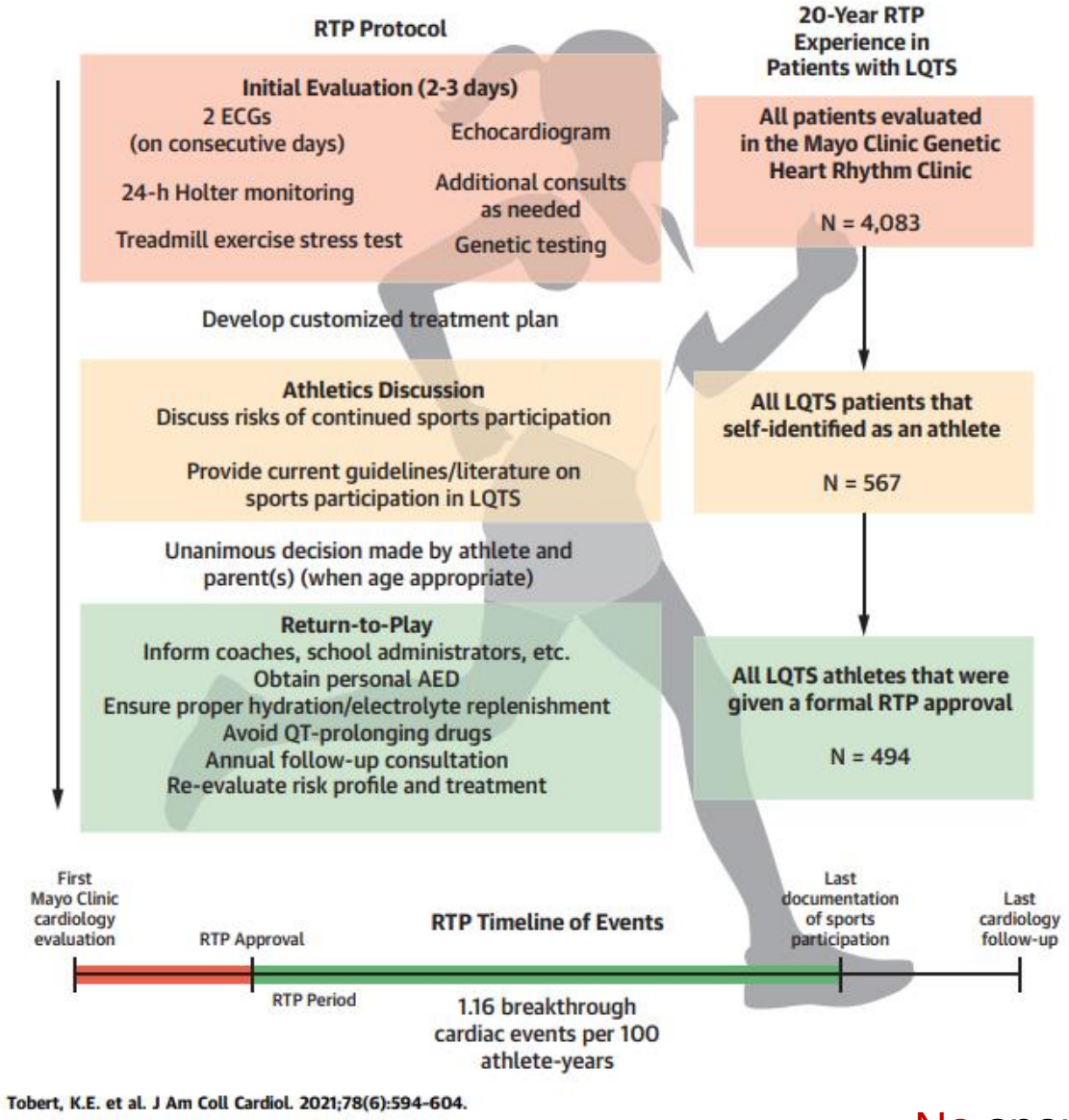
Br J Sports Med. 2013;47:28-33

Current guideline for athletes (2015 ACC/AHA)

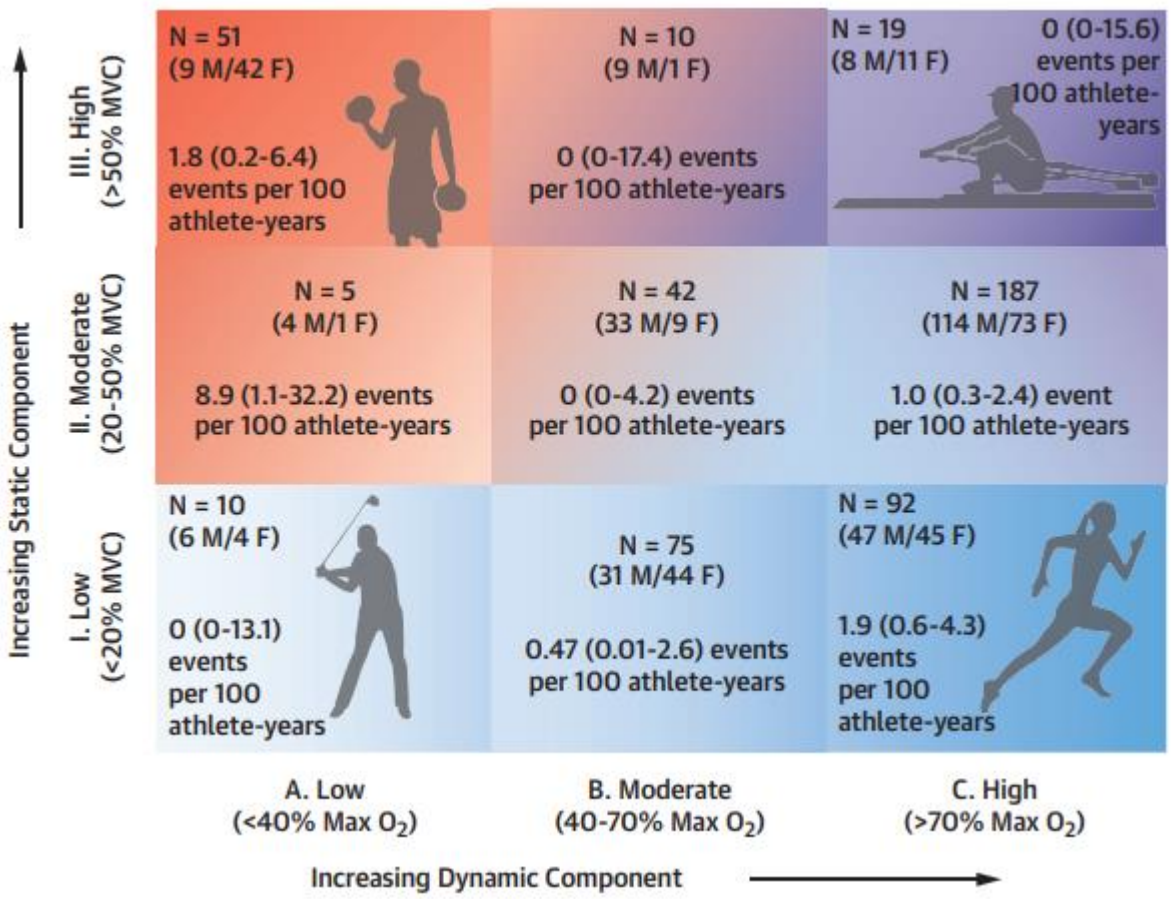
Symptomatic athletes with any suspected or diagnosed cardiac channelopathy be restricted from all competitive sports until comprehensive evaluation completed, the athlete and family well informed, treatment program implemented, asymptomatic on therapy for 3 months **(Class IC)**

Genotype-positive/phenotype-negative LQTS can participate in all competitive sports with appropriate precautionary measures : Avoidance of QT prolonging drug, Electrolyte/hydration replenishment and avoidance of dehydration, avoidance or treatment of hyperthermia, acquisition of personal AED **(Class IIa)**

For symptomatic LQTS or ECG manifest LQTS, competitive sports participation may be considered after institution of treatment and appropriate precautionary measures ; asymptomatic on treatment for at least 3months (Except competitive swimming for LQT1 patient) **(Class IIb)**



Tobert, K.E. et al. J Am Coll Cardiol. 2021;78(6):594-604.



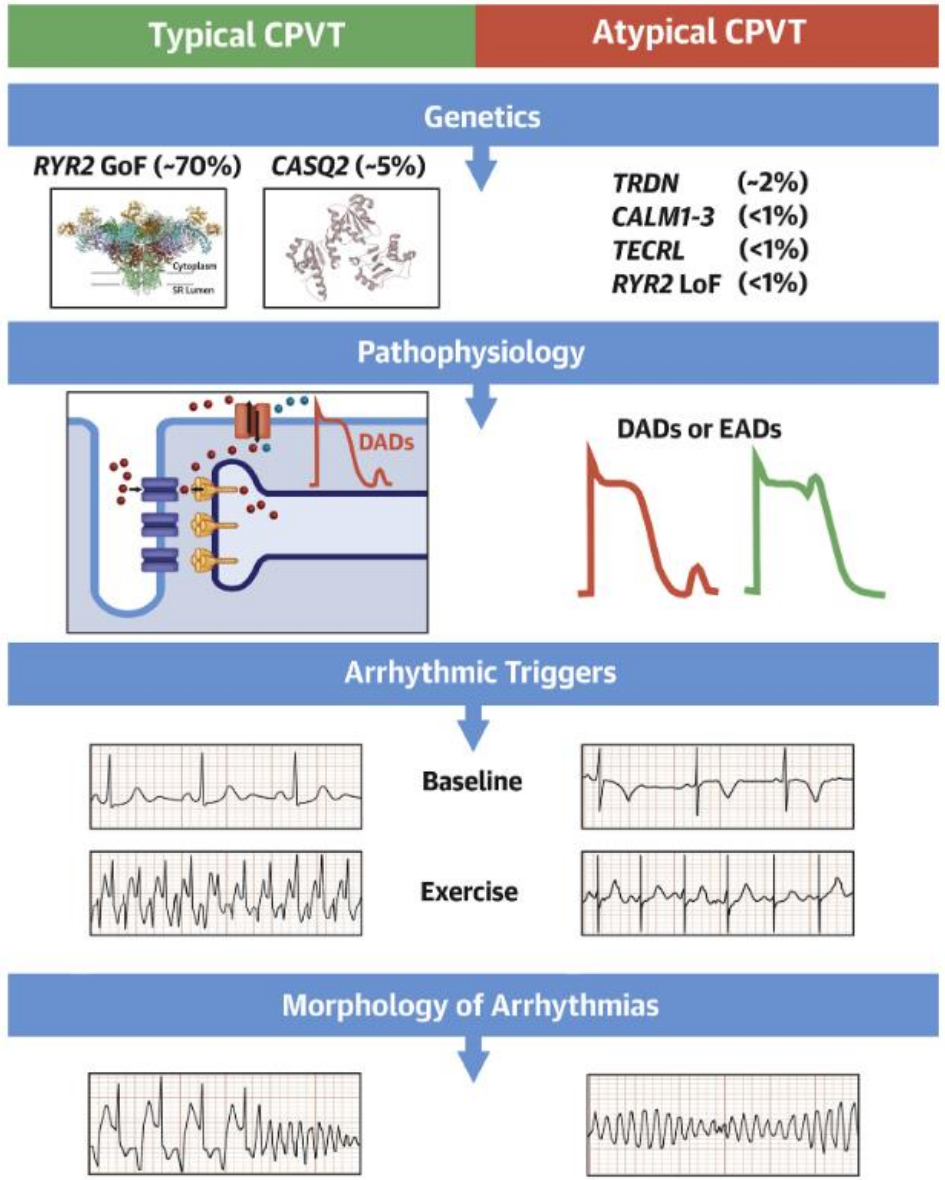
N=494

No sports associated mortality

1.16 nonlethal events per 100 athlete-years

CPVT

- 1:10,000, lack of accurate population data
- Mortality rate up to 31% by 30 years
- 0 to 1.4% of all SCD in athletes
- Physical & emotional stress are triggers



JACC May 2021:2592

Br J Sports Med 2021;55:1196–1203

KHRS 2023

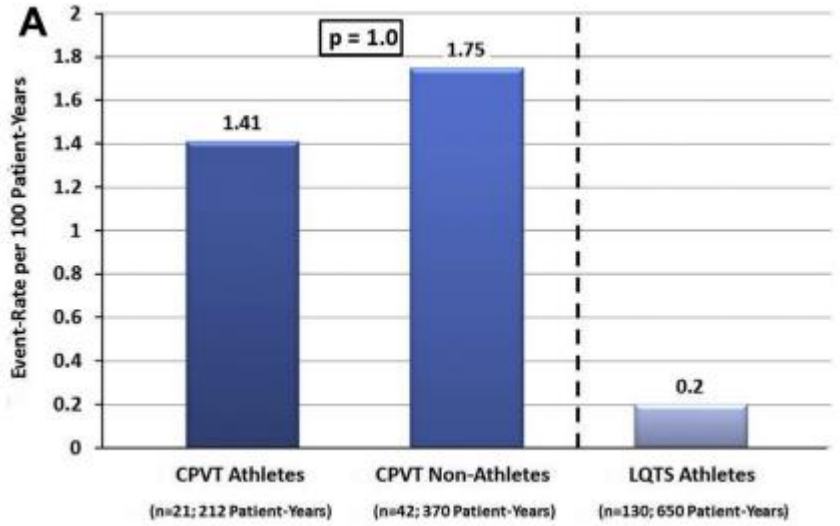
Competitive Sports Participation in Patients With Catecholaminergic Polymorphic Ventricular Tachycardia

A Single Center's Early Experience

Stuart A. Ostby, BS,[a] J. Martijn Bos, MD, PhD,[b,c] Heidi J. Owen, RN,[b] Philip L. Wackel, MD,[b] Bryan C. Cannon, MD,[b] Michael J. Ackerman, MD, PhD[a,b,c,d]

- N=63, 21 Athletes vs 42 non-athletes
- All received CPVT therapy

  (79% BB, 35% Flecainide, 33% ICD, 30% LCSD)
- 16 (76%) vs 18 (43%) experienced CPVT trigger event prior to diagnosis (p=0.02)
- During follow up, 3 (14%) vs 7 (14%) events occurred (p=1.00)
- No event resulted in death



JACC Clin Electrophysiol 2016 Jun;2(3):253-262

Guidelines for athletes

2005 The Bethesda Conference Guidelines

All patients with CPVT should be restricted from competitive sports with the possible exception of minimal contact, class IA category (billiards, bowling, cricket, curling, golf…)

2015 AHA Guidelines

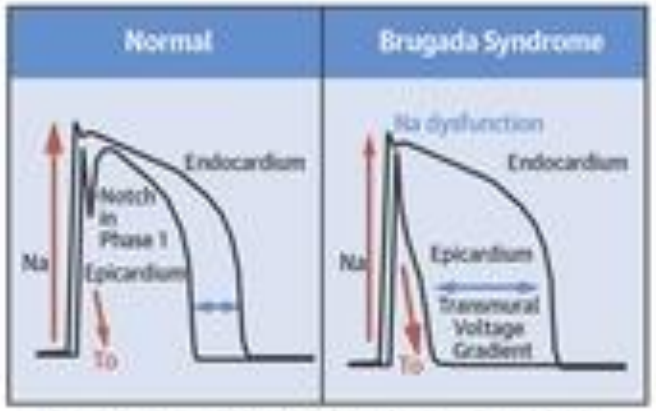
CPVT athletes should be restricted from competitive sports participation if they have exercise-induced PVCs in bigeminy, couplets, or NSVT

2022 ESC Guidelines

Patients with CPVT should avoid competitive sports, strenuous exercise, and stressful environments

BrS

Coved type ST-segment in V1-V2



Loss of function of sodium channels

- 1:2000
- >80% during sleep or rest
- “Brugada-like” RBBB pattern commonly observed in endurance sports athletes
- But, rare case reported SCD in athletes

J Am Coll Cardiol 2018; 72:1046-1059.

Brugada Syndrome, Exercise, and Exercise Testing

Shihab Masrur, MD; Sarfaraz Memon, MD; Paul D. Thompson, MD
Division of Cardiology, Hartford Hospital, Hartford, Connecticut

- 2 large studies and several case reports including 166
- 4 cases of syncope after exercise
- 57% ST elevation during early recovery phase
- No reports of exercise-related sudden death

Clin. Cardiol. 38, 5, 323–326 (2015)

Current guideline for athletes (2015 ACC/AHA)

Symptomatic athletes with any suspected or diagnosed cardiac channelopathy be restricted from all competitive sports until comprehensive evaluation completed, the athlete and family well informed, treatment program implemented, asymptomatic on therapy for 3 months **(Class IC)**

Genotype-positive/phenotype-negative BrS can participate in all competitive sports with appropriate precautionary measures : Avoidance of drug that exacerbate the BrS, Electrolyte/hydration replenishment and avoidance of dehydration, avoidance or treatment of hyperthermia, acquisition of personal AED **(Class IIa)**

For symptomatic or ECG manifest BrS, competitive sports participation may be considered after institution of treatment and appropriate precautionary measures ; asymptomatic on treatment for at least 3months **(Class IIb)**

SQTS

- QTc < 300ms 1:3000 in Japan, lack of population data
- 83% during rest or sleep, 17% during stress or exertion
- Rare case reported SCD in athletes

Clin Cardiol 31(6):270–274

Cardiology 94 (2) (2000) 99–102.

The prevalence and significance of a short QT interval in 18 825 low-risk individuals including athletes

Harshil Dhutia,[1] Aneil Malhotra,[1] Sameer Parpia,[2] Vincent Gabus,[1] Gherardo Finocchiaro,[1] Greg Mellor,[1] Ahmed Merghani,[1] Lynne Millar,[1] Rajay Narain,[1] Nabeel Sheikh,[1] Elijah R Behr,[1] Michael Papadakis,[1] Sanjay Sharma[1]

- Athlete 8939 (47%)
- 0.1% QTc ≤ 320ms, 0.2% QTc ≤ 330ms in athletes
- Athletes shorter QTc than non-athletes
- No reports of syncope or death

Br J Sports Med 2016;50:124–129.

Current guideline for athletes (2015 ACC/AHA)

Symptomatic athletes with any suspected or diagnosed cardiac channelopathy be restricted from all competitive sports until comprehensive evaluation completed, the athlete and family well informed, treatment program implemented, asymptomatic on therapy for 3 months **(Class IC)**

Genotype-positive/phenotype-negative SQTS can participate in all competitive sports with appropriate precautionary measures : Electrolyte/hydration replenishment and avoidance of dehydration, acquisition of personal AED **(Class IIa)**

For symptomatic or ECG manifest SQTS, competitive sports participation may be considered after institution of treatment and appropriate precautionary measures ; asymptomatic on treatment for at least 3months **(Class IIb)**

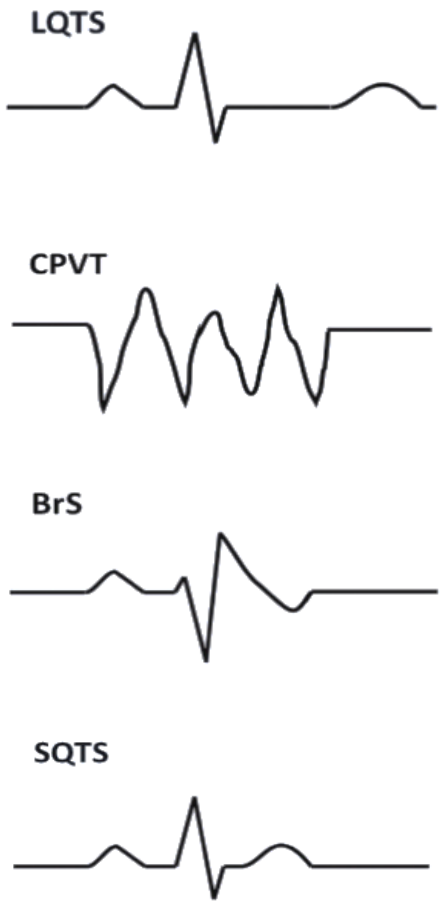
Summary

- **Inherited arrhythmia** accounts for ~ 40% of all SCD in athletes

- The prevalence of inherited arrhythmia in athletes does not seem to be significantly different from that in general population

- Based on recent data, guidelines for restricting sports participation for athletes with inherited arrhythmia have recently been relaxed.

Summary



Suspected, diagnosed athletes	Asymptomatic athletes	Symptomatic or ECG manifest athletes
Restricted all competitive sports until • Evaluation • Education • Asymptomatic on therapy for 3 months	**Consider all competitive sports participation with** Avoid QT prolonging drugs for **LQTS** Avoid exacerbate drug of **BrS** Avoid dehydration and electrolyte imbalance for **All** Avoid hyperthermia for **LQTS** or **BrS** Acquisition of personal AED for **All**	Consider after asymptomatic on 3months treatment (except swimming in LQT1) Not recommend for patients with exercise induced PVC bigeminy, cuplets, NSVT Consider after asymptomatic on 3months treatment Consider after asymptomatic on 3months treatment