Sudden Cardiac Arrest in the Young

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Ochsner Medical Center Hospital for Children

Objectives

- Discuss incidence and prevalence of Sudden Cardiac Arrest (SCA)
- Review causes of SCA
- Describe measures to screen for and prevent SCA in the young
- Provide resources for healthcare providers caring for patients with history or risk of SCA

Disclosures

- No disclosures

What is Sudden Cardiac Arrest?

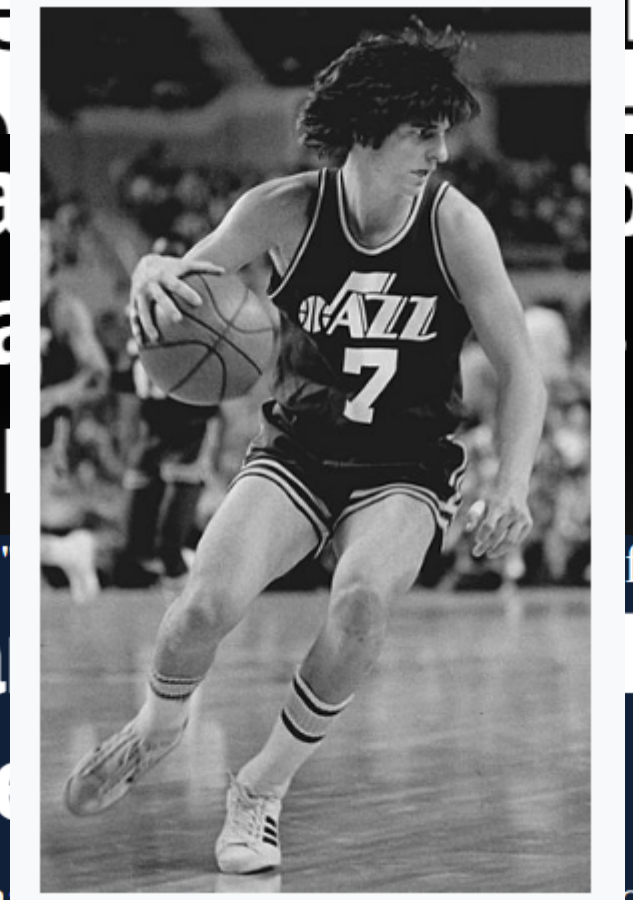
- SCA is a condition in which the heart abruptly stops providing cardiac output
- Most commonly due to ventricular fibrillation or Torsades de Pointe, but can also be due to bradycardic conditions such as asystole, sinus node arrest, or atrioventricular block

Definitions and Acronyms

- SCA that results in death is **sudden cardiac death, or SCD**
- Heritable syndromes that result in SCA or SCD are referred to as **sudden arrhythmic death syndromes (SADS)**
- Sudden death of unclear cause is referred to **as sudden unexplained death** (SUD, or SUDi if under 1 year old)

Wally Pontiff re[illegible] LSU folk hero at 10-yea[illegible] death

Bronny Ja[illegible] ongenital heart defect tha[illegible] cardiac arrest, a spokespe[illegible]



Socc[illegible] on-fi[illegible]

Eriksen, a 29-year-old midfielder, [illegible] for further examination," the

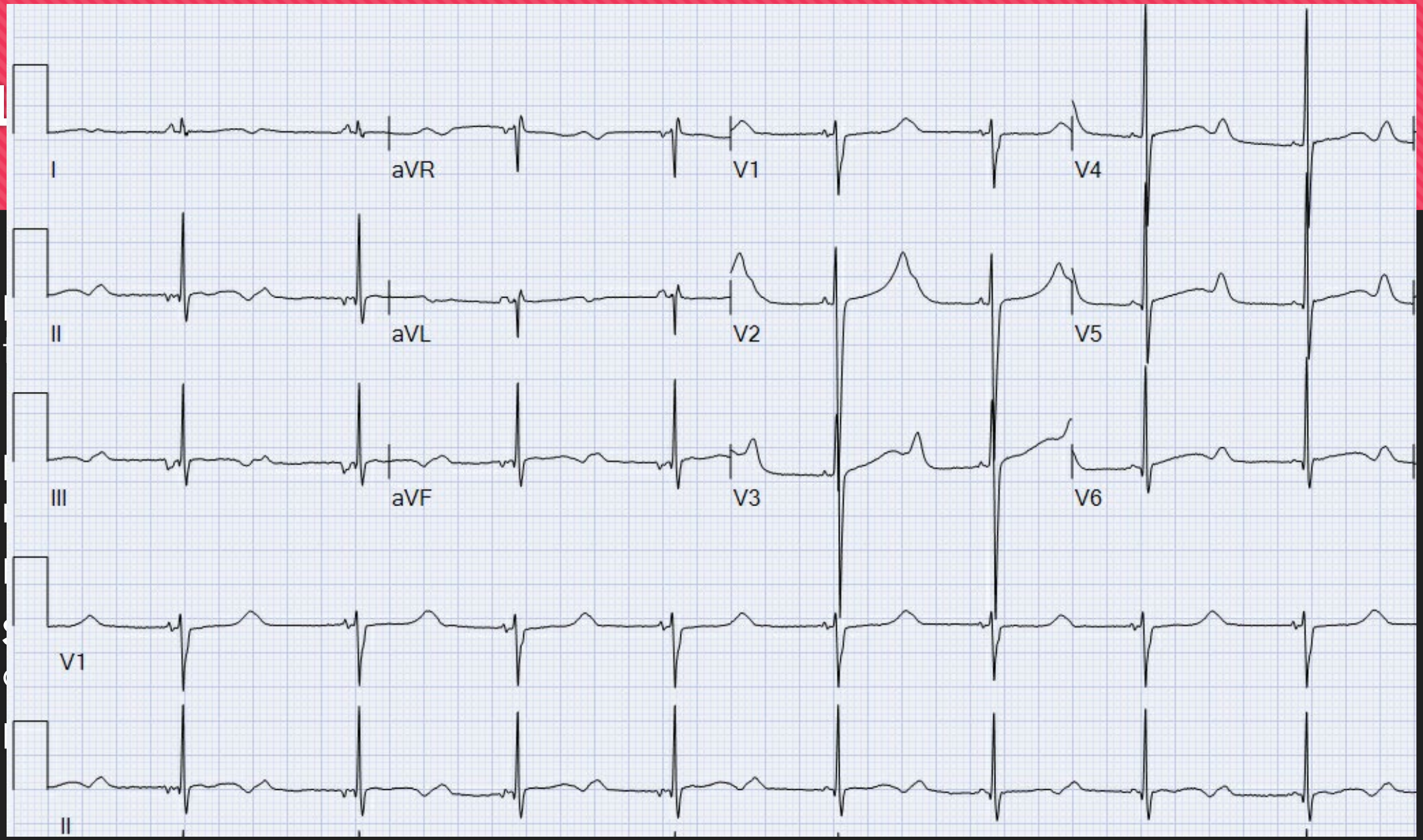
Buffalo Bills' Da[illegible] heart was most likely healthy be[illegible]

The right hit at the wrong time may have triggered a rare phenomenon known as commotio cordis, heart experts suggest.

SCA Statistics

- 210,000 deaths from sudden cardiac arrest per year (all ages and causes) (SADS Foundation)
- 1:200,000 HS athletes die of SCA (JAMA)
- 10-12% of SIDS may be related to Long QT Syndrome
- Long QT Syndrome approximately 3x more common than childhood leukemia (SADS Foundation)

Causes of SCA in the Young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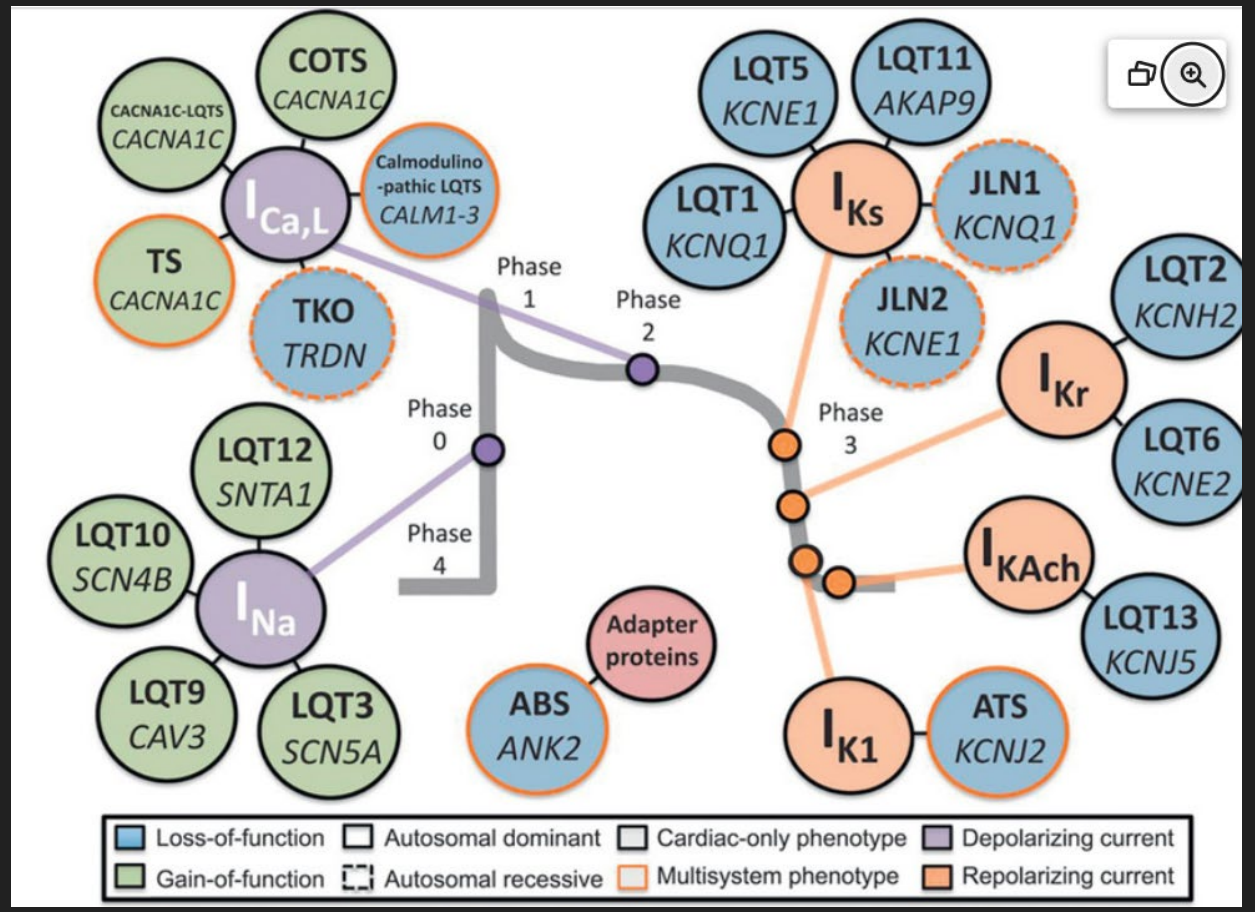
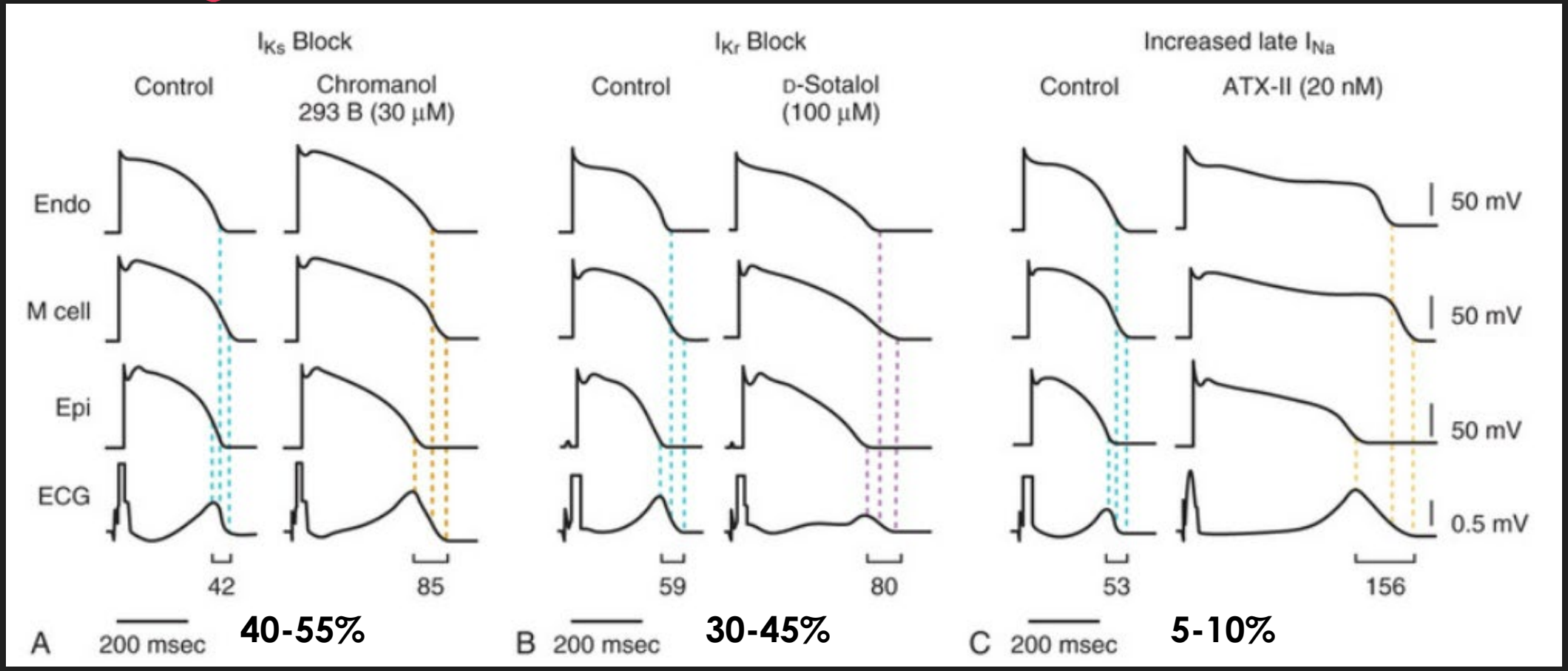
Long QT Syndrome

- Originally named based on clinical features and heritance pattern
  - AD: Romano-Ward Syndrome
  - AR with hearing loss: Jervell-Lange-Nielsen Syndrome
- Now named based on affected gene
  - LQTS 1 through 17; >600 individual mutations so far identified



Most Common Forms of LQTS



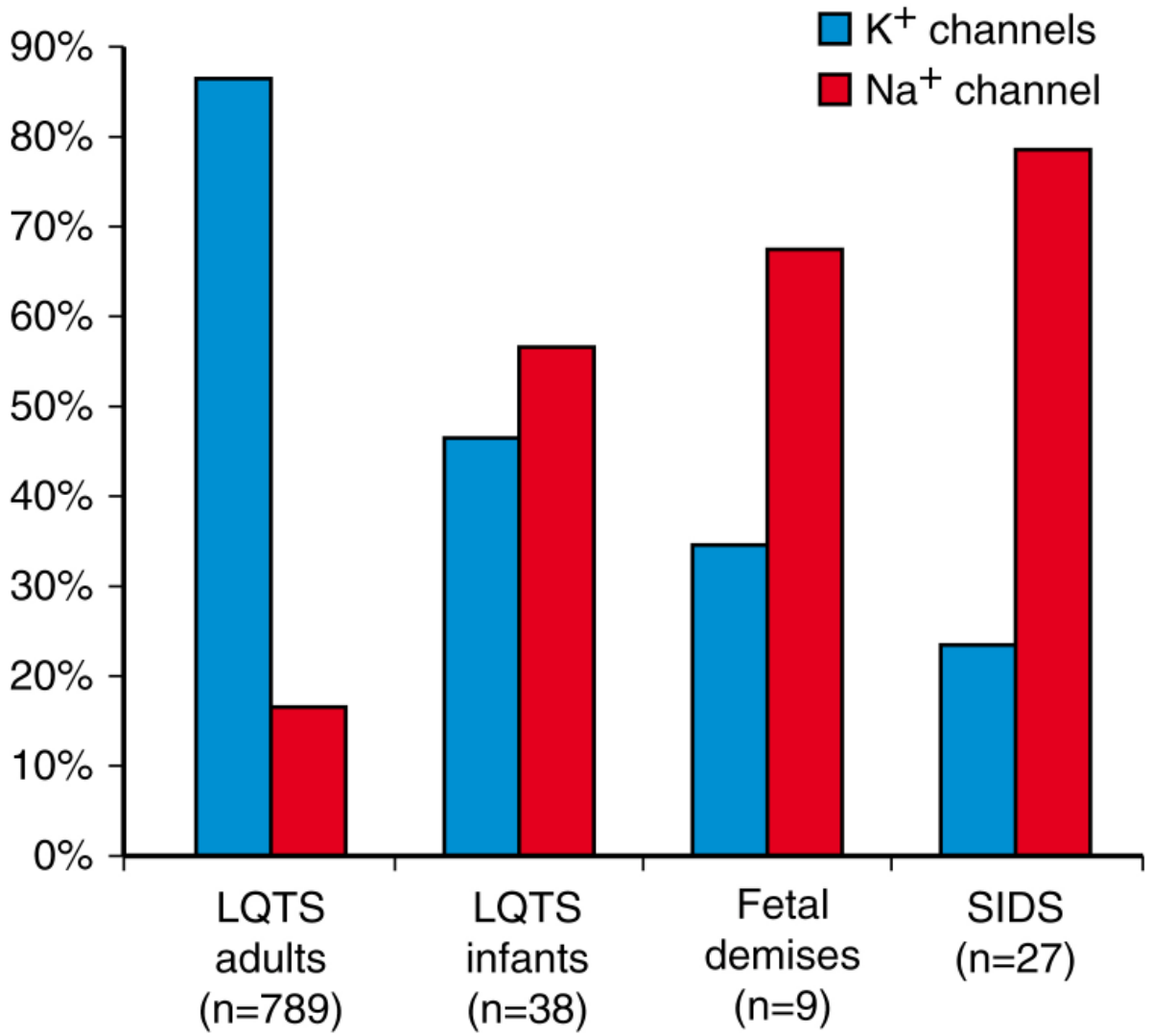


FIGURE 93.1 **Distribution of mutation carriers in the two main potassium (*K*$^+$) channel genes (*KCNQ1* and *KCNH2*; in *blue*) and in the sodium (*Na*$^+$) channel gene (*SCN5A*; in *red*), for four different populations: long QT syndrome (*LQTS*) adult patients, LQTS infants with life-threatening arrhythmias in utero or within the first year of life, intrauterine fetal demises (*IUFDs*), and sudden infant death syndrome (*SIDS*) victims.**
The distribution of variants between the potassium and sodium channel genes is similar in SIDS, IUFD, and LQTS infant populations, all significantly different from the adults ($p < .0005$). From Crotti L, Ghidoni A, Insolia R, Schwartz PJ. The role of the cardiac sodium channel in perinatal early infant mortality. *Card Electrophysiol Clin*. 2014;6:749-759.

Diagnosing LQTS

- Diagnosis made based on clinical and molecular criteria
- Genetic testing is useful for:
  - Confirming suspected disease (75-80% gene identified in LQTS)
  - Screening asymptomatic family members to assess future risk
  - Provide potential prognostic guidance related to clinical LQTS diagnosis

			Points
Electrocardiographic Findings[a]			
A	QTc[b]	≥480 ms	3
		460–479 ms	2
		450–459 (male) ms	1
B	QTc[b] 4th minute of recovery from exercise stress test ≥ 480 ms		1
C	Torsades de pointes[c]		2
D	T wave alternans		1
E	Notched T wave in three leads		1
F	Low heart rate for age[d]		0.5
Clinical History			
A	Syncope[c]	With stress	2
		Without stress	1
B	Congenital deafness		0.5
Family History			
A	Family members with definite LQTS[e]		1
B	Unexplained sudden cardiac death below age 30 among immediate family members[e]		0.5

SCORE:

≤1 point: low probability of LQTS

1.5–3 points: intermediate probability of LQTS

≥3.5 points: high probability

Treatment Specific to LQTS

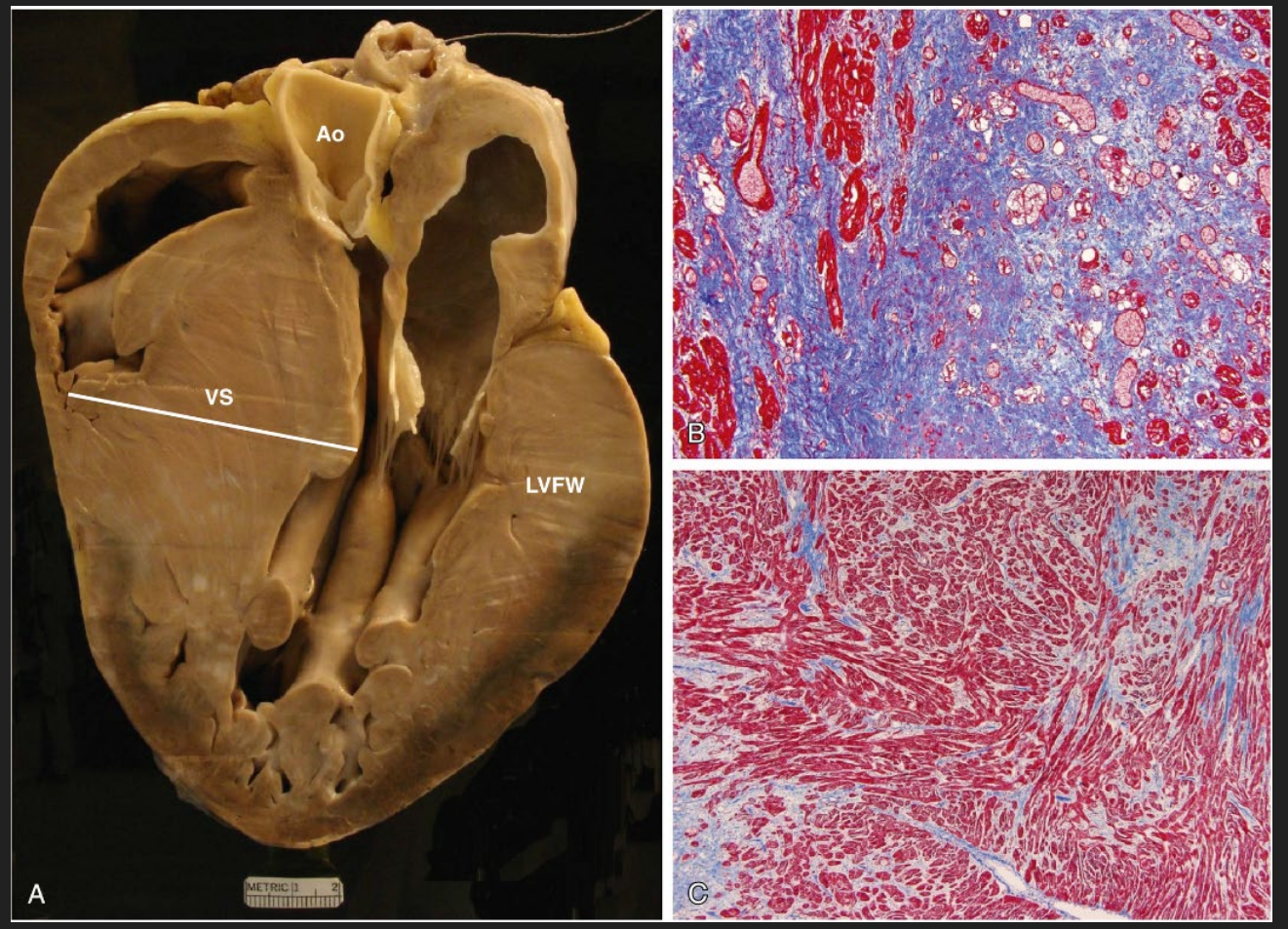
- **Beta-blockade**
  - Nadolol (1-1.5 mg/kg/day) or propranolol (3-4 mg/kg/day) specifically
  - Greatly reduces arrhythmia risk; compliance is crucial
- Left Cardiac Sympathetic Denervation, Pacing, and ICD implant may be needed; based on patient characteristics and tolerance of therapy

Treatment Specific to LQTS

- Asymptomatic carriers ("gene positive, phenotype negative") have lower risk of ventricular arrhythmias (VA's), but beta-blockade and cardiology follow up recommended
- Exercise restriction is controversial
  - Most agree with some form of competitive sport limitation for LQT1, but for other types with less adrenergic-related arrhythmias, joint decision making is generally advised.

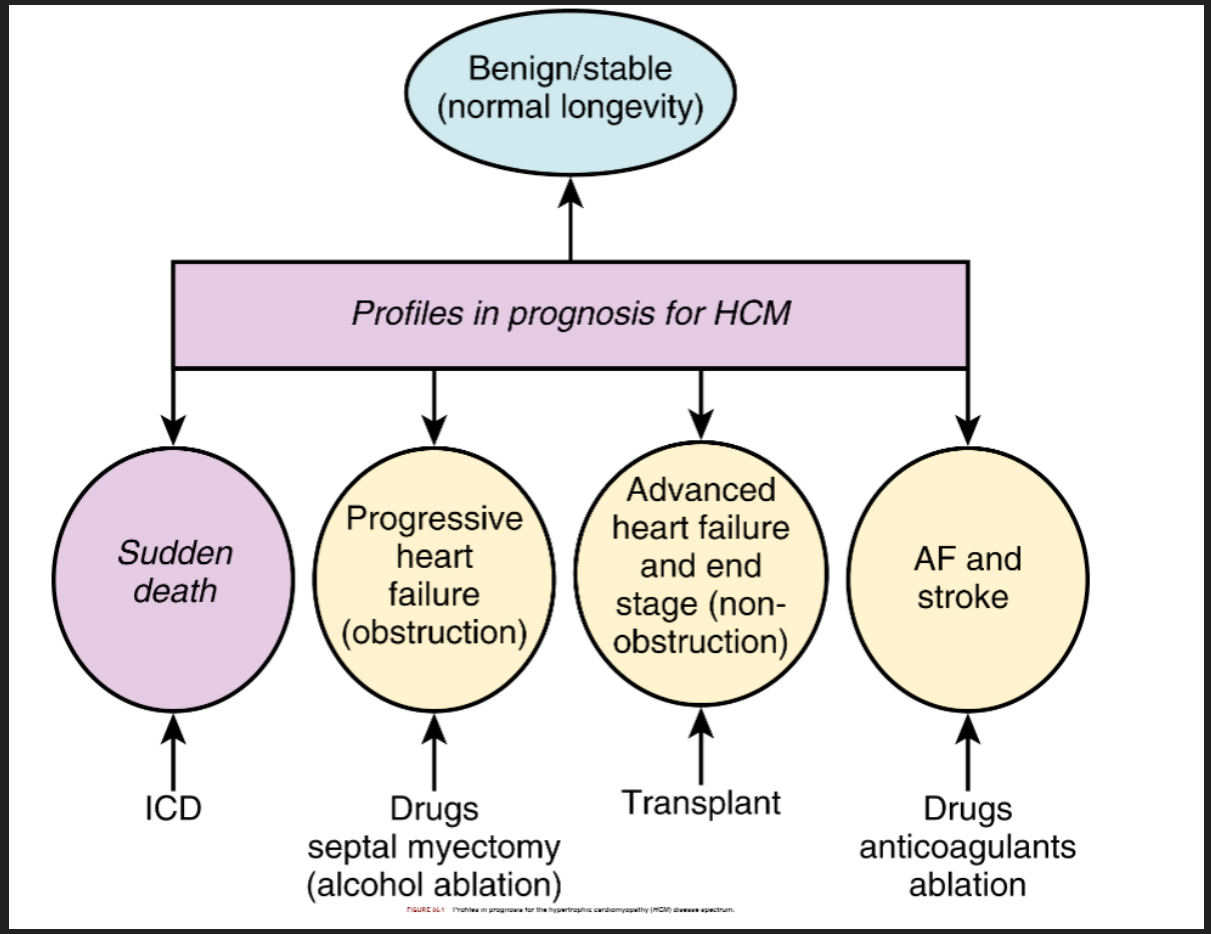
Hypertrophic Cardiomyopathy (HCM)

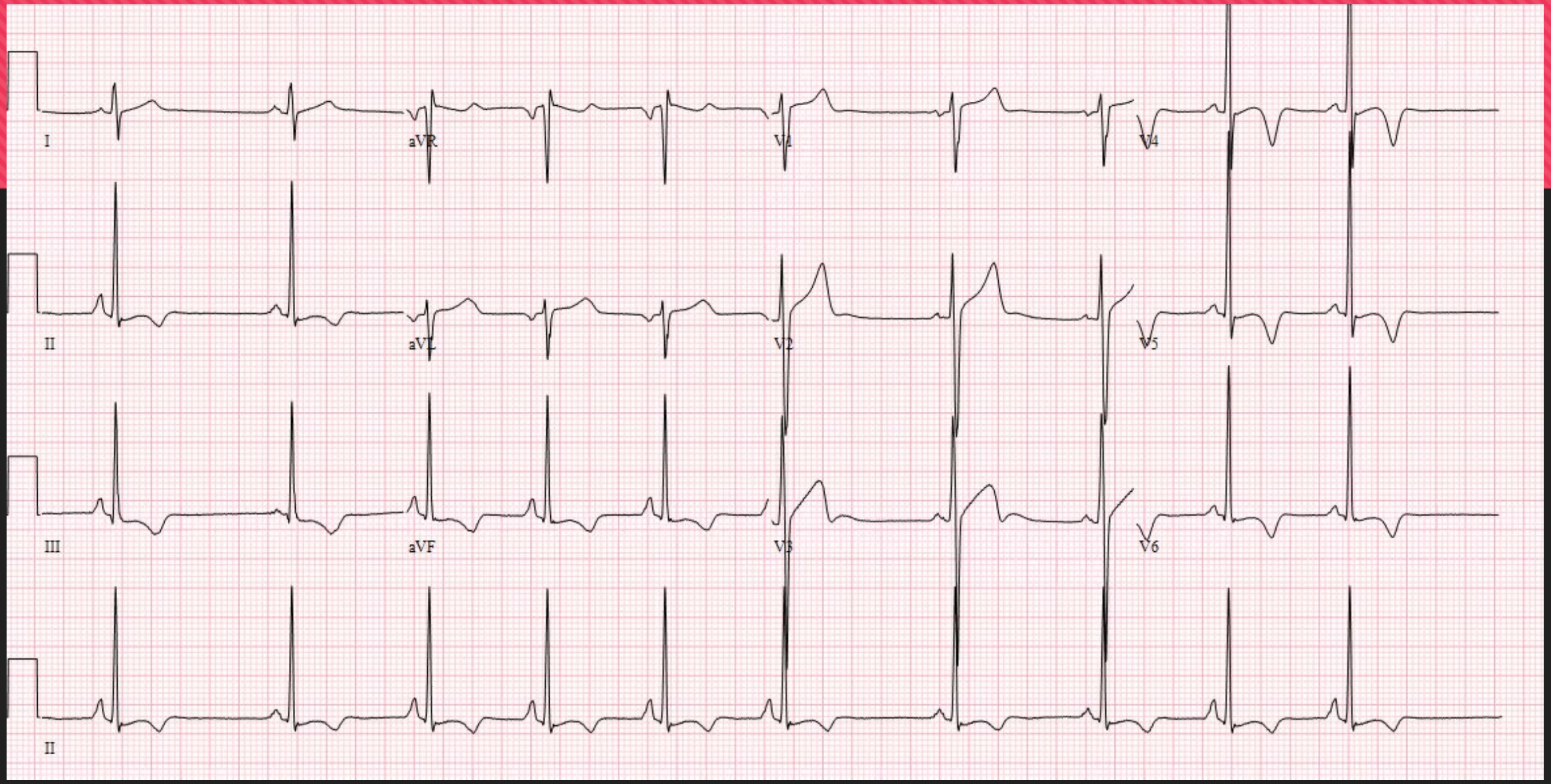
- Genetic disease effecting cardiac myocytes, resulting in "extensive areas of myocyte disarray" resulting in a "chaotic architecture."
  - This results in areas of myocardial fibrosis and scarring, which is proarrhyhtmic.
  - That said, the triggers for lethal VA's are poorly understood



Hypertrophic Cardiomyopathy

- Relatively high prevalence in the population, **estimates 1:500 to 1:200 individuals**
- **Most common cause of SCD in the young**, including competitive athletes
  - HOWEVER, HCM-related SCA events occur in a small minority of patients (~5%); other adverse cardiac complications more common (AF, heart failure)
- SCD risk difficult to predict, but considered 1-1.5%/year
  - SCD risk skews younger rather than older

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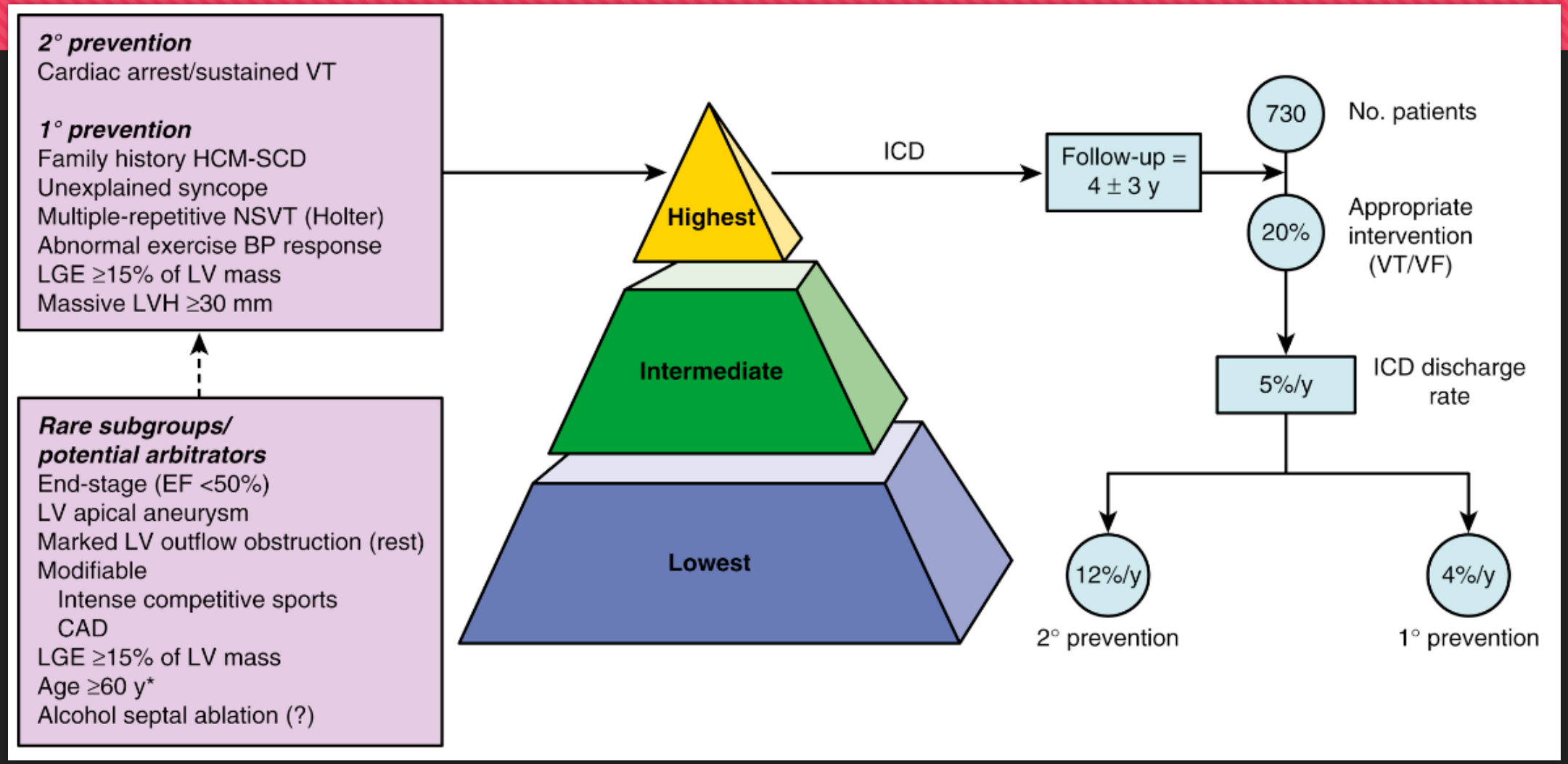
Hypertrophic Cardiomyopathy

- **Autosomal Dominant** genetic heritance pattern; 50% chance of passing risk to offspring
- Because of the volume of information regarding SCA in athletes with HCM, **competitive athletics are generally prohibited**

Treatment of HCM

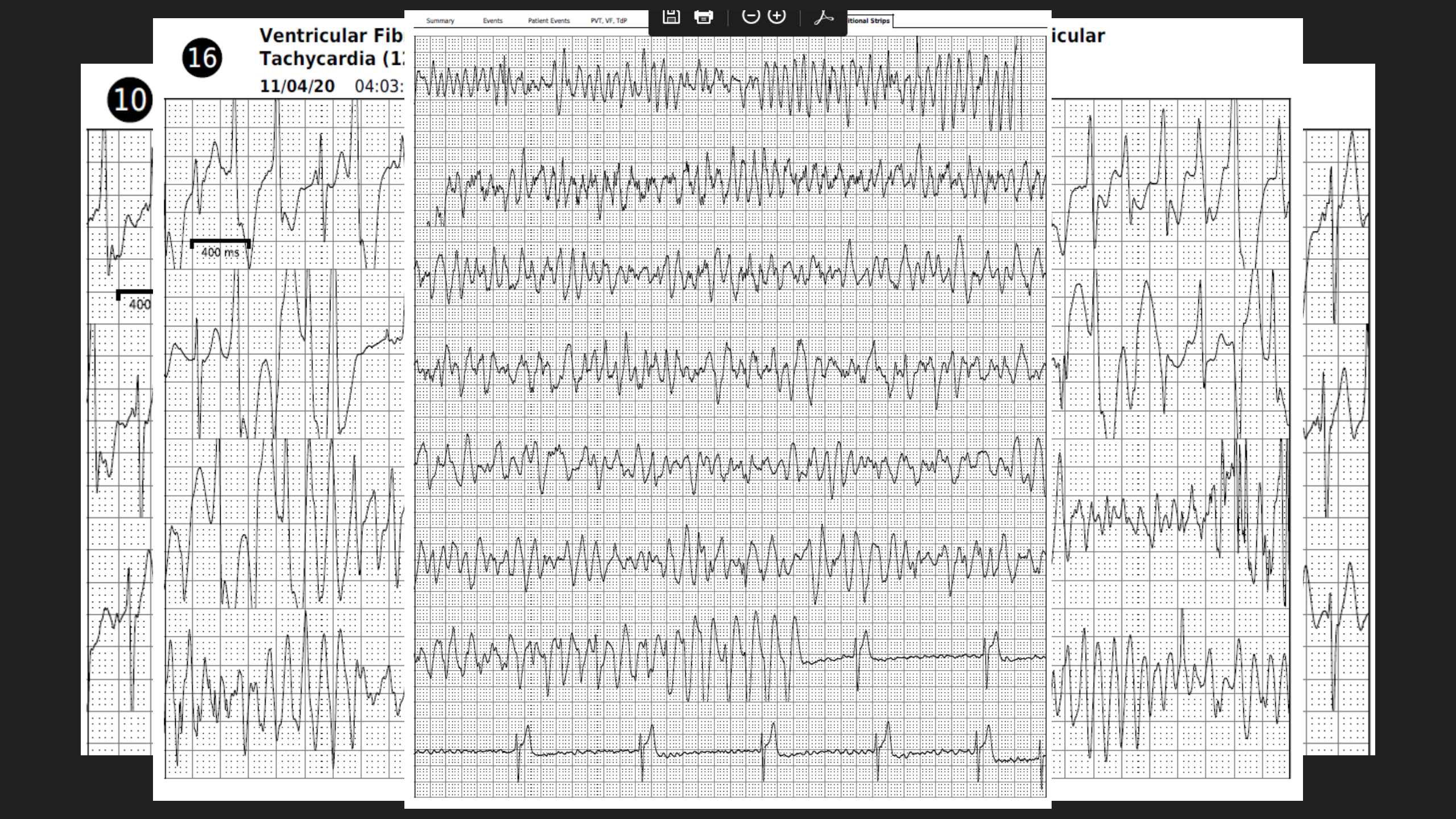
- Overall treatment requires comprehensive approach, with regards to outflow tract obstruction, heart failure, anticoagulation need, and prevention of SCA
- **No drug therapy is effective for preventing SCA**
- ICD implantation is the only measure proven to prevent SCA

Patient Selection for ICD Implant



Catacholaminergic Polymorphic Ventricular Tachycarida (CPVT)

gene	Locus	Inheritance	No. of mutations identified	Phenotype	Frequency
RYR2	1q42-43	Autosomal dominant	>130	CPVT (exon 3 deletion: sinoatrial node, atrioventricular node dysfunction, atrial fibrillation, atrial standstill, dilated cardiomyopathy)	≈60%
CASQ2	1p13.3-p11	Autosomal recessive	12	CPVT	<5%
TRDN	6q22-23	Autosomal recessive	3	CPVT	<5%
Unknown	7p14-22	Autosomal recessive	Not applicable	CPVT, mild QTc interval prolongation	Unknown
KCNJ2	17q24.3	Autosomal dominant		CPVT phenocopy	<5%


Summary
Events
Patient Events
PVT, VF, TdP
16
Ventricular Fib
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400 ms
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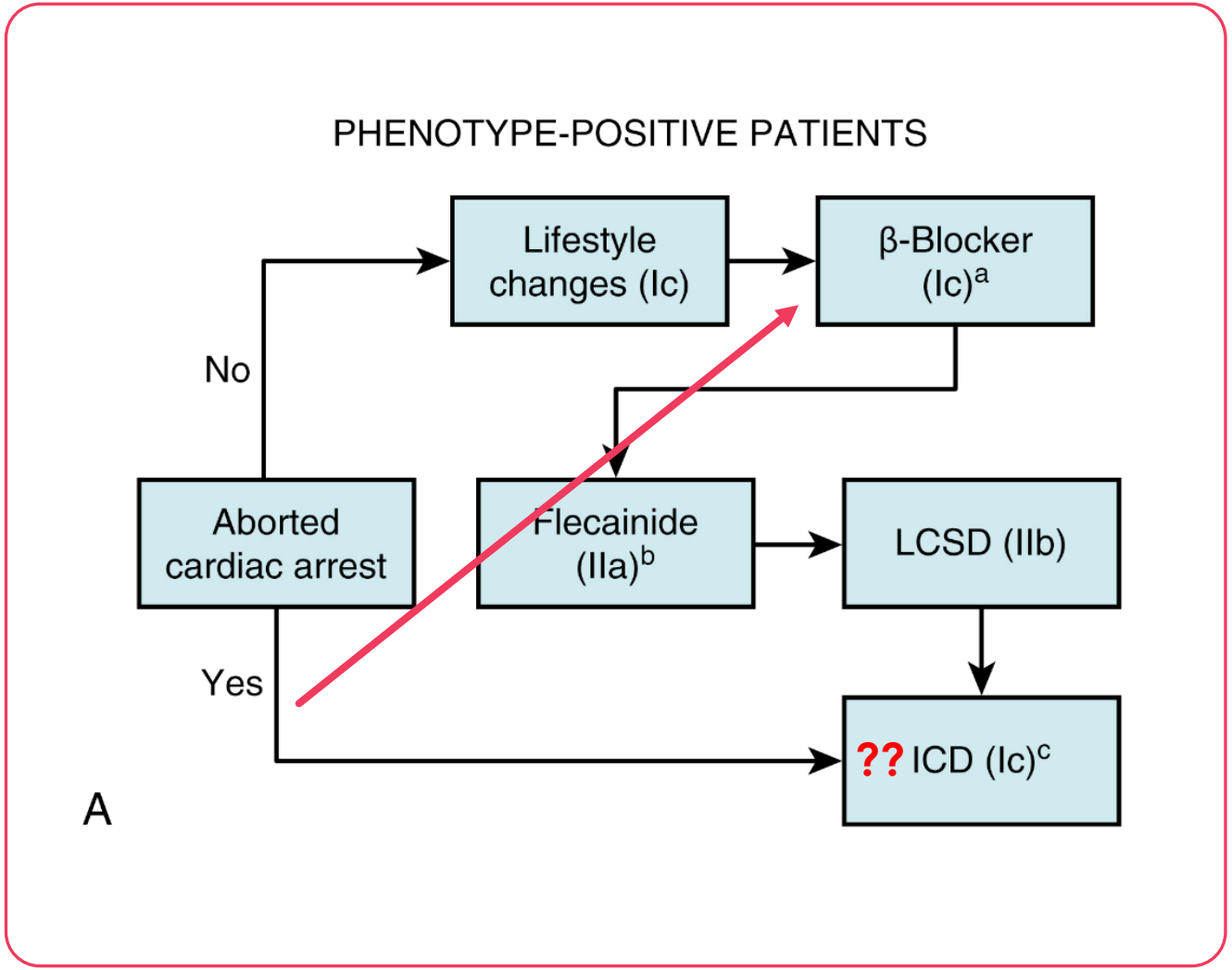
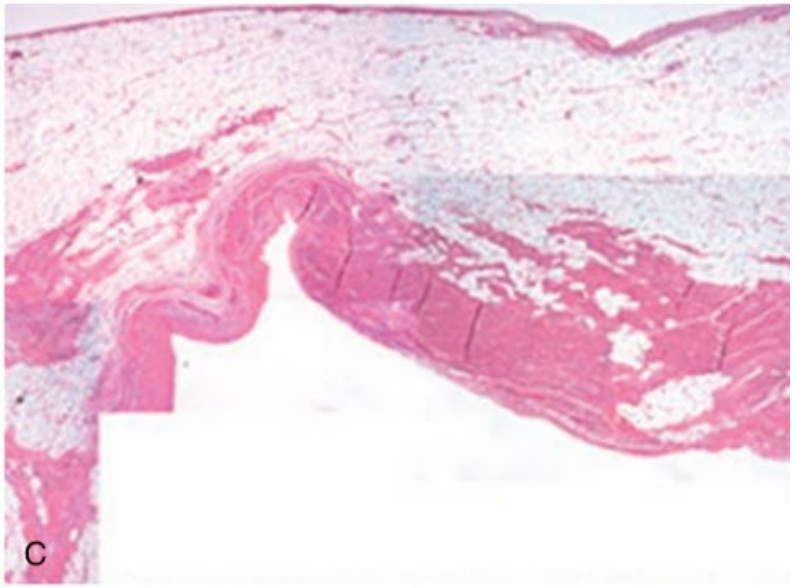
CPVT Treatment

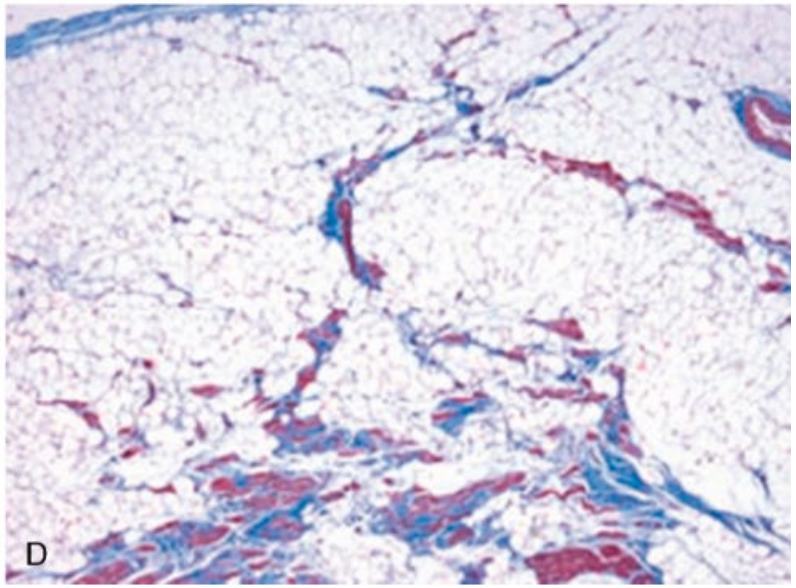
- Beta-blocker is primary therapy
- Competitive athletics prohibited



Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

- Inherited **(AD)** cardiomyopathy characterized by VA's, increased risk of SCD, and abnormalities of the right ventricular structure and function
- Myocyte replacement with fibro-fatty infiltrate due to malfunction of "adherence" proteins





ARVC

- Prevalence 1:5000, presentation generally second to fifth decade of life
- **Syncope,** palpitations, SCD are common presentations
- **Family history**

ARVC

- Clinical diagnosis made based on combination of imaging, electrocardiogram, genetic, histologic, and family history findings
- Treatment based on cardiac function and symptoms, heart failure management and ICD implant as indicated
- **Competitive athletics is prohibited**

Brugada Syndrome

JACC Vol. 20, No. 6
November 15, 1992:1391–6

Right Bundle Branch Block, Persistent ST Segment Elevation and Sudden Cardiac Death: A Distinct Clinical and Electrocardiographic Syndrome

A Multicenter Report

PEDRO BRUGADA, MD, JOSEP BRUGADA, MD*†

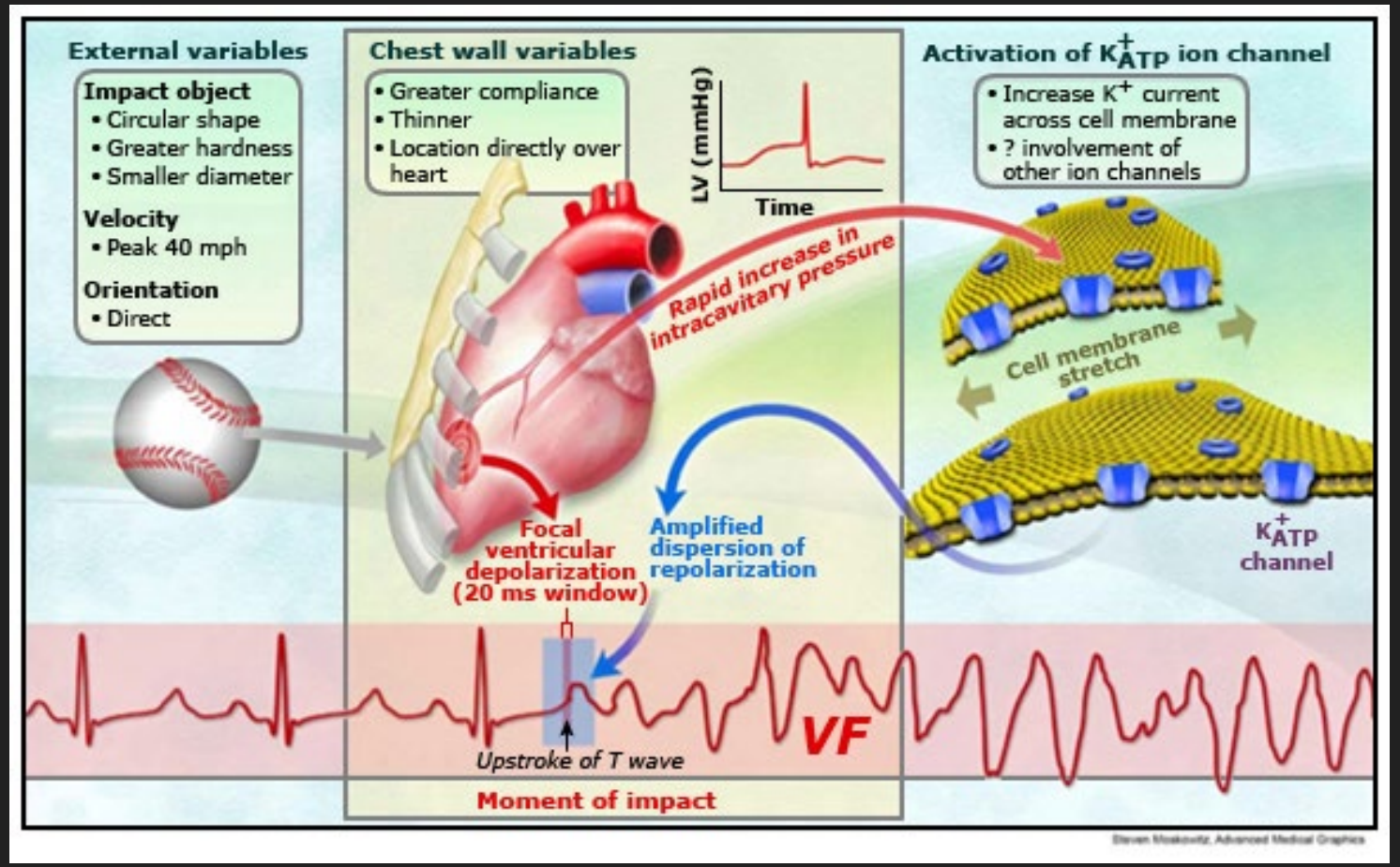
Aalst, Belgium and Barcelona, Spain

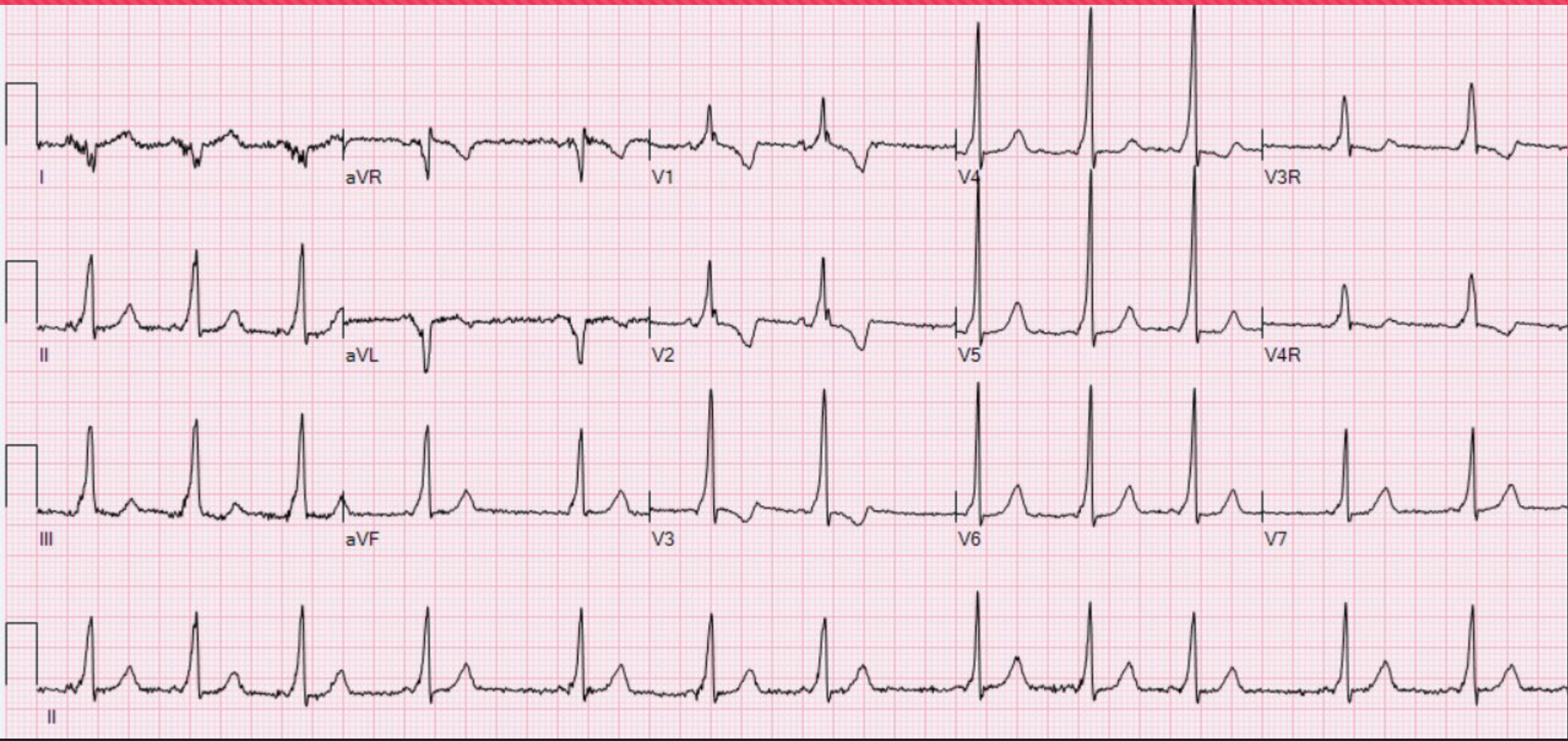
Brugada Syndrome

- Best estimate of prevalence is 1:2000 **worldwide**
  - Varies in different populations (**Highest in SE asia)**
- Brugada pattern can be transient
- **Fever** tends to bring out Brugada pattern, and subsequently increases arrhythmia risk
- **VA's** tend to be at rest; exercise restriction not generally warranted
- Risk stratification difficult to assess

Commotio Cordis

- Sudden cardiac arrest/death due to blunt (nonpenetrating) precordial impact
- Prevalence unknown; 10-20 reported per year
- Mechanism not entirely clear; timing within the cardiac cycle appears to be a significant factor

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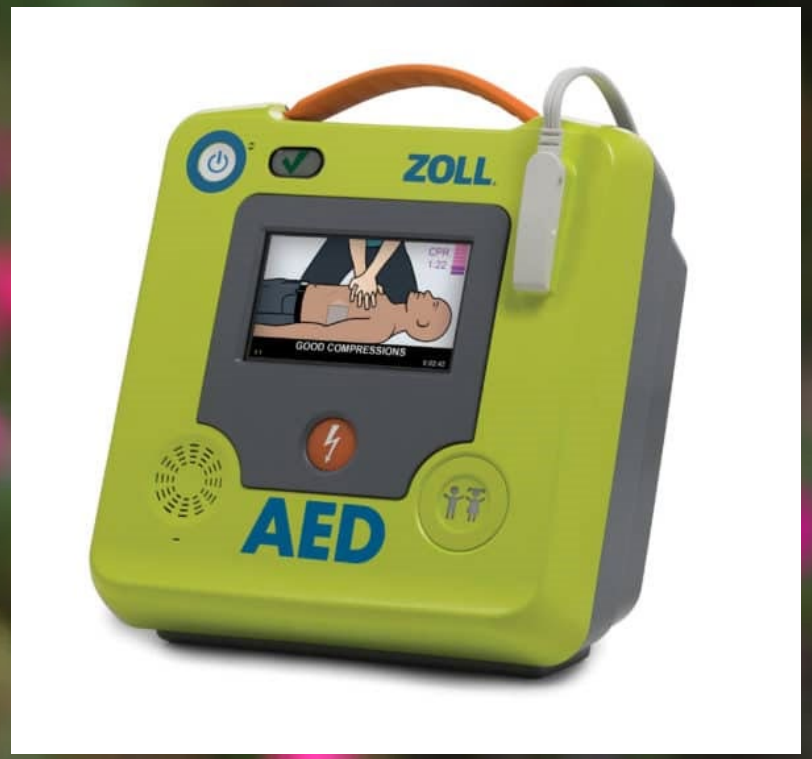
Wolff-Parkinson-White Syndrome

- Most common presentation is SVT
  - Abrupt onset palpitations, lightheadedness, chest pain with tachycardia
- WPW is a **rare** cause of SCA, with risk estimated at 0.13% per year
  - Mechanism generally considered rapidly conducted AF, but this may not account for all cases
- Risk stratification remains elusive; **in most cases, catheter-based electrophysiology study recommended for risk assessment and therapeutic ablation as indicated**

Idiopathic Ventricular Fibrillation (IVF)

- "a resuscitated cardiac arrest victim… with documentation of VF, in whom cardiac, respiratory, metabolic, and toxicological etiologies have been excluded" (2013 HRS/EHRA/APHRS definition)
- Multiple theories and areas of research focusing on new diagnoses that may explain IVF
- ICD implant generally advised
- Genetic evaluation recommended



Arrhythmogenic right ventricular cardiomyopathy in Boxer dogs: the diagnosis as a link to the human disease

ANNINA S. VISCHER,[1,2] DAVID J. CONNOLLY,[3] CAROLINE J. COATS,[1] VIRGINIA LUIS FUENTES,[3] WILLIAM J. MCKENNA,[4] SILVIA CASTELLETTI,[1,5] and ANTONIOS A. PANTAZIS[1,6]

Strategies to Prevent Sudden Cardiac Death

Screening for Risk Factors

Treatment of Sudden Cardiac Arrest

Screening for Risk Factors

- History of **syncope, especially exertional**
- Family history of early "heart attacks" or cardiac arrest (<40-50 years old, especially if multiple)
- Family history of unexplained deaths
  - Single car car wrecks
  - Drowning in people who otherwise swim well
- Palpitations, chest pain are very nonspecific, but lean toward evaluation if there are questions

Screening for Risk Factors

Life threatening cardiac arrhythmia and sudden death during electronic gaming: An international case series and systematic review

Case series and systematic review

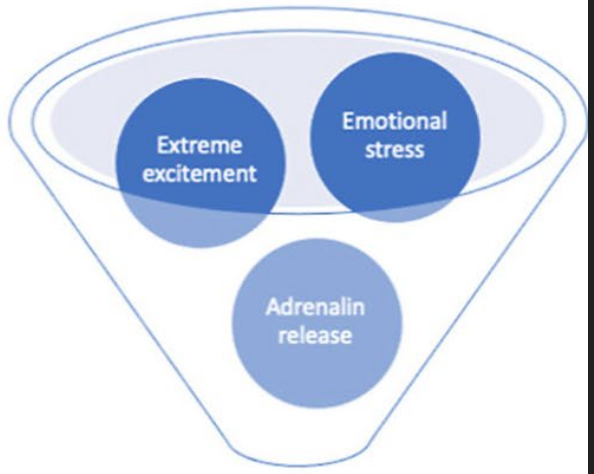
Twenty-two patients with suspected or proven cardiac arrhythmia during electronic gaming

Including six resuscitated cardiac arrests and four sudden deaths

Nineteen pro-arrhythmic diagnoses

CPVT n=10	LQTS n=4	Other n=5

Proposed mechanisms



Risk of arrhythmic events in susceptible individuals

Investigate syncope occurring during electronic gaming

Consider counselling regarding safe participation in electronic gaming after a pro-arrhythmic cardiac diagnosis

Standardized Diagnostic Screening

METHODS

We prospectively studied sudden deaths among athletes and nonathletes (35 years of age or less) in the Veneto region of Italy from 1979 to 1996. The causes of sudden death in both populations were compared, and the pathological findings in the athletes were related to their clinical histories and electrocardiograms. Cardiovascular reasons for disqualification from participation in sports were investigated and follow-up was performed in a consecutive series of 33,735 young athletes who underwent preparticipation screening in Padua, Italy, during the same period.

RESULTS

Of 269 sudden deaths in young people, 49 occurred in competitive athletes (44 male and 5 female athletes; mean [±SD] age, 23±7 years). The most common causes of sudden death in athletes were arrhythmogenic right ventricular cardiomyopathy (22.4 percent), coronary atherosclerosis (18.4 percent), and anomalous origin of a coronary artery (12.2 percent). Hypertrophic cardiomyopathy caused only 1 sudden death among the athletes (2.0 percent) but caused 16 sudden deaths in the nonathletes (7.3 percent). Hypertrophic cardiomyopathy was detected in 22 athletes (0.07 percent) at preparticipation screening and accounted for 3.5 percent of the cardiovascular reasons for disqualification. None of the disqualified athletes with hypertrophic cardiomyopathy died during a mean follow-up period of 8.2±5 years.

CONCLUSIONS

The results show that hypertrophic cardiomyopathy was an uncommon cause of death in these young competitive athletes and suggest that the identification and disqualification of affected athletes at screening before participation in competitive sports may have prevented sudden death.

Not screened (non-athle
(???)

220

SCA:
269

eened:
3,735

neligible for
Sports:
621
(22 HCM)

TABLE 2. CAUSES OF SUDDEN DEATH IN ATHLETES AND NONATHLETES 35 YEARS OF AGE OR LESS IN THE VENETO REGION OF ITALY, 1979 TO 1996.

CAUSE	ATHLETES (N=49)	NONATHLETES (N=220)	TOTAL (N=269)
	number (percent)		
Arrhythmogenic right ventricular cardiomyopathy	11 (22.4)	18 (8.2)*	29 (10.8)
Atherosclerotic coronary artery disease	9 (18.4)	36 (16.4)	45 (16.7)
Anomalous origin of coronary artery	6 (12.2)	1 (0.5)†	7 (2.6)
Disease of conduction system	4 (8.2)	20 (9.1)	24 (8.9)
Mitral-valve prolapse	5 (10.2)	21 (9.5)	26 (9.7)
Hypertrophic cardiomyopathy	1 (2.0)	16 (7.3)	17 (6.3)
Myocarditis	3 (6.1)	19 (8.6)	22 (8.2)
Myocardial bridge	2 (4.1)	5 (2.3)	7 (2.6)
Pulmonary thromboembolism	1 (2.0)	3 (1.4)	4 (1.5)
Dissecting aortic aneurysm	1 (2.0)	11 (5.0)	12 (4.5)
Dilated cardiomyopathy	1 (2.0)	9 (4.1)	10 (3.7)
Other	5 (10.2)	61 (27.7)	66 (24.5)

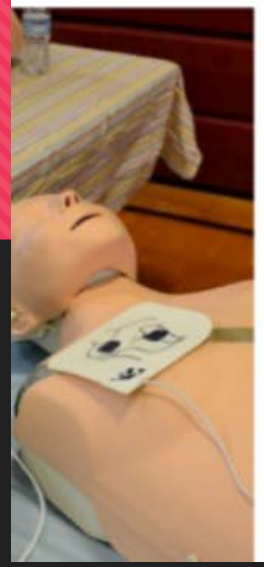
*P=0.008 for the comparison with the athletes.

†P<0.001 for the comparison with the athletes.

Parent Heart Watch
THE NATIONAL VOICE
PROTECTING YOUTH FROM SUDDEN CARDIAC ARREST

DONATE JOIN SPONSOR MEMBER LOGIN

STORIES NEWS CONTACT



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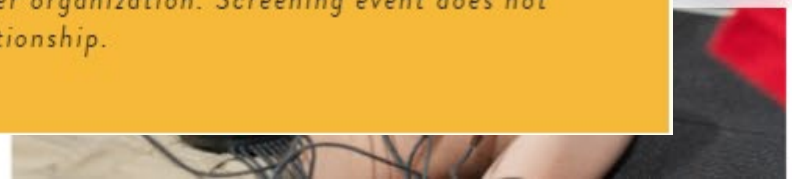
2025

Heart Screening Consists Of:

- Online Registration
- Personal & Family Heart History
- Height, Weight & Blood Pressure Measurements
- 12-lead Electrocardiogram (ECG)
- Auscultation (listening to heart)
- Limited Echocardiogram for those with abnormal results or risk factors
- Results Interpreted by Physician
- Registrants will learn Hands-Only CPR & AED use

Screening protocol may vary per organization. Screening event does not establish a patient/doctor relationship.



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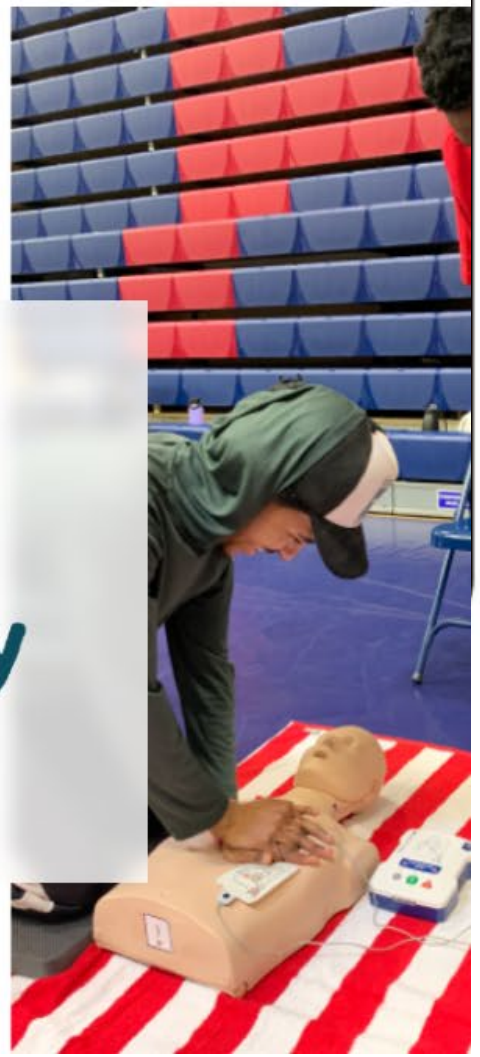

Treatment of Sudden Cardiac Arrest

- Regardless of change in policy or practice, screening will never achieve 100% identification of possible sudden cardiac arrest conditions
- Awareness of and improved response to SCA
  - CPR training
  - AED availability and use

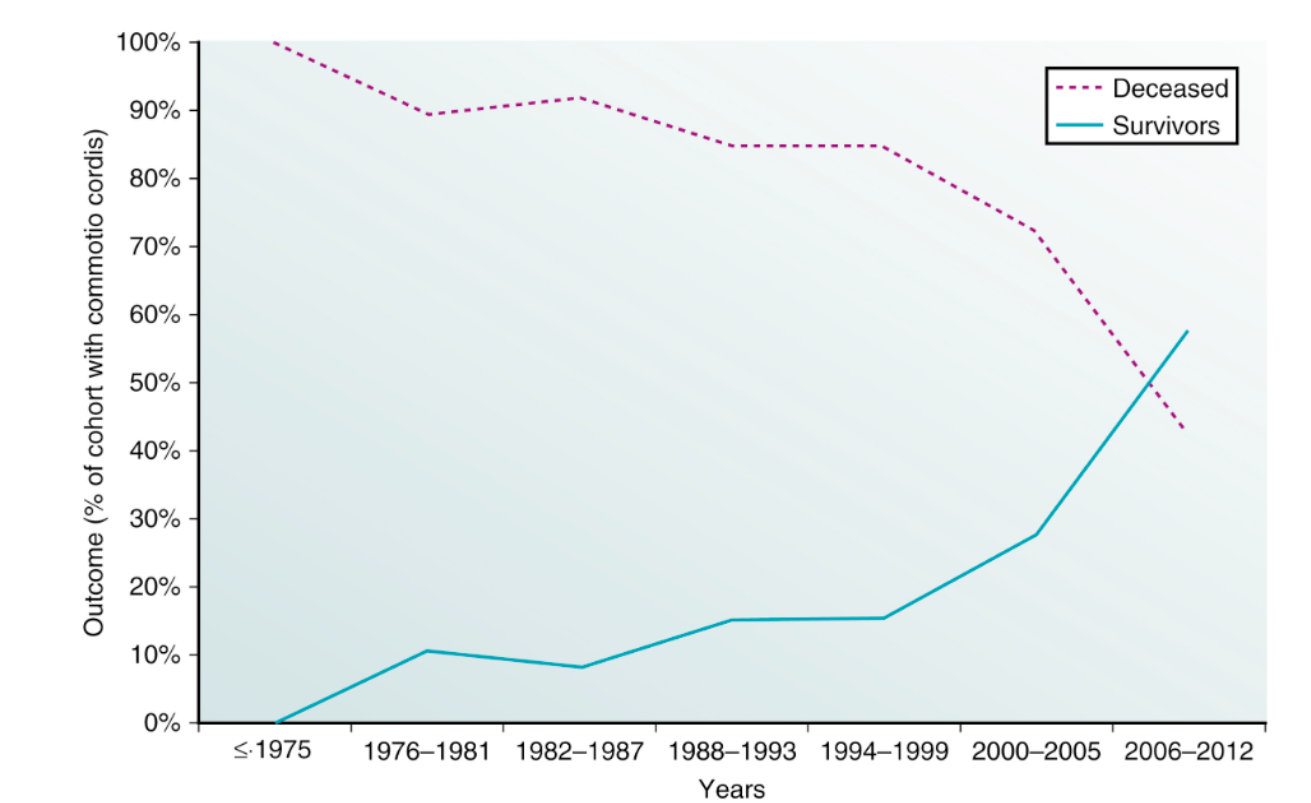

FIGURE 108.9 Resuscitation, originally thought to be rare in commotio, is now routinely reported, likely due to increased recognition that ventricular fibrillation is the cause of sudden collapse after chest wall impact as well as the increased penetration of automated external defibrillator and knowledge of cardiopulmonary resuscitation in the community. From Maron BJ, Haas TS, Ahluwalia A, Garberich RF, Mark Estes NA 3rd, Link MS. Increasing survival rate from commotio cordis. *Heart Rhythm*. 2013;10:219-223.

Education, Awareness, and Advocacy

English

SADS FOUNDATION
SUDDEN ARRHYTHMIA DEATH SYNDROMES

SADS Conditions If You Suspe

MISSISSIPPI LEGISLATURE
2023 Regular Session
To: Public Health and Welfare; Appropriations
By: Senator(s) Hopson, Jackson, McCaughn

Senate Bill 2750

(As Sent to Governor)

AN ACT TO CREATE NEW SECTION 41-60-34, MISSISSIPPI CODE OF 1972, TO ESTABLISH THE AUTOMATED EXTERNAL DEFIBRILLATORS IN PUBLIC AND CHARTER SCHOOLS GRANT PROGRAM TO BE ADMINISTERED BY THE DEPARTMENT OF HEALTH FOR THE PURPOSE OF PROVIDING FUNDS TO ENTITIES TO SUPPLY AEDS IN PUBLIC AND CHARTER SCHOOLS; TO REQUIRE THE DEPARTMENT TO PROMULGATE RULES AND REGULATIONS FOR THE PROGRAM; TO SET CERTAIN REQUIREMENTS OF THE PROGRAM; AND FOR RELATED PURPOSES.

Join us in November 2024 for an opportunity to learn from the country's top medical experts, hear about the latest research, and connect with others.

Resources

- Pediatric and Adult Congenital Electrophysiology Society (PACES)
- SADS.org
- Project ADAM
  - CodeAna.org
- Parent Heart Watch
- Crediblemeds.org
  - LQTS and BrS drug reference

Thank you!

References Not Listed in Slides

- Zipes et al., "*Cardiac Electrophysiology: From Cell to Bedside, 7th Edition.*" 2018, Elsevier