

MUERTE SÚBITA DURANTE LA ACTIVIDAD FÍSICA EN EL NIÑO

Antonio J. Cartón

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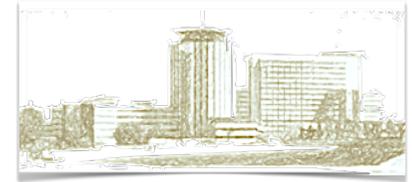
Hospital Universitario La Paz, Madrid

El problema clínico



Durante el ejercicio puede producirse muerte súbita en niños y adolescentes predispuestos.

Sudden Cardiac Arrest during Participation in Competitive Sports



Cameron H. Landry, M.D., Katherine S. Allan, Ph.D.,
 Kim A. Connelly, M.B., B.S., Ph.D., Kris Cunningham, M.D., Ph.D.,
 Laurie J. Morrison, M.D., and Paul Dorian, M.D., for the Rescu Investigators*

Table 4. Causes of Sudden Cardiac Arrest among Competitive and Noncompetitive Athletes, According to Age Group.

Variable	Age Group			
	12–17 yr	18–34 yr	35–45 yr	All
Competitive				
No. of athletes	4	9	3	16
Percent of athletes who survived	50.0	44.4	33.3	43.8
Diagnosis				
Ischemic*	0	0	3	3
Primary arrhythmic	0	6	0	6
Structural†	2	3	0	5
Commotio cordis	2	0	0	2
Noncompetitive				
No. of athletes	9	18	31	58
Percent of athletes who survived	66.7	50.0	35.5	44.8
Diagnosis				
Ischemic*	0	5	21	26
Primary arrhythmic	4	5	0	9
Unknown	2	2	0	4
Structural‡	3	6	8	17
Other§	0	0	2	2

Incidencia: 0.76 casos por 100 000 atletas-años

Comunicadas de hasta 3-4 c/ 10⁵ atletas-años

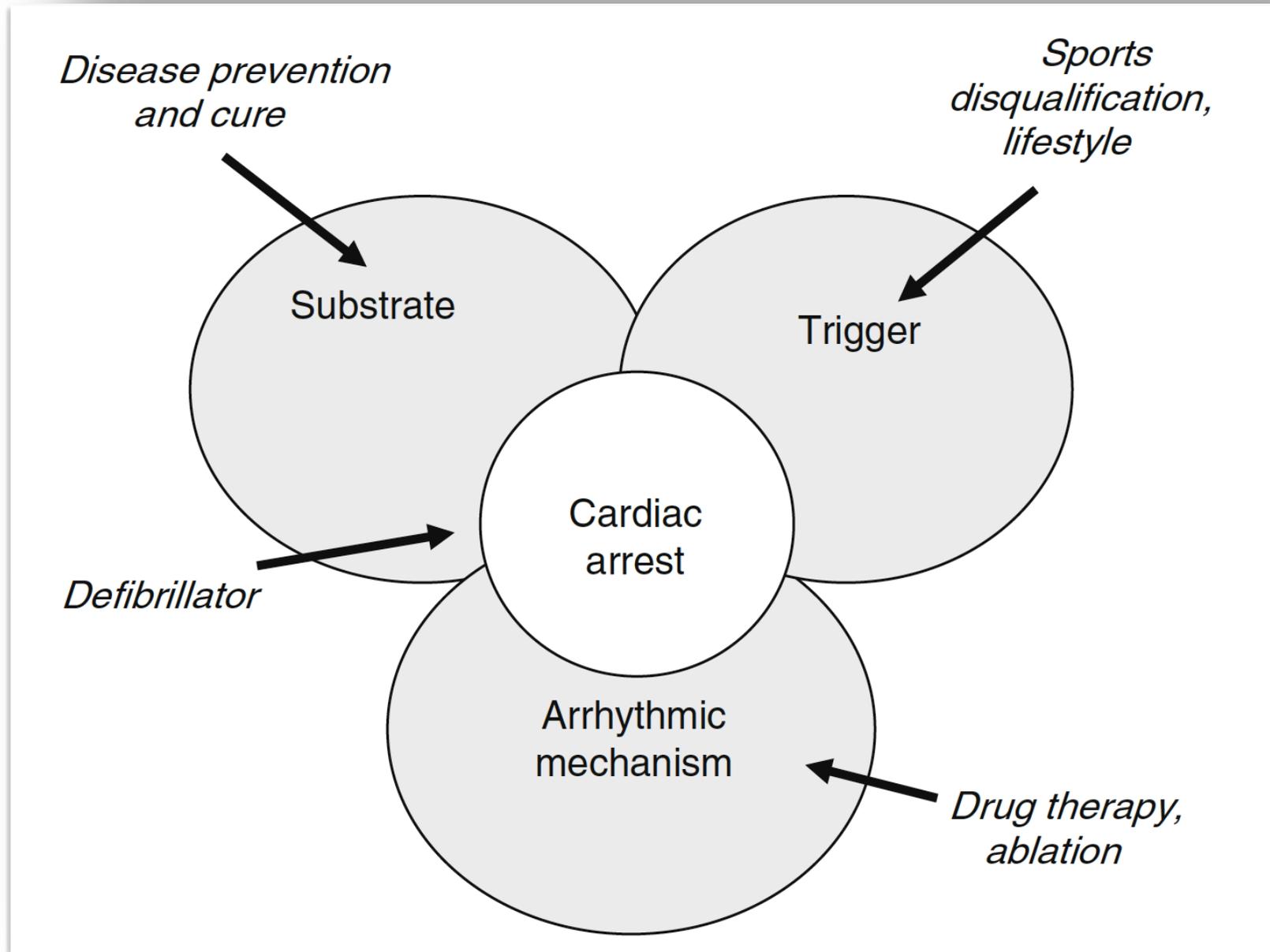
Origen cardiaco arrítmico y no solo...

Landry et al. *N Engl J Med* 2017;377:1943-53.

El problema clínico



El ejercicio es un desencadenante de **ARRITMIAS MORTALES** en niños y adolescentes predispuestos.



PREGUNTAS



- **¿CUÁLES SON LOS INDIVIDUOS PREDISPUUESTOS?
¿QUÉ ENFERMEDADES TIENEN? ¿LOS PODEMOS
IDENTIFICAR? ¿CÓMO?**
- **¿QUÉ ARRITMIAS APARECEN?**
- **¿QUÉ DEPORTES PUEDEN PRECIPITAR LAS
ARRITMIAS? ¿HAY DEPORTES MÁS SEGUROS QUE
OTROS?**

En contexto...

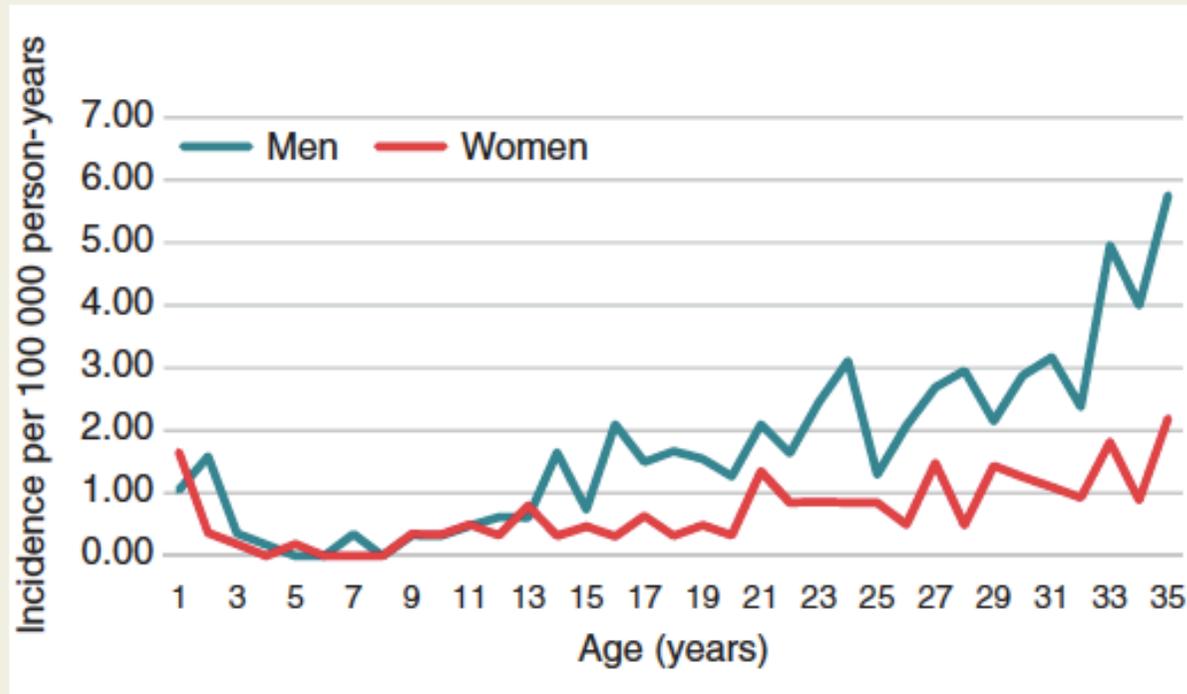
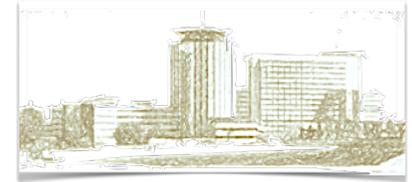
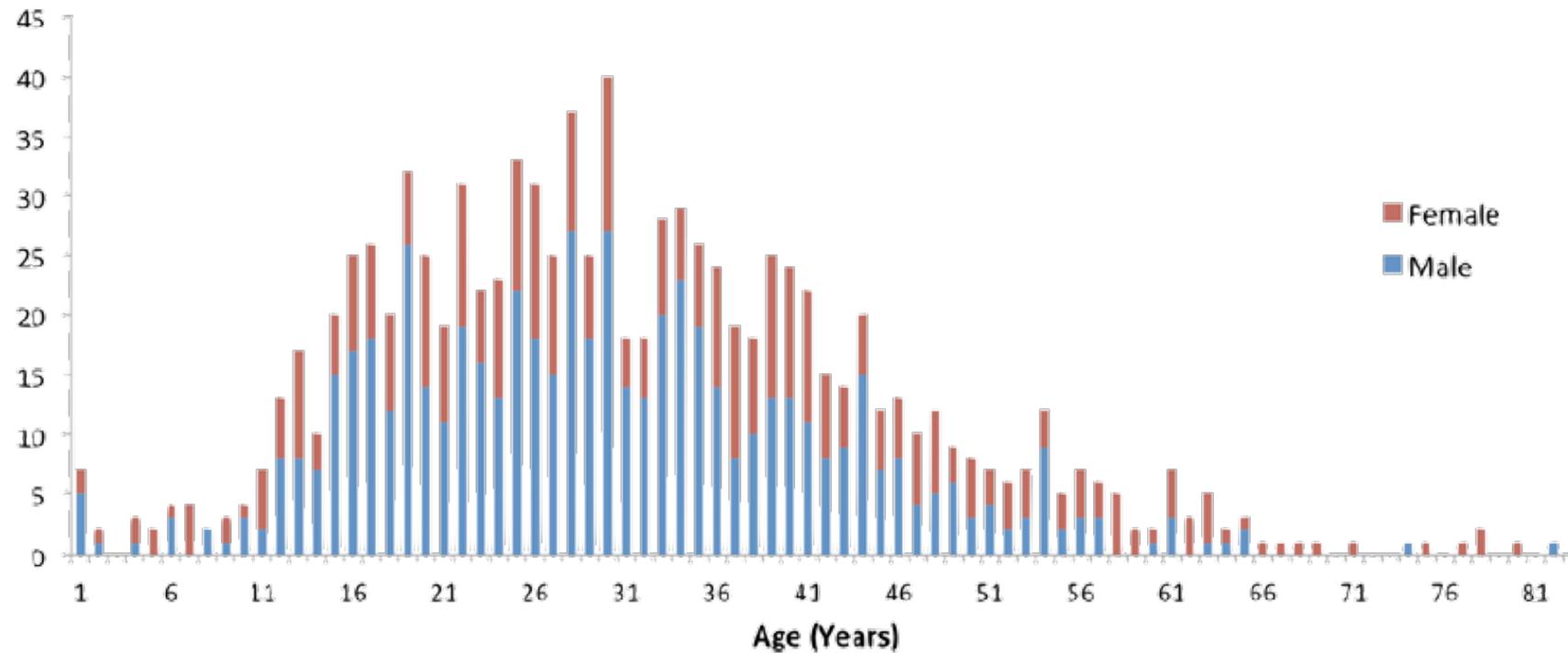
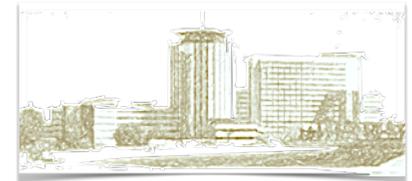


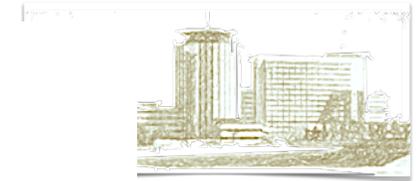
Figure 3 Trends in sudden cardiac death by age and sex in persons aged 1–35 years in Sweden during 2000–10.

Sudden cardiac death among the young in Sweden from 2000 to 2010: an autopsy-based study

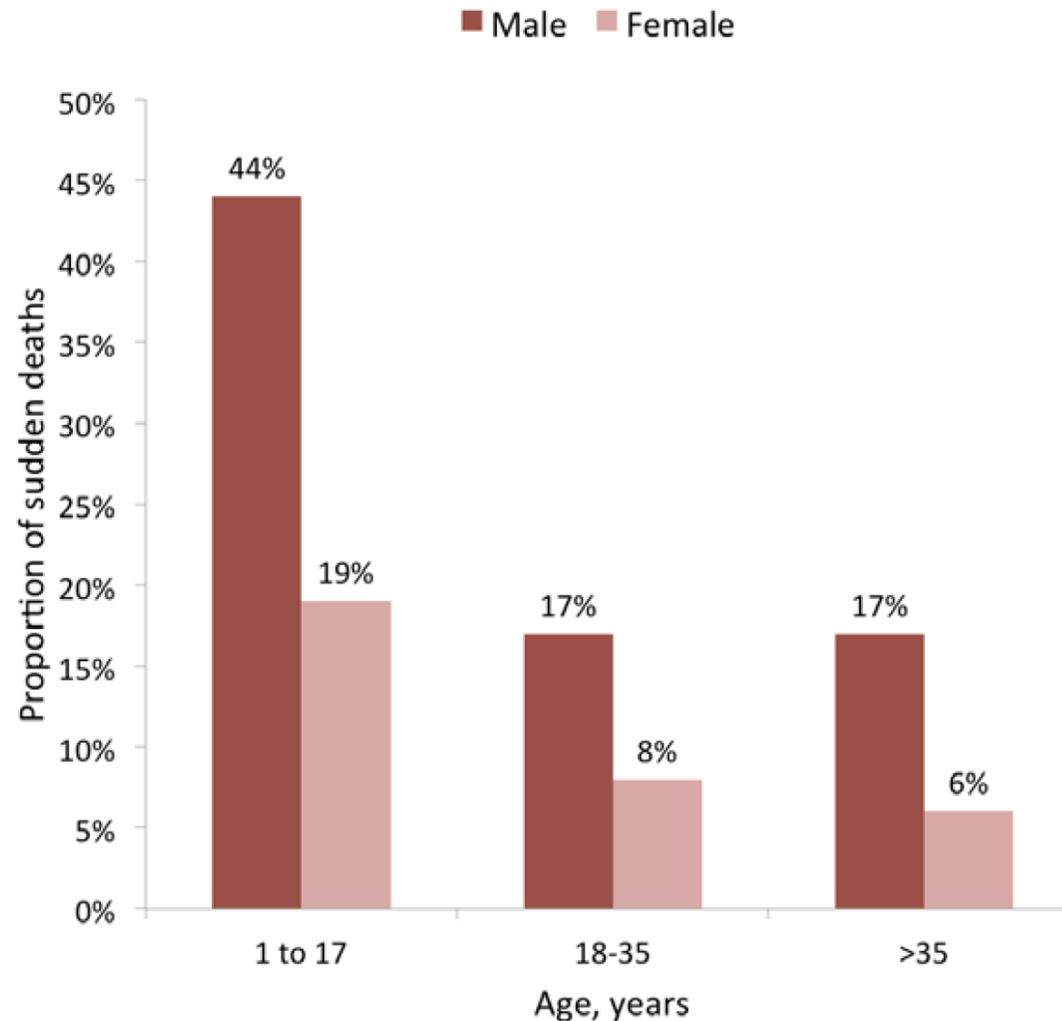


Reino Unido. n=2156

☞ Características demográficas de las víctimas y circunstancias que rodean a la muerte súbita cardiaca con corazón normal

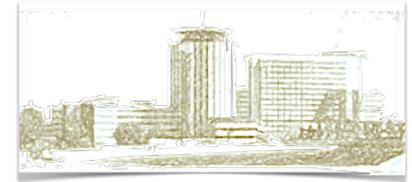


% de muertes durante ejercicio/estrés emocional



☞ La muerte en los síndromes de muerte súbita sucede más frecuentemente en REPOSO o en el SUEÑO.

☞ La muerte durante el EJERCICIO o el stress es más frecuente en varones y menores de 18 años.

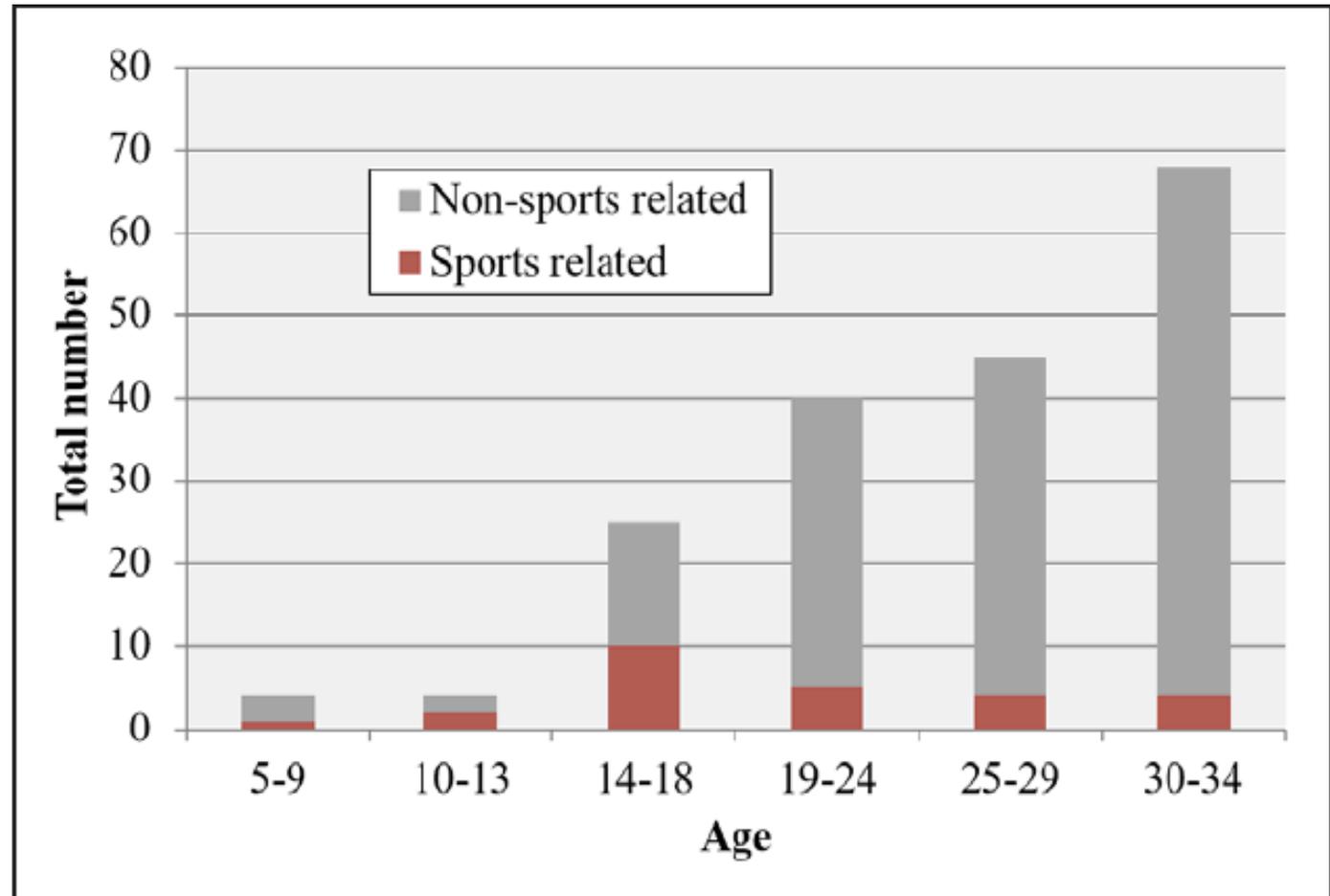


3775 SCAs

5-35 años

2002-2015

Portland (1M hab.)



El deporte fue un desencadenante MINORITARIO de muerte súbita de origen cardiaco (14%)



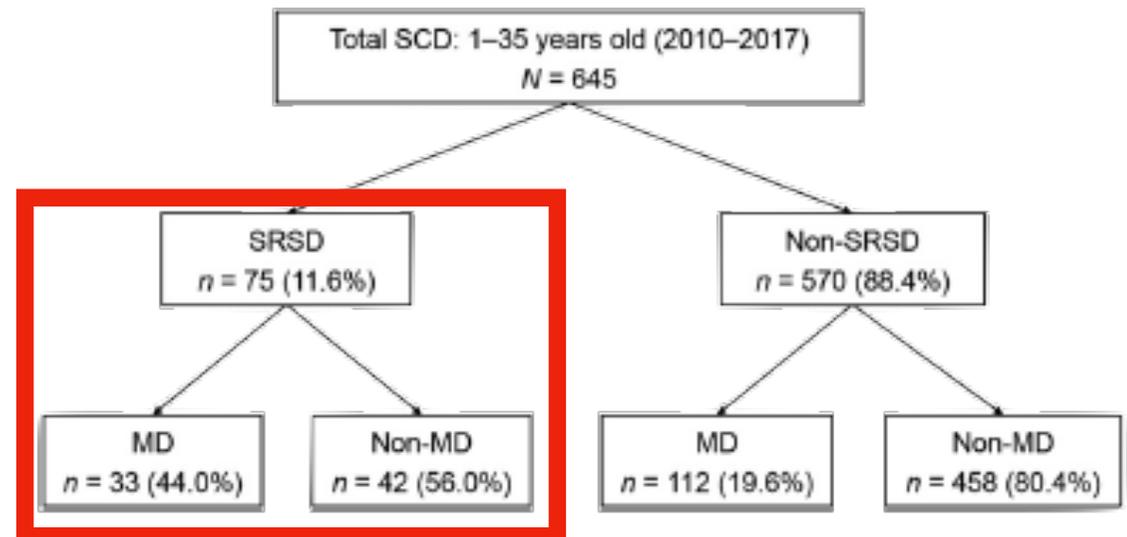
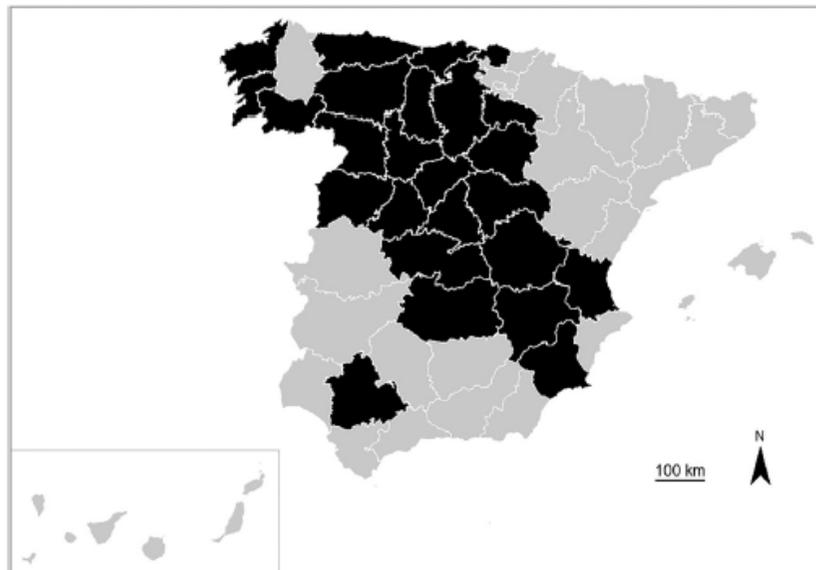
La muerte súbita sucedió, sobre todo, sin síntomas previos



Sports-related sudden cardiac death due to myocardial diseases on a population from 1–35 years: a multicentre forensic study in Spain

Benito Morentin ^a, M. Paz Suárez-Mier^b, Ana Monzó^c, Pilar Molina ^c and Joaquín S. Lucena ^d

^aSection of Forensic Pathology, Basque Institute of Legal Medicine, Bilbao, Spain; ^bHistopathology Service, Department of Madrid, National Institute of Toxicology and Forensic Sciences, Spain; ^cForensic Pathology Service, Institute of Legal Medicine and Forensic Sciences, Valencia, Spain; ^dForensic Pathology Service, Institute of Legal Medicine and Forensic Sciences, Seville, Spain



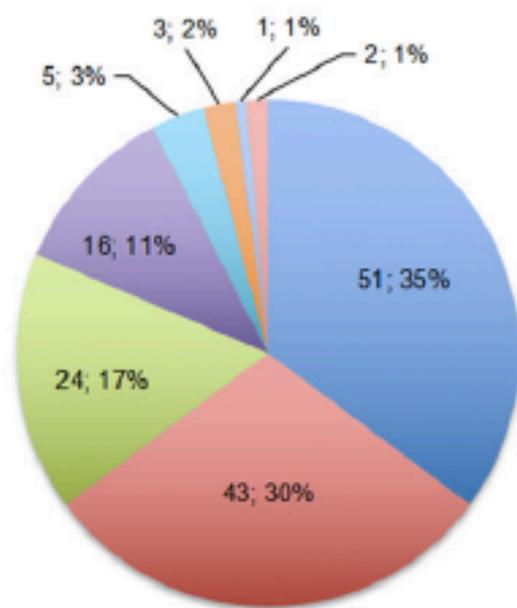


Sports-related sudden cardiac death due to myocardial diseases on a population from 1–35 years: a multicentre forensic study in Spain

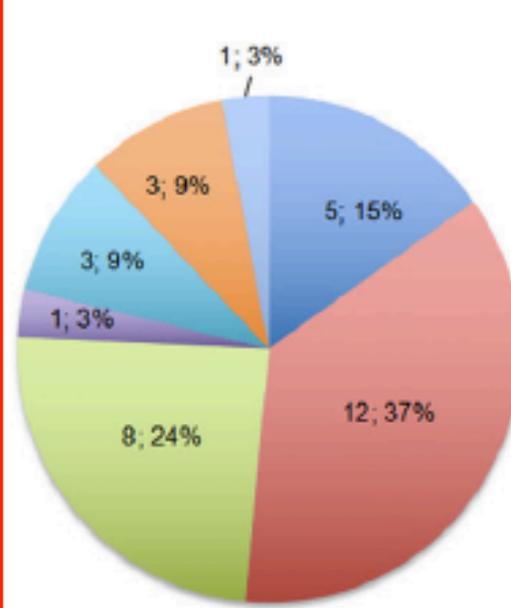
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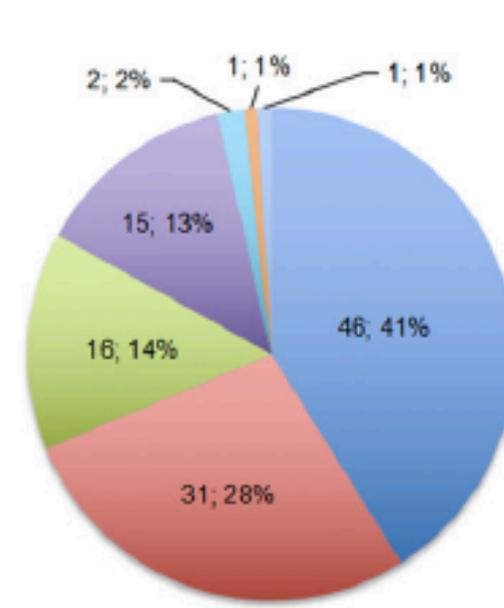
Total cases of SCD by MD (1–35 year-old) (*n* = 145)



Cases of MD in SRSD (*n* = 33)



Cases of MD in non-SRSD (*n* = 112)



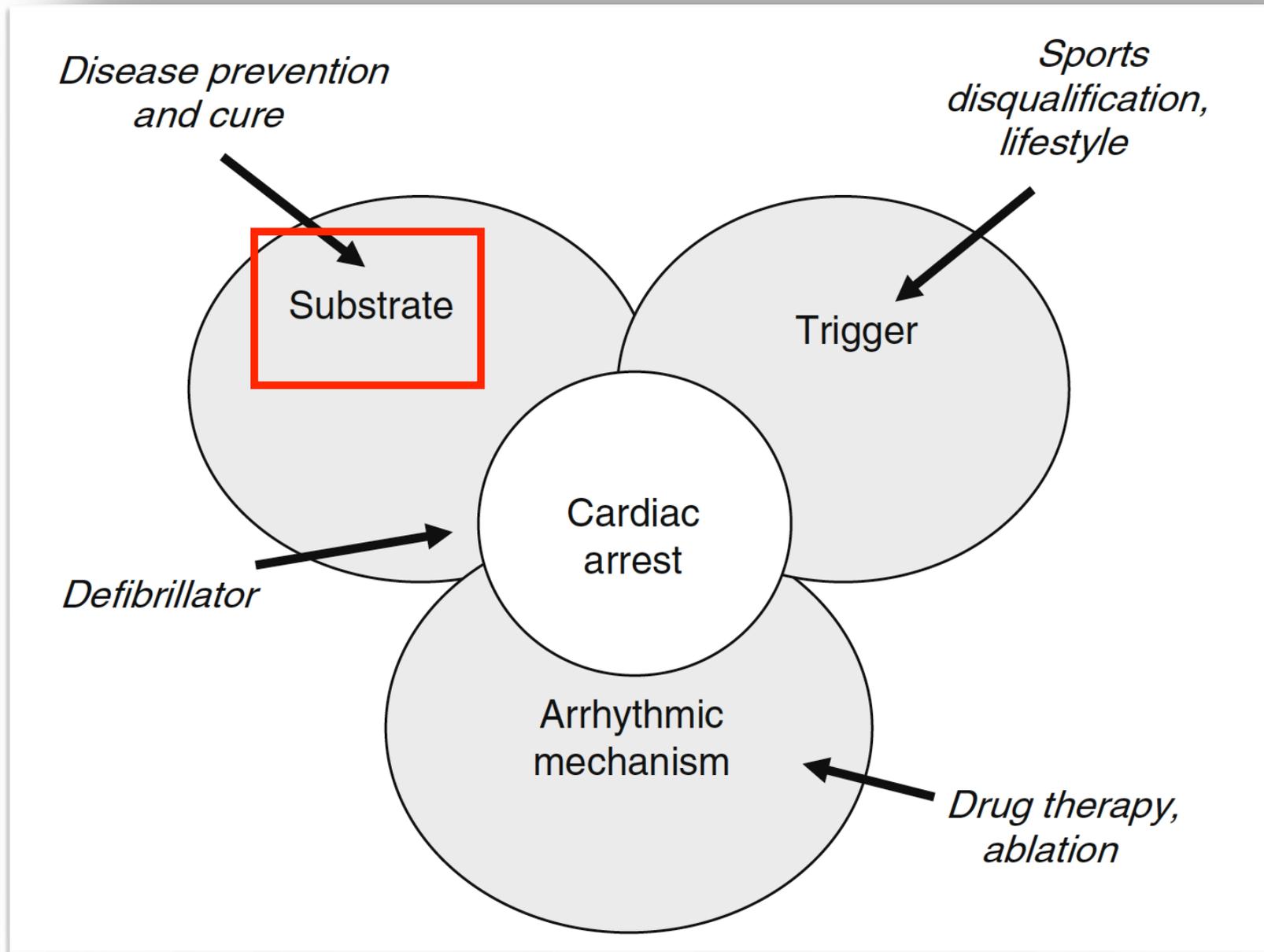
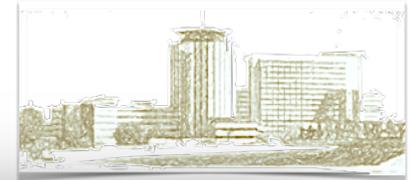
■ Myocarditis ■ Arrhythmogenic cardiomyopathy ■ Hyperthrophic cardiomyopathy ■ Dilated cardiomyopathy
 ■ Idiopathic left ventricle hypertrophy ■ Mixed phenotypes ■ Non-compacted cardiomyopathy ■ Others

Figure 3. Causes of sudden cardiac death (SCD) due to myocardial diseases (MD) and their relation to exercise. SRSD: sports-related sudden death; non-SRSD: non-sports related sudden death; MD: myocardial diseases.

El problema clínico



- ☑ El ejercicio (el deporte) es **UN** desencadenante de **ARRITMIAS MORTALES** en individuos predispuestos



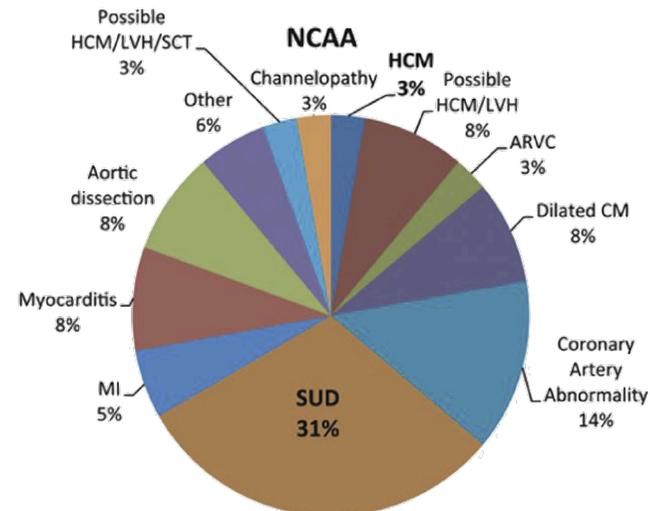
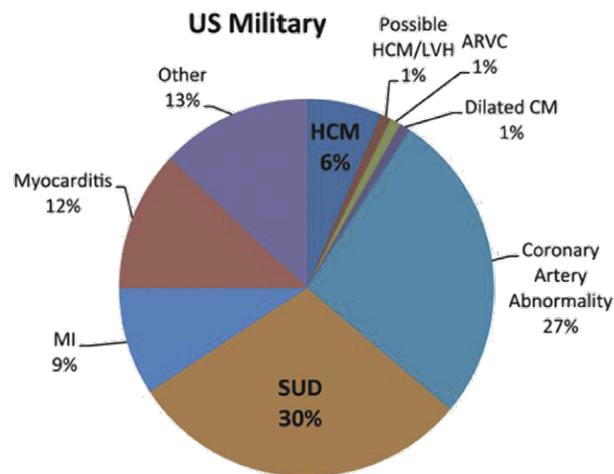
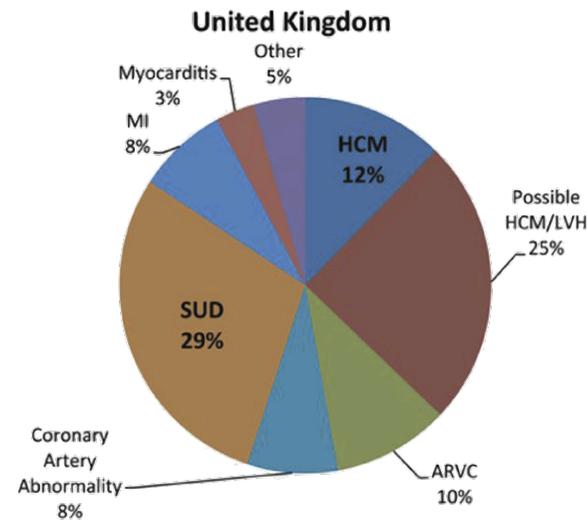
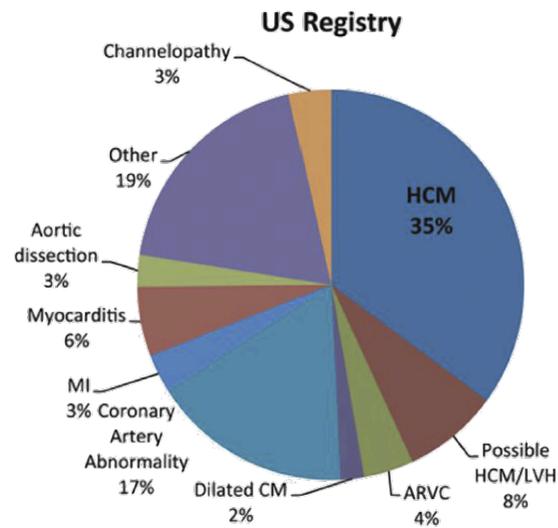
PREGUNTAS

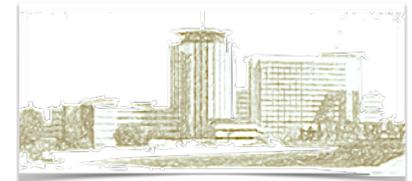


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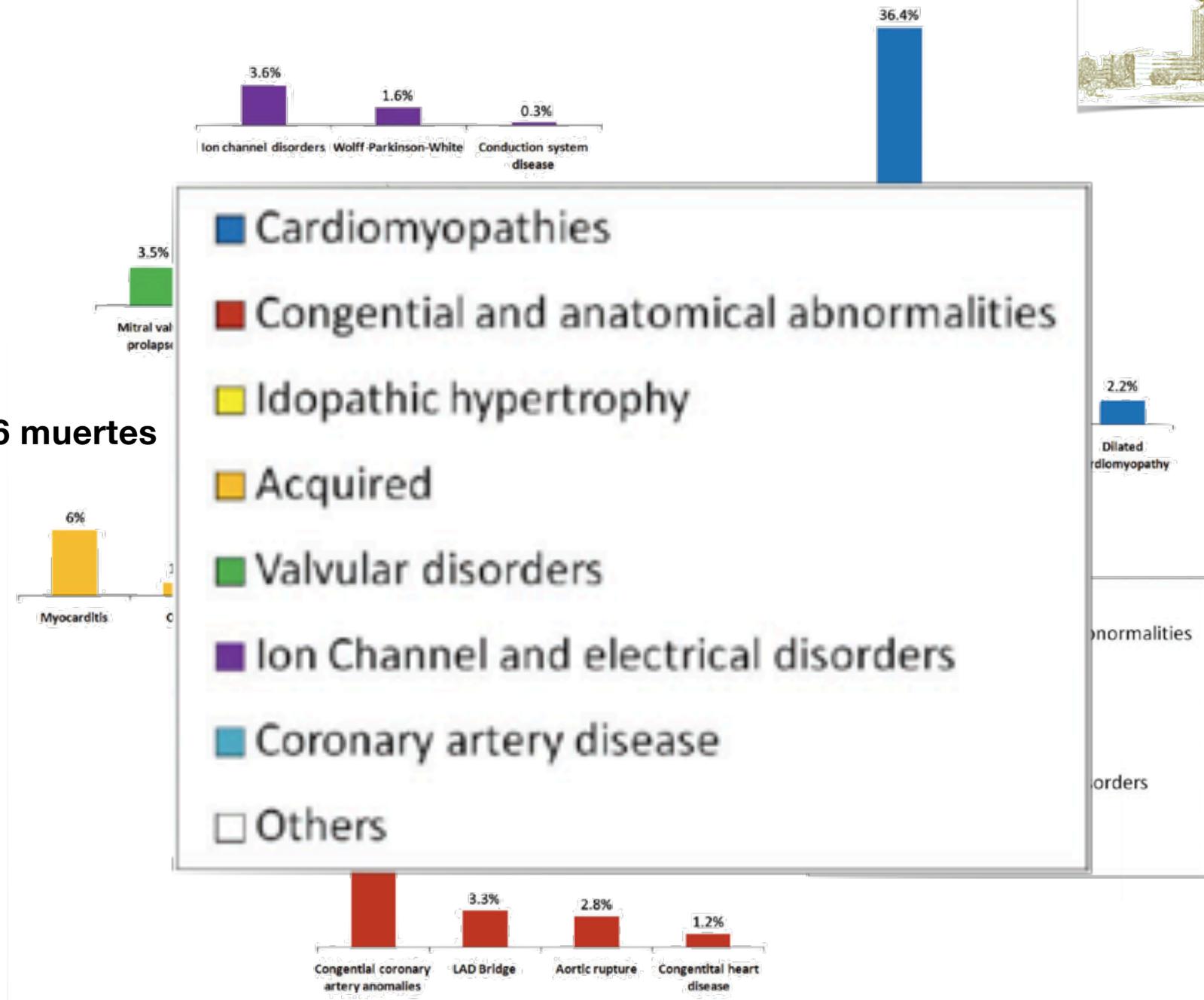


Variabilidad de fuentes de datos





1866 muertes



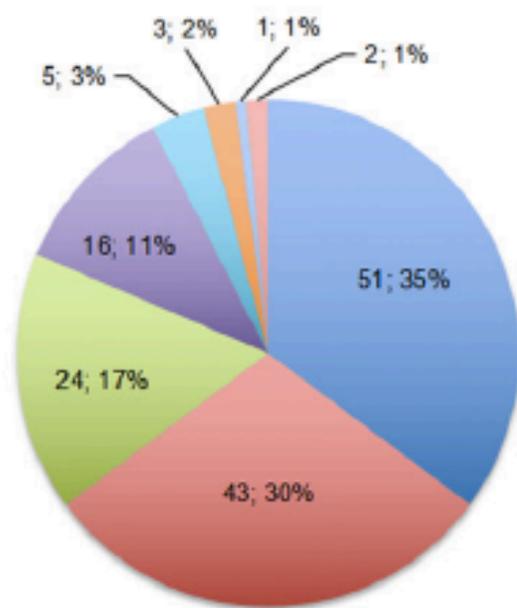


Sports-related sudden cardiac death due to myocardial diseases on a population from 1–35 years: a multicentre forensic study in Spain

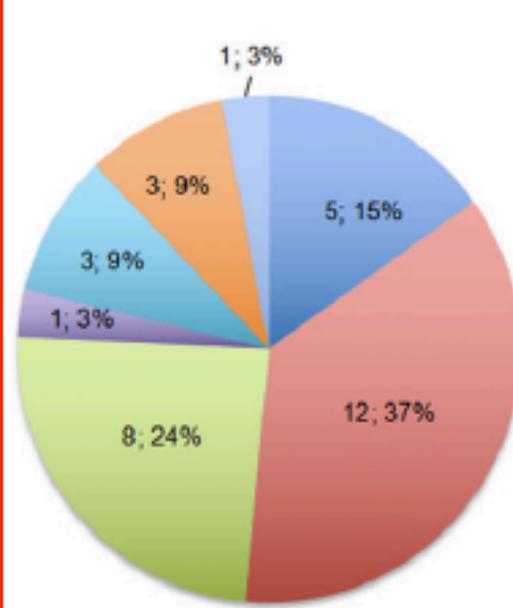
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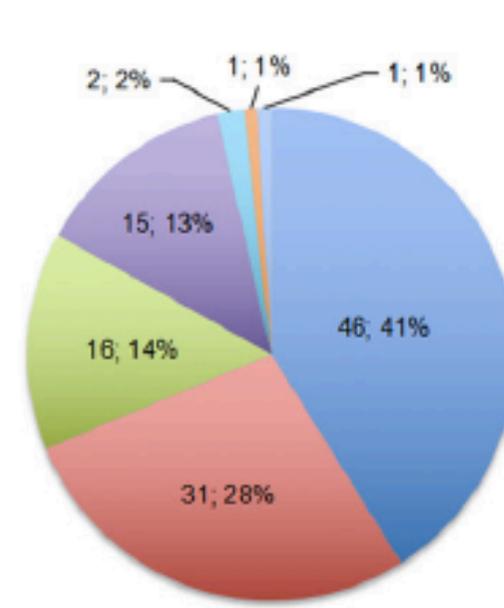
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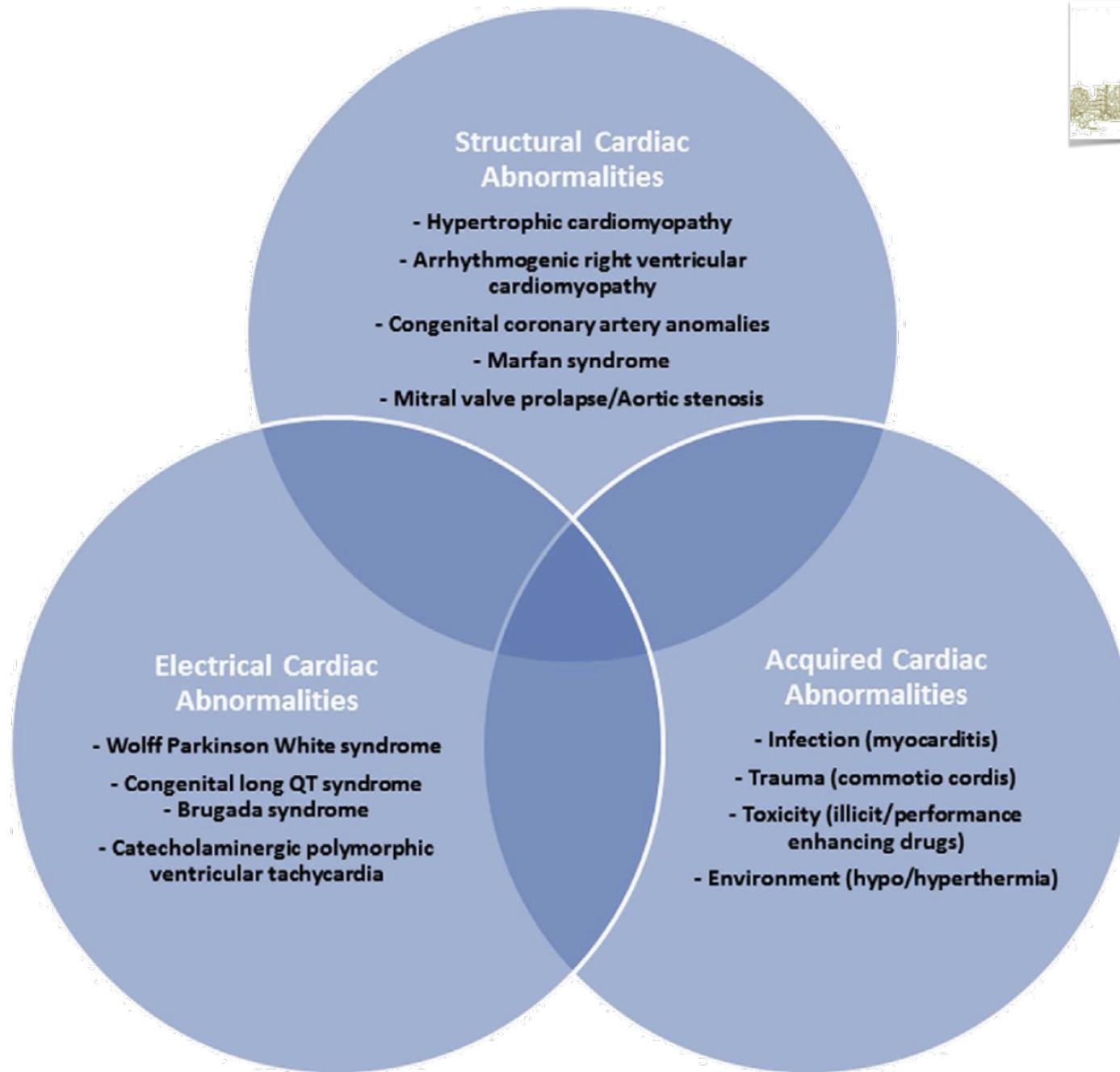


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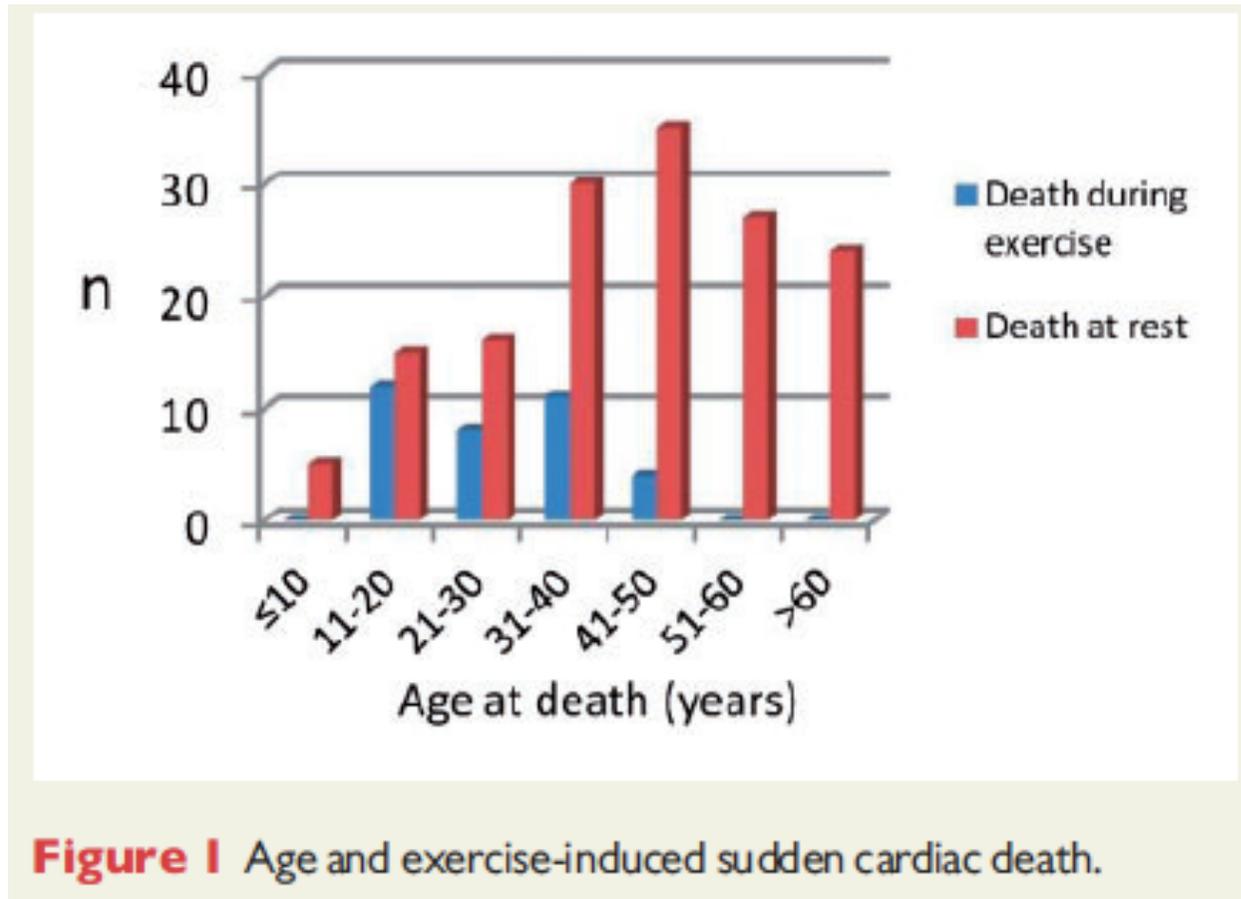
Estimaciones epidemiológicas

- Anomalías de las arterias coronarias: 1:100?
- Miocardiopatía hipertrófica: 1:500
- Wolf-Parkinson-White: 1:750
- Síndrome de QT largo: 1:2⁵⁰⁰
- Brugada: 1:2⁰⁰⁰-1: 5⁰⁰⁰
- TVP catecolaminérgica: 1: 10⁰⁰⁰
- Marfan: 1: 5⁰⁰⁰

SUMA PARCIAL 1:200-1:300



Miocardiopatía hipertrófica



Circunstancias de aparición de la muerte súbita en la MCH

Anomalías coronarias



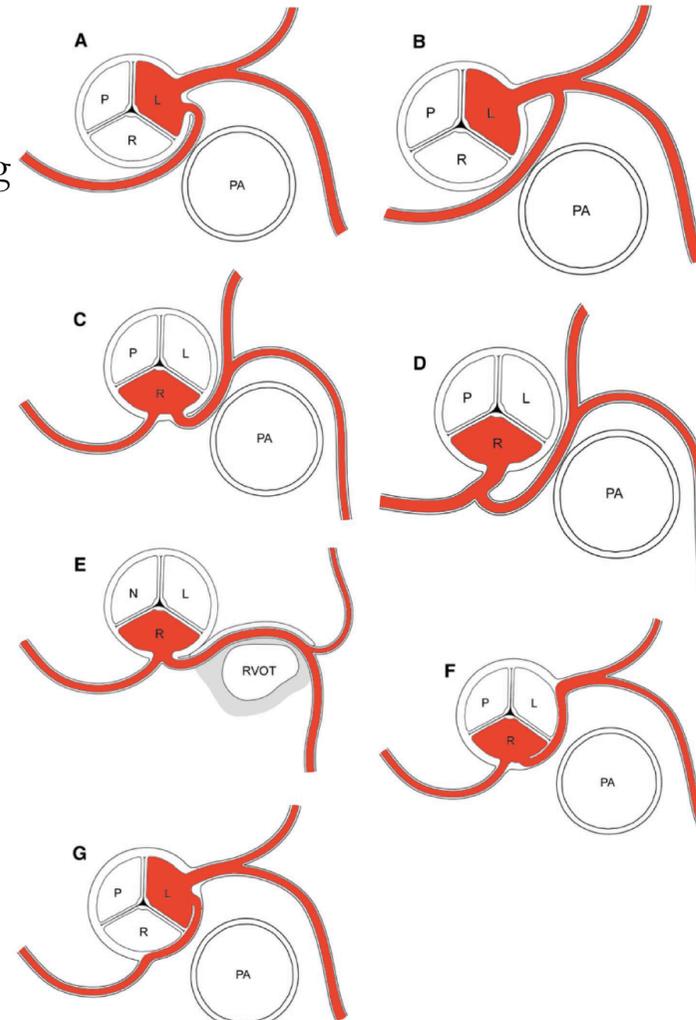
- Del 12 al 33% de atletas con muerte súbita.
- Origen de TCI del seno derecho de Valsalva y origen del seno izquierdo

-CARACTERÍSTICAS DE ALTO RIESGO

- Ostium tipo rendija
- Trayecto angulado
- Trayecto interarterial

-Isquemia aguda durante el ejercicio

-Isquemia crónica: fibrosis==>SUSTRATO
ARRITMOGÉNICO



Marfan

CAUSA ARRÍTMICA MUY RARA*

Table 1
Findings in Sudden and/or Unexpected Death in Marfan Syndrome

A. Causes of death related directly to Marfan syndrome

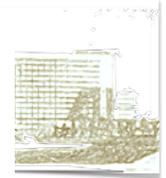
1. Aortic dissection
 - i. Hemopericardium/cardiac tamponade.
 - ii. Hemothorax.
 - iii. Vascular compromise.
 - a. Coronary artery occlusion.
 - b. Other artery occlusion.
 - iv. Arteriovenous shunting.
2. Other vessel dissection
 - i. Coronary artery dissection with myocardial infarct.
 - ii. Aortic branch dissections.
3. Aneurysm rupture with hemorrhage
 - i. Intracranial.
 - ii. Ductus arteriosus.
 - iii. Other.
4. Mitral valve prolapse
5. Aortic valve dilatation with left ventricular failure
6. Ventricular arrhythmias
7. Endocarditis
 - i. Native valves.
 - ii. Prosthetic valves.
8. Postvascular/valvular surgical complications
9. Atlanto-occipital instability with cord/brainstem compression

B. Conditions/Activities exacerbating underlying pathological features of Marfan syndrome

1. Pregnancy
2. Exercise
3. Trauma
4. Drug abuse

C. Lethal events not necessarily related to Marfan syndrome

1. Accidents
2. Homicides
3. Coincidental natural diseases





M. arritmogénica de VD

Circumstances of death

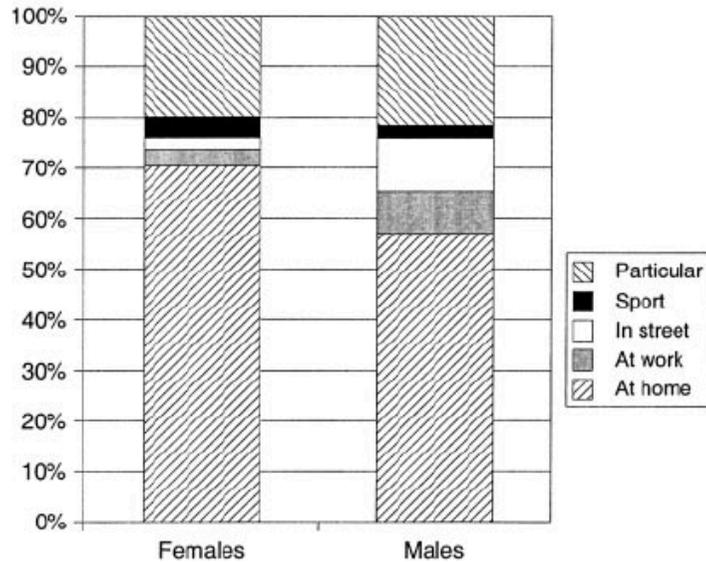
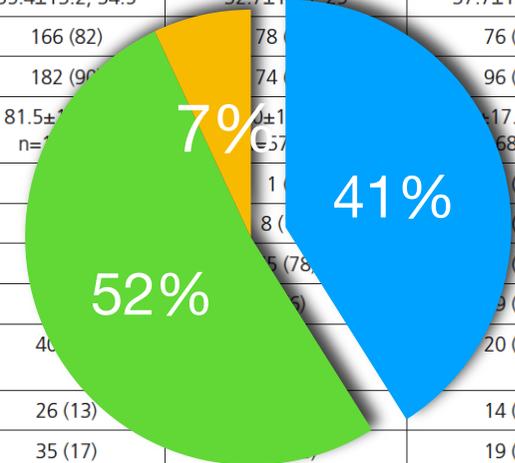


Figure 3. Circumstances of death. Percentage of 5 types of circumstances of death in 98 female and 102 male cases of ARVC/D. For explanation of “Particular” circumstances, see “Circumstances of Death” in Results.

Characteristics	Overall	Death During Exertion	Death at Rest/ During Sleep
n (%)	202	83 (41)	105 (52)
Age at death, y	35.4±13.2, 34.5	32.7±12.5, 29	37.7±13.1, 37
Male, n (%)	166 (82)	78 (41)	76 (72)
White, n (%)	182 (90)	74 (41)	96 (91)
Body weight, median	81.5±17.5, n=197	81.5±17.5, n=57	81.7±17.8, 80.55, n=68 (65)
Diagnosis of cardiomyopathy antemortem, n (%)	1 (0.5)	1 (1)	1 (1)
Previous syncope, n (%)	8 (4)	8 (8)	11 (11)
Asymptomatic status, n (%)	155 (77)	55 (58)	76 (76)
FH SCD (<35 y old), n (%)	19 (9)	19 (23)	9 (9)
Macroscopically normal appearance of the heart at autopsy	40 (20)	20 (24)	20 (19)
Dissecting aortic aneurysm	26 (13)	14 (17)	14 (13)
Dissecting aortic aneurysm with aortic valve disease	35 (17)	19 (23)	19 (18)
Biventricular hypertrophy	141 (70)	57 (69)	72 (69)



■ Durante ejercicio
■ Reposo/sueño
■ Otros



WPW y muerte súbita

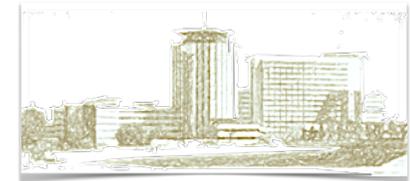
TABLE 1 Characteristics of the Study Population

Age (yrs)	Sex	Symptoms and Medical History	Circumstances of Death	HW (g)	MWT (mm)	LV Fibrosis	RV Fibrosis	Autopsy Diagnosis
Asymptomatic								
7	M	—	Unknown	200		No	No	Normal heart
48	M	—	At rest	526	20	Yes	No	HCM, mild CAD
50	F	—	During sleep	384	18	No	No	HCM
26	M	—	During exertion	510	20	Yes	No	HCM
18	M	—	At rest	390	19	No	No	Normal heart
Symptomatic								
20	M	Palpitation, on flecainide	At rest	237	9	No	No	Normal heart
26	M	Palpitation	At rest	361	16	No	No	Normal heart
20	M	Palpitation	At rest	444	14	No	No	Normal heart
20	F	Palpitation	At rest	311		No	No	Normal heart
46	M	Palpitation, on beta-blocker	At rest	474	16	No	No	Cardiac sarcoid
33	M	Palpitation	At rest	532	21	No	No	Enlarged LV
16	M	Palpitation	At rest	366	25	No	No	HCM
36	M	Syncope	At rest	498		Yes	No	Idiopathic fibrosis
55	F	Palpitation, on beta-blocker	At rest	385	13	No	No	Normal heart
Previous Ablation								
28	M	Palpitation	During sleep	316	19	No	No	LVH, mild CAD
27	M	Asymptomatic	During exertion	426	21	No	No	Normal heart
24	M	Asymptomatic	At rest	578		No	No	ILVH
65	M	Palpitation	At rest	486	12	No	No	Normal heart
28	M	Palpitation	During sleep	302	12	No	No	Normal heart

CAD = coronary artery disease; HCM = hypertrophic cardiomyopathy; HW = heart weight; ILVH = idiopathic left ventricular hypertrophy; LV = left ventricle; LVH = left ventricular hypertrophy; MWT = maximal wall thickness; RV = right ventricle.

Life-Threatening Event Risk in Children With Wolff-Parkinson-White Syndrome

A Multicenter International Study



Susan P. Etheridge, MD,^a Carolina A. Escudero, MD,^b Andrew D. Blaufox, MD,^c Ian H. Law, MD,^d Brynn E. Dechert-Crooks, RN, MSN,^e Elizabeth A. Stephenson, MD,^f Anne M. Dubin, MD,^g Scott R. Ceresnak, MD,^g Kara S. Motonaga, MD,^g Jonathan R. Skinner, MBChB, MD,^h Luciana D. Marcondes, MD,^h James C. Perry, MD,ⁱ Kathryn K. Collins, MD,^j Stephen P. Seslar, MD,^k Michel Cabrera, MD,^l Orhan Uzun, MD,^m Bryan C. Cannon, MD,ⁿ Peter F. Aziz, MD,^o Peter Kubuš, MD,^p Ronn E. Tanel, MD,^q Santiago O. Valdes, MD,^r Sara Sami, MD,^r Naomi J. Kertesz, MD,^s Jennifer Maldonado, MBA, CCRP,^d Christopher Erickson, MD,^t Jeremy P. Moore, MD,^u Hiroko Asakai, MD,^f LuAnn Mill, RN, BSN,^t Mark Abcede, MBA, CCRP,ⁱ Zebulun Z. Spector, MD,^k Shaji Menon, MD,^a Mark Shwayder, MD,^g David J. Bradley, MD,^e Mitchell I. Cohen, MD,^v Shubhayan Sanatani, MD^w

22 centros, 6 países, 25 años de evolución

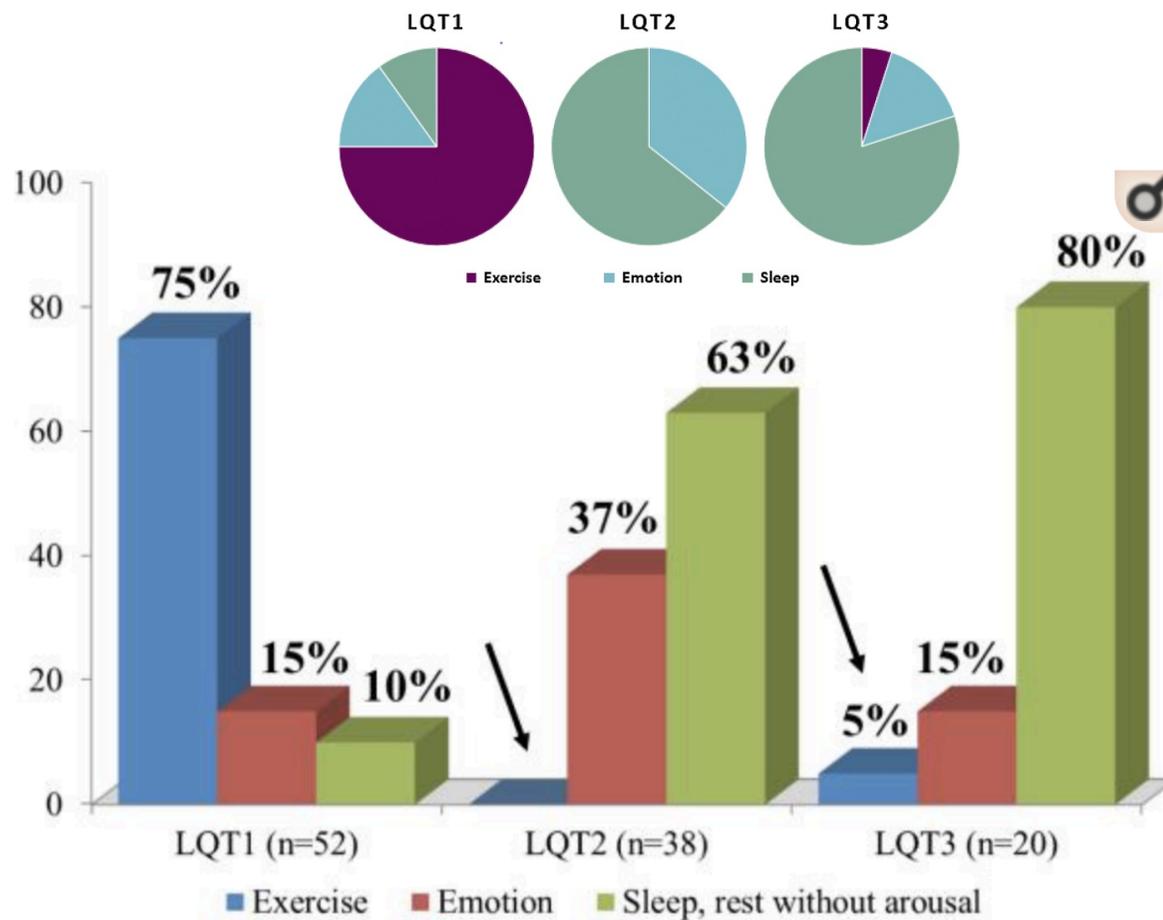
912 pacientes < 21 años

“The LTE occurred **most often at rest or with noncompetitive activity** and was equally likely to be rapidly conducted pre-excited atrial fibrillation and aborted sudden death.”

TABLE 2 Clinical Characteristics of Case Subjects (N = 96)

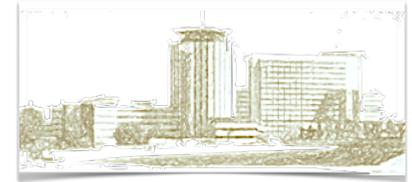
Age at LTE (yrs)	14.1 ± 3.9 (range 0.4 months-21 yrs)
Known WPW	40 (42)
LTE as presenting symptom	62 (65)
LTE diagnosis	
Pre-excited atrial fibrillation	47 (49)
Aborted sudden death	43 (45)
Sudden death	6 (6)
Activity at time of LTE	
Rest	37 (39)
Active, noncompetitive	33 (34)
Active, competitive	10 (10)
Unknown	16 (17)
Outcome of the LTE	
Full/near full recovery	82 (85)
Recovery with neurological injury	5 (5)
Death	9 (9)

SQTL



Triggers for all lethal and non-lethal cardiac events in the three genotypes

ONLINE FIRST



RESEARCH LETTER

**Competitive Sports Participation in Athletes
With Congenital Long QT Syndrome**

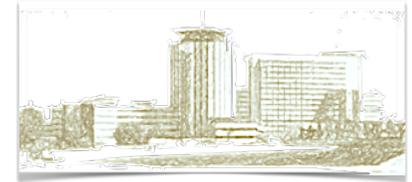
Pronóstico de pacientes con SQTL que decidieron seguir haciendo deporte a pesar de las recomendaciones en contra

Eventos globales: **0.003 atleta-año** (1 evento: 331 atleta-años; IC 95% , 1:92-1:2763)

Comentario

Con un seguimiento de más de 650 atleta-años, informan de una tasa baja de eventos cardiacos en SQTL durante el deporte.

Limitaciones: muestra pequeña, seguimiento limitado, generabilidad desconocida.



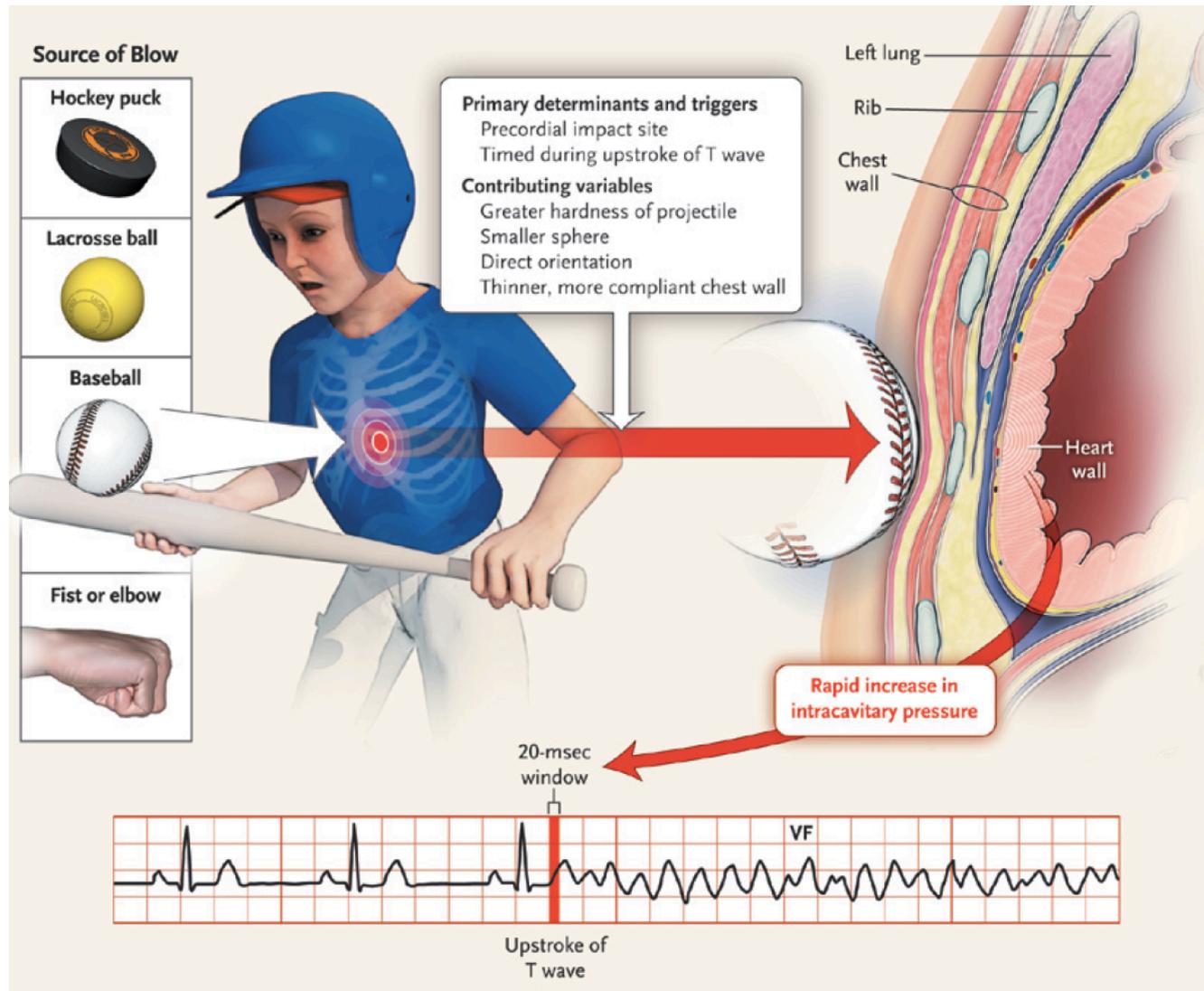
Exercise-induced syncope and Brugada syndrome

[Anjan S Batra](#),¹ [Rachel Watson](#),² and [Anthony C McCanta](#)¹

ANÉCDOTA: “patient who presented with exercise-induced syncope, ventricular tachycardia during an exercise test, and was found to be both genotypically and phenotypically positive for BrS”.

- Síntomas de SBr: tradicionalmente en reposo, a menudo en el sueño (aumento del tono vagal; disminución de la actividad simpática)
- Es muy atípico presentar síntomas en esfuerzo: no se suele restringir la actividad; la ergometría no suele ser parte de las pruebas realizadas.

Commotio cordis





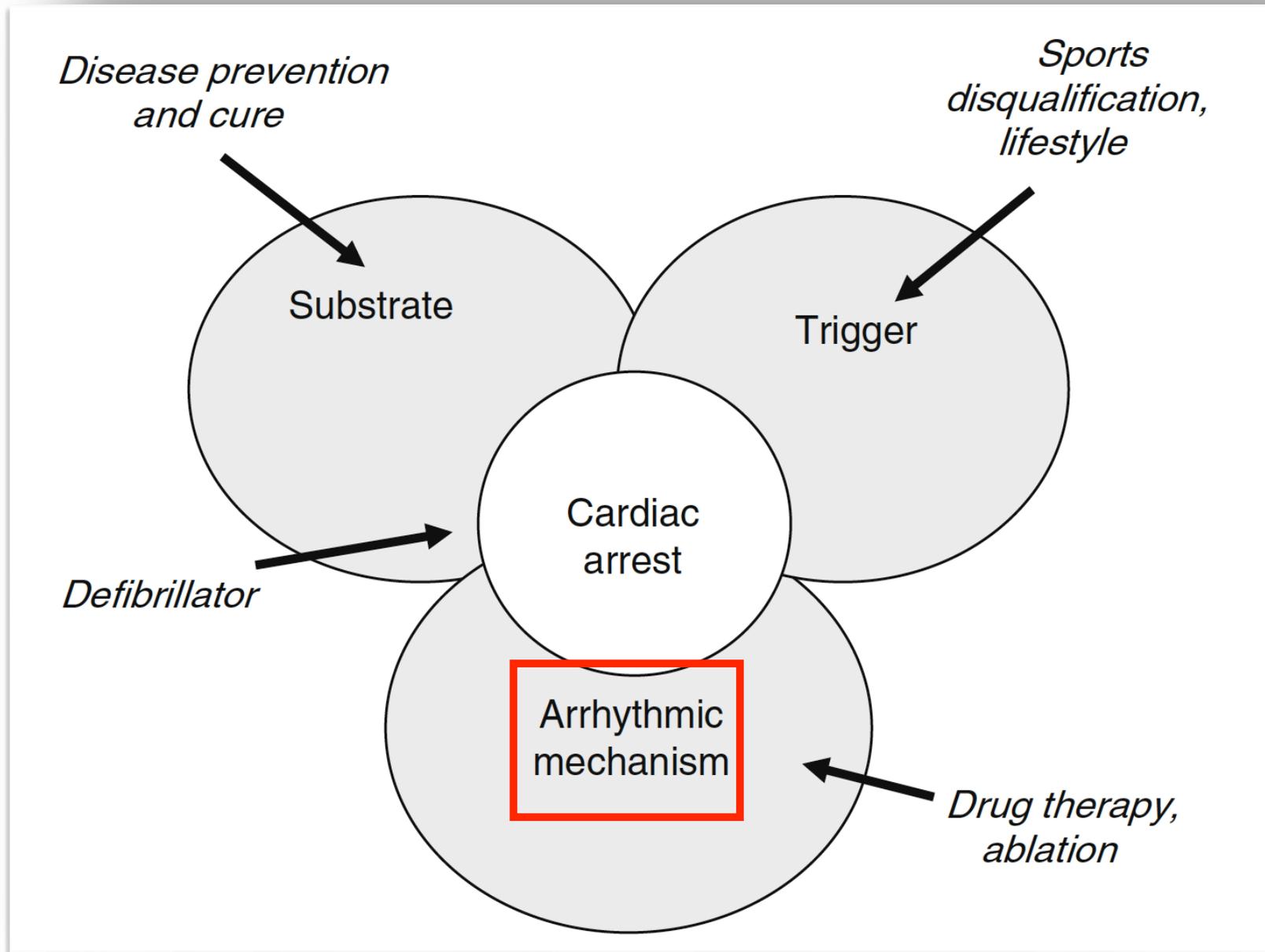
Cardiopatías congénitas

ORIGINAL ARTICLE

Nationwide Study of Sudden Cardiac Death in People With Congenital Heart Defects Aged 0 to 35 Years

Dinamarca

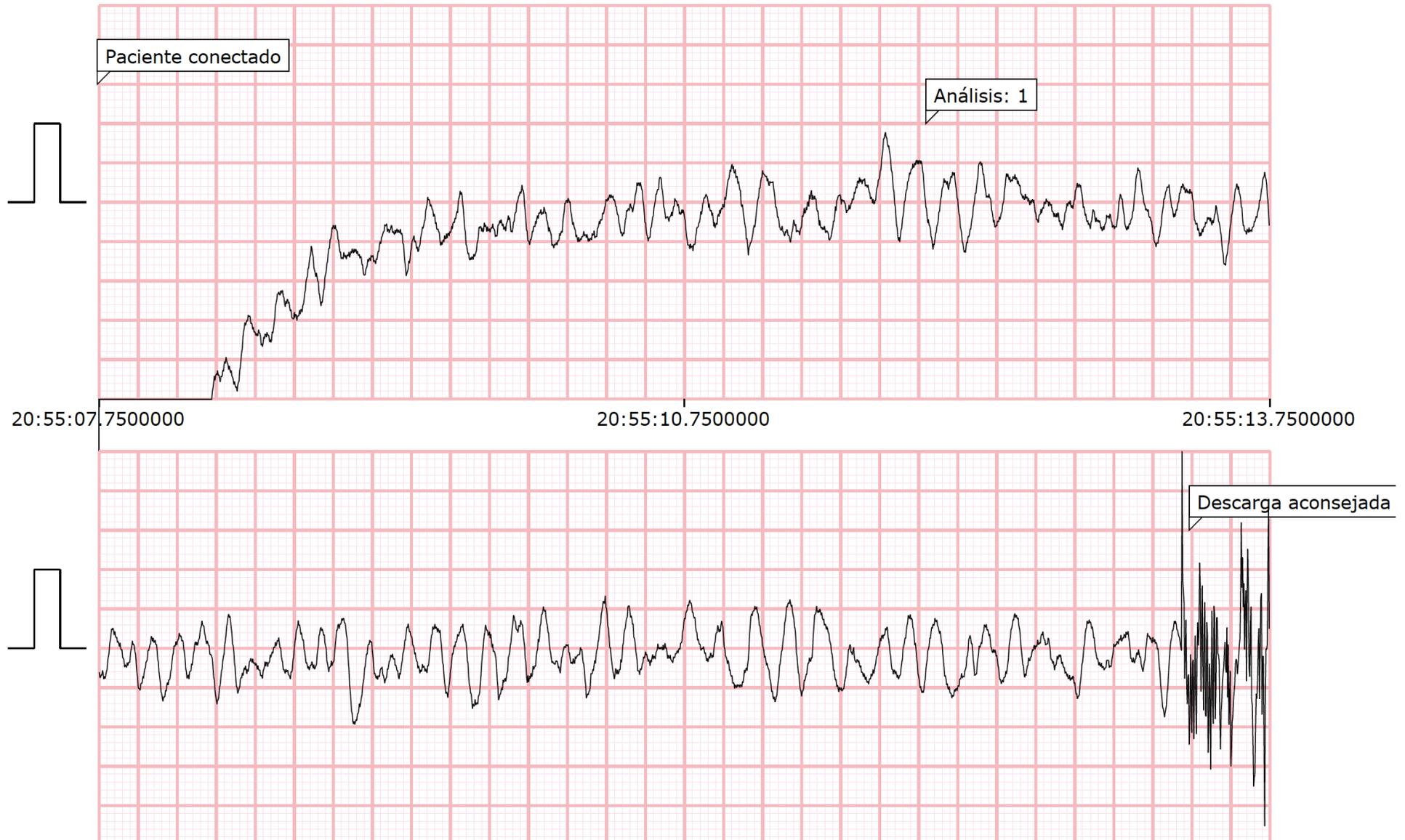
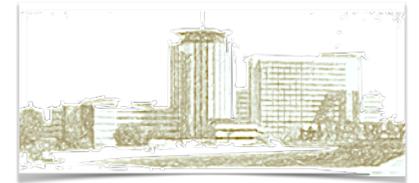
- ☞ La proporción de muerte súbita asociada a CC en el joven fue del 11% (mayor de la previamente conocida)
- ☞ La muerte súbita **asociada a la actividad física** en pacientes con CC FUE INFRECUENTE: 4% en pacientes con CC conocida, pero 18% en pacientes sin CC conocida

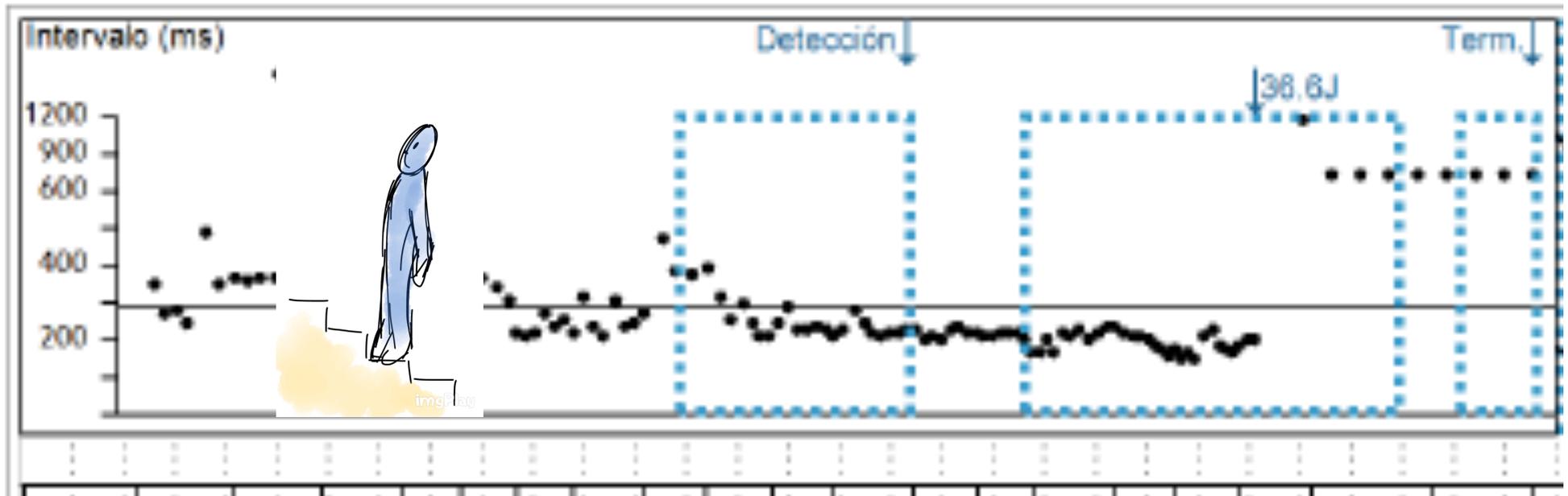
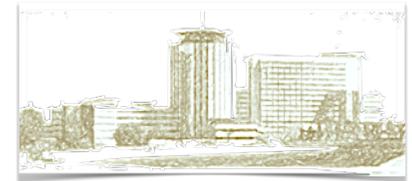


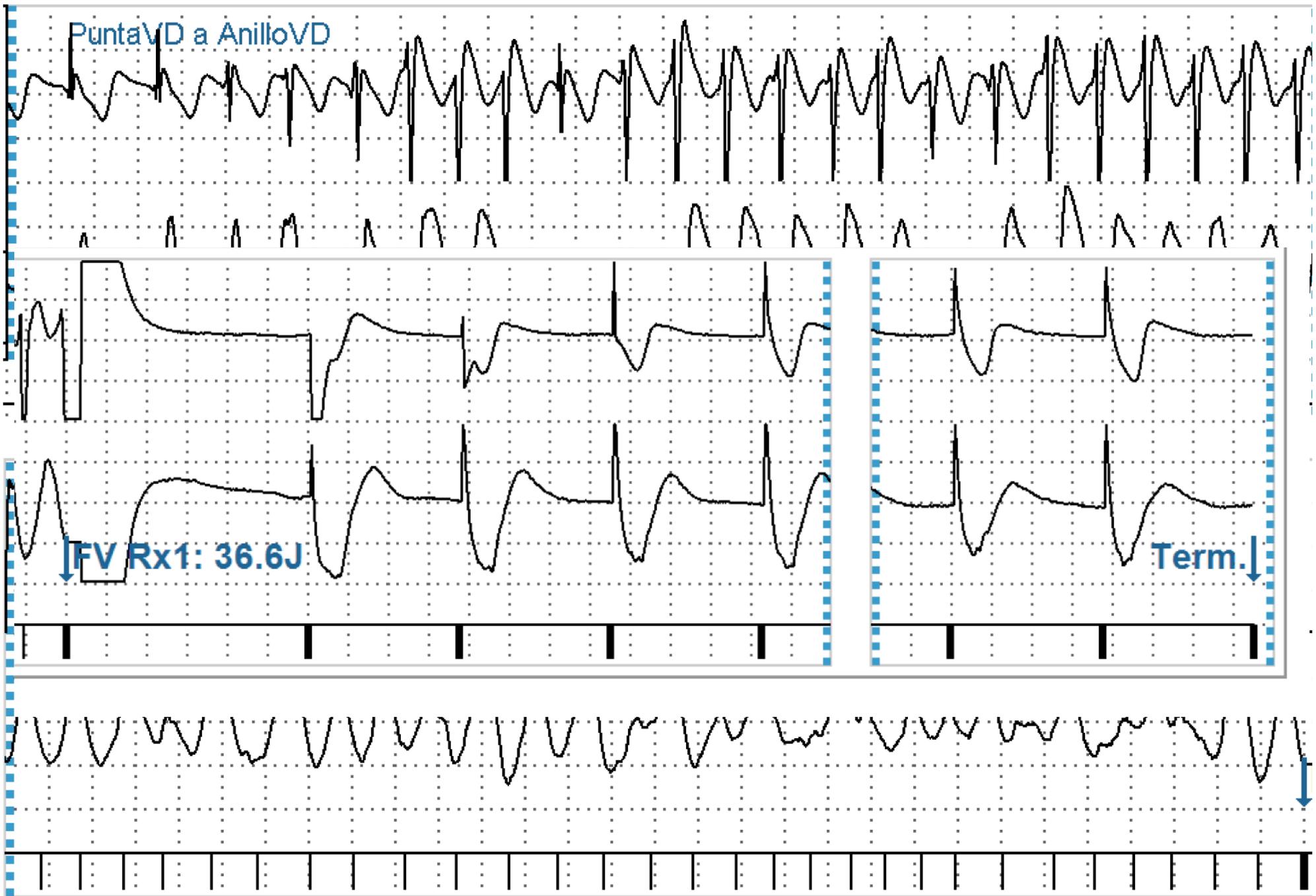
PREGUNTAS

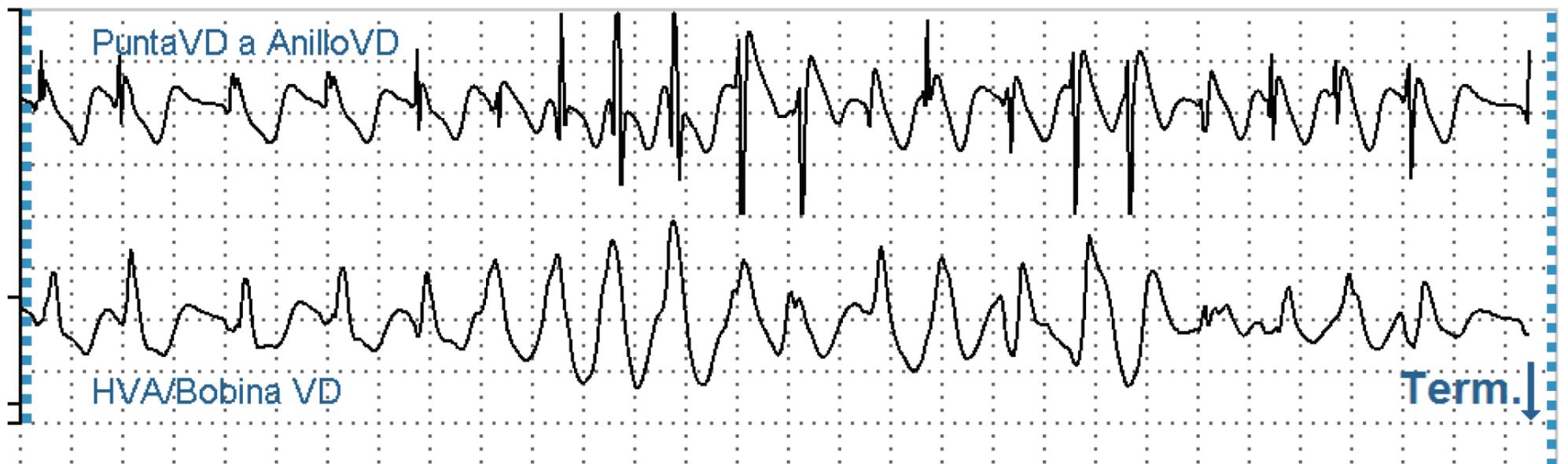


- ¿CUÁLES SON LOS INDIVIDUOS PREDISPUUESTOS? ¿QUÉ ENFERMEDADES TIENEN? ¿LOS PODEMOS IDENTIFICAR? ¿CÓMO?
- ☑ **¿QUÉ ARRITMIAS APARECEN?**
- ¿QUÉ DEPORTES PUEDEN PRECIPITAR LAS ARRITMIAS? ¿HAY DEPORTES MÁS SEGUROS QUE OTROS?









Ritmos registrados...

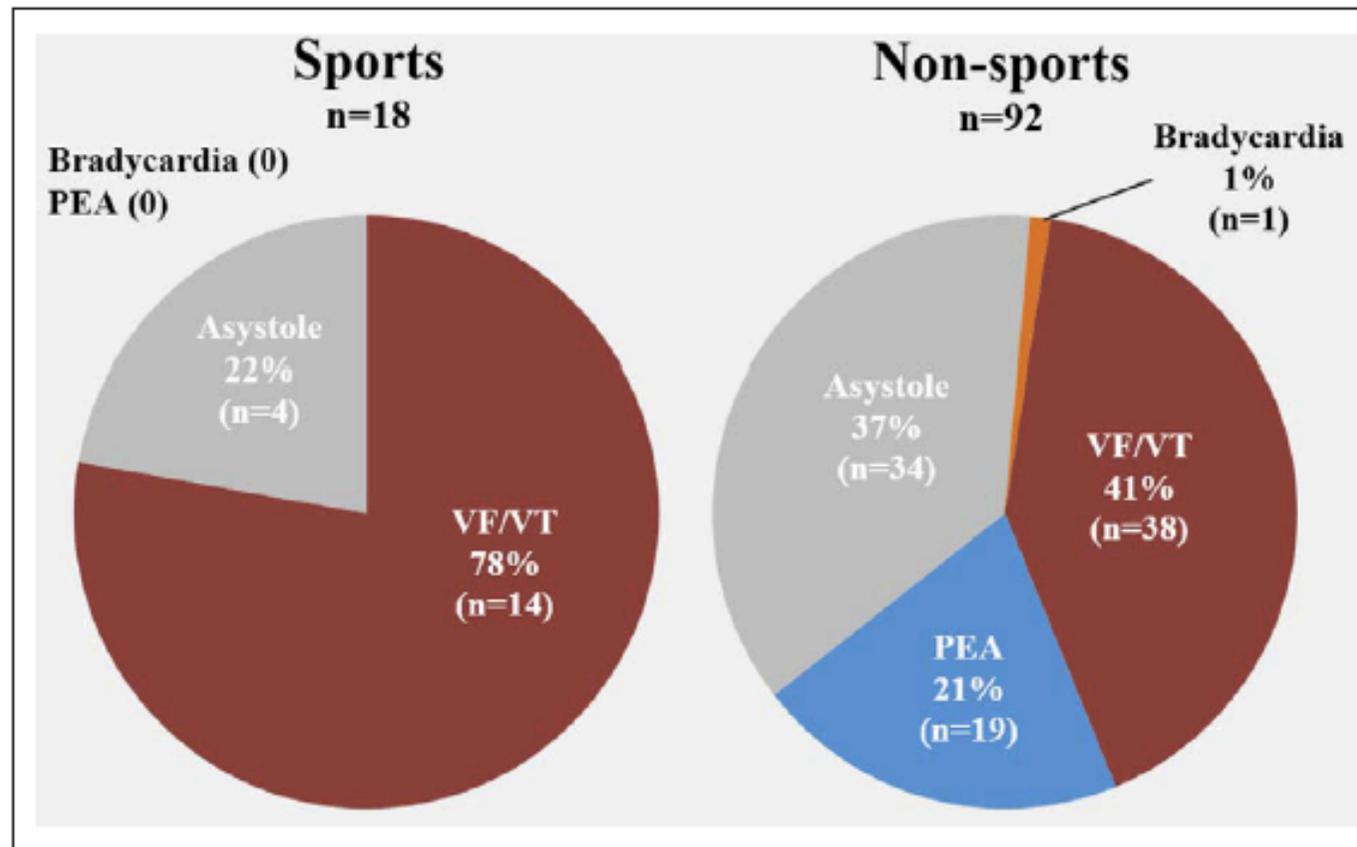
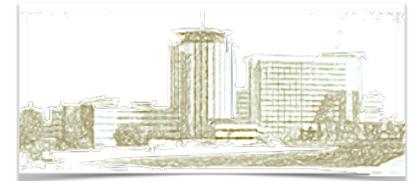
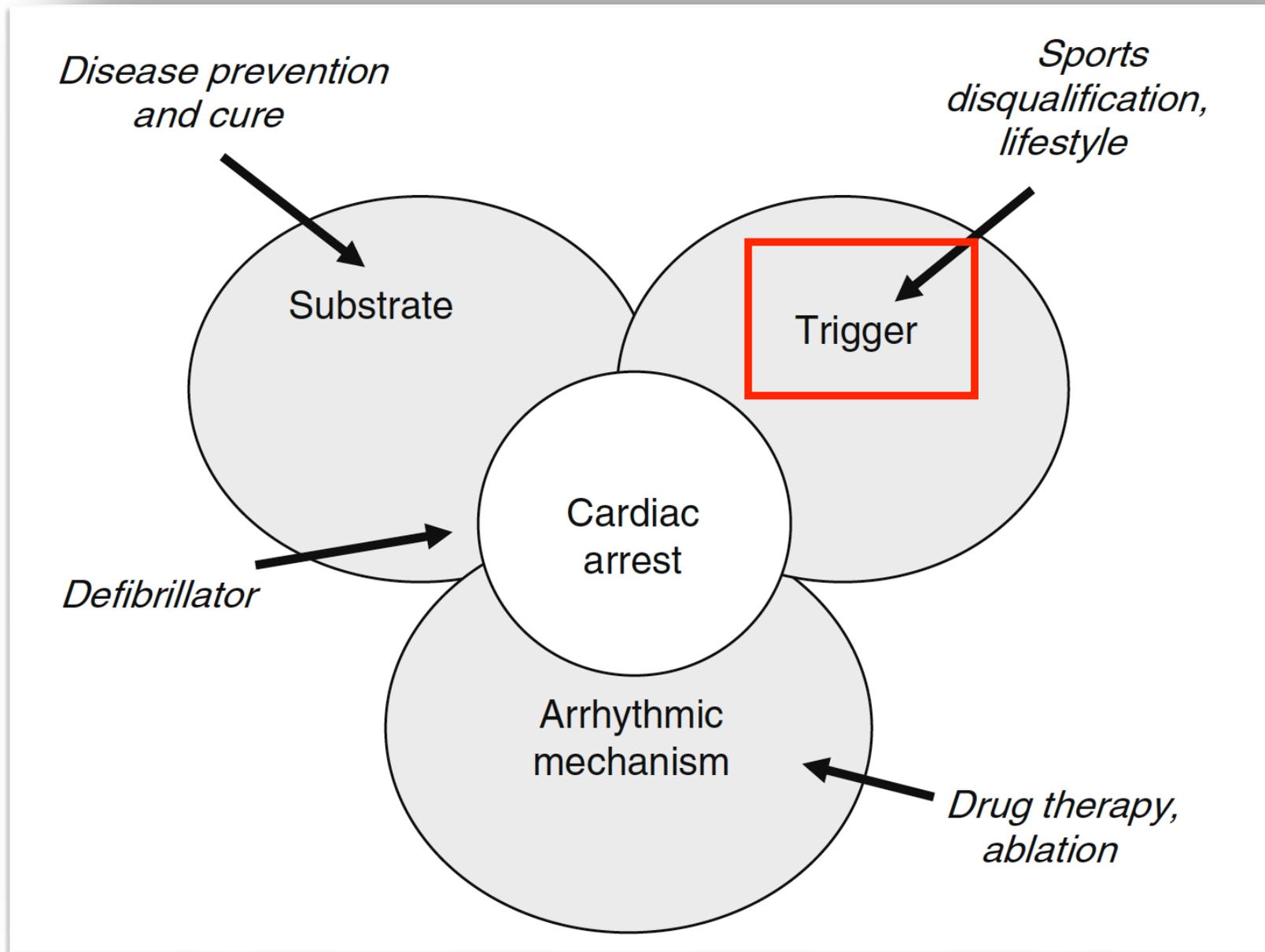
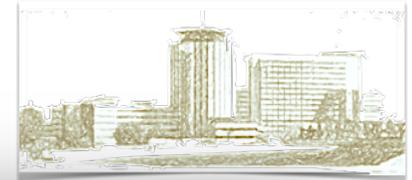


Figure 2. Initial rhythm recorded during presentation with sudden cardiac arrest.

Shockable rhythm (VF/VT) was significantly more common among sports-related than nonsports SCAs. Among the subset of 21 sports-related and 105 nonsports-related cases with resuscitation attempted in the field, 18 sports-related and 92 nonsports-related cases had initial rhythm available from ECG recordings in the field or from EMS reports. PEA indicates pulseless electrical activity; SCA, sudden cardiac arrest; VF, ventricular fibrillation; and VT, ventricular tachycardia.



PREGUNTAS

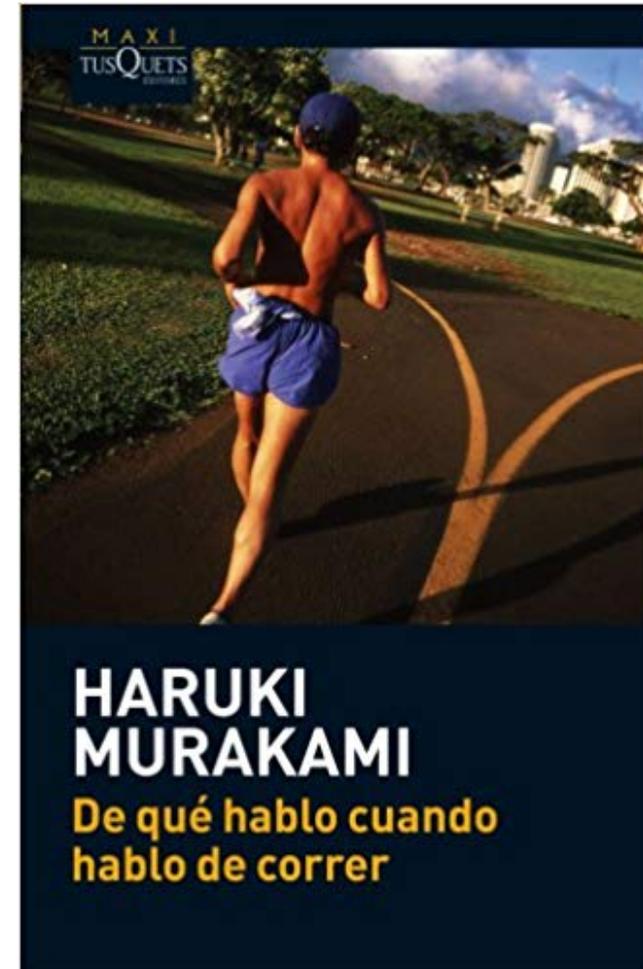


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Términos



- Reposo
- Deporte recreativo
- Deporte de competición
- Deporte profesional



Términos



- ☞ D. de COMPETICIÓN: deporte organizado o individual, donde hay un incentivo en el resultado. Los participantes se involucran en un nivel alto de entrenamiento y de competición, y pueden no tener la voluntad o el deseo de limitar su actividad (instituto, universidad, profesional)
- ☞ D. RECREATIVO: Participan por deseo de bienestar físico o por diversión, sin presión por el resultado. Los niveles de actividad pueden ser altos.



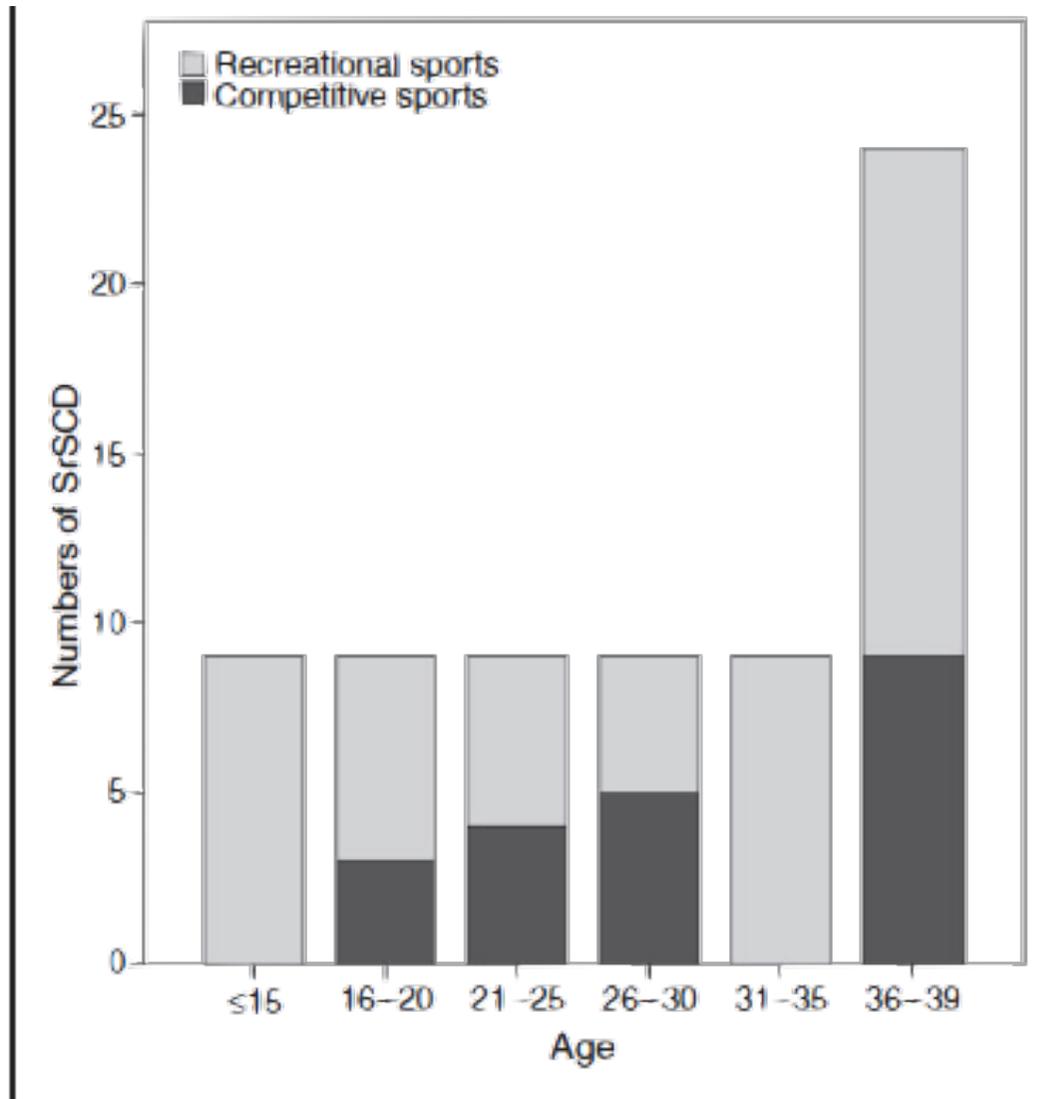
Athlete	Gender	Sport	Age (Years)	Reported Cause	Link
1	Male	Soccer	31	Bradyarrhythmia	http://www.espn.com/soccer/fiorentina/story/3408662/davide-astori-died-natural-death-from-heart-issue-autopsy-shows
2	Male	Basketball	26	Natural	http://www.espn.com/mens-college-basketball/story/_/id/18668751/former-syracuse-boston-celtics-center-fab-melo-dies-brazil
3	Female	Biathlon	21	Acute heart failure	https://www.reuters.com/article/us-biathlon-russia-yakimkina-idUSKBN0LR0Z720150223
4	Male	Soccer	25	SCA	http://www.espn.com/soccer/italian-serie-b/story/2950188/doctors-found-guilty-of-manslaughter-over-death-of-piermario-morosini
5	Male	Soccer	22	SCA	https://www.theguardian.com/football/2007/aug/28/europeanfootball.sevilla
6	Male	Soccer	30	Heart attack	http://news.bbc.co.uk/2/hi/americas/3967127.stm
7	Male	Soccer	24	Heart attack	https://www.theguardian.com/football/2004/jan/26/newssstory.sport4
8	Male	Soccer	28	Heart attack	https://www.theguardian.com/world/2003/jun/26/football
9	Male	Swimming	26	Atherosclerotic coronary artery disease	https://www.reuters.com/article/us-swimming-oen/norwegian-swim-champ-dale-oen-died-of-heart-disease-official-idUSBRE85B10820120612
10	Male	Volleyball	37	Heart attack	https://www.foxsports.com/olympics/story/vigor-bovolenta-dies-age-37-italian-volleyball-player-olympic-silver-medalist-heart-attack-032512
11	Female	Volleyball	26	Thrombosis	http://www.fivb.org/viewPressRelease.asp?No=34247&Language=en#.W65-fmhKjDc
12	Male	Soccer	26	Heart failure	https://www.theguardian.com/football/2009/aug/09/espnyol-dani-jarque-dies
13	Male	Soccer	35	Heart failure	https://www.theguardian.com/football/2007/dec/30/newssstory.sport2
14	Male	Soccer	26	Heart failure	https://www.bbc.com/sport/football/36675570
15	Male	Soccer	25	Heart collapse	http://edition.cnn.com/2010/SPORT/football/03/09/football.africa.deaths/index.html
16	Male	Soccer	27	Heart failure	https://www.theguardian.com/football/2013/aug/01/christian-benitez-died-heart-failure

SCA: sudden cardiac arrest; the Google database was searched combining the words "sudden death" and "sport" (in the link news and all) since 2003. The access date on the abovementioned websites was 31 November 2018.

☞ El deporte competitivo es muy mediático (¿nos sesga?)



Competitivo vs. recreativo



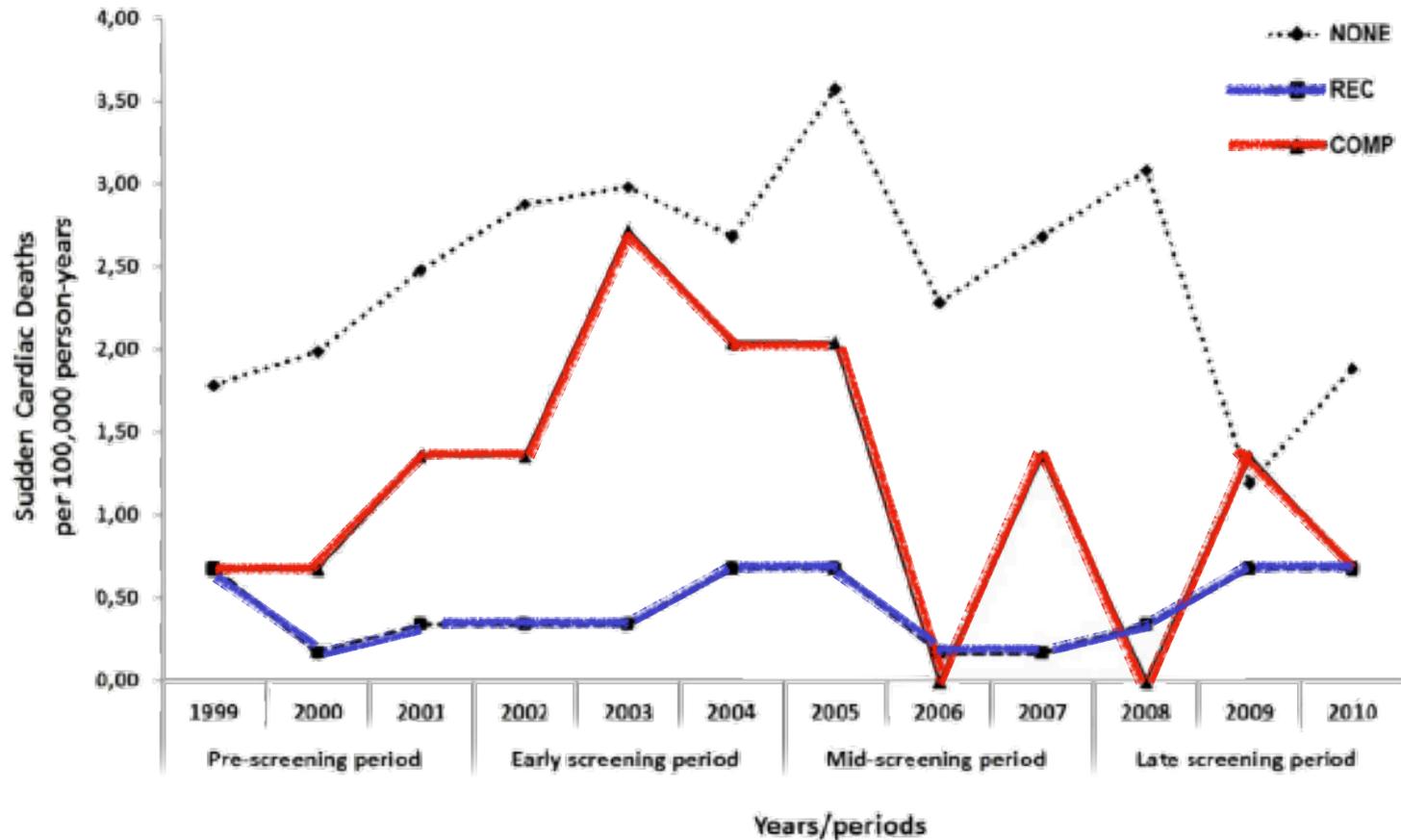
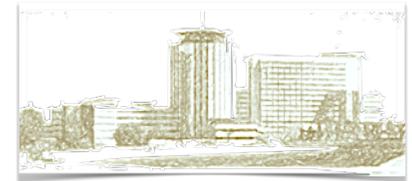


Fig 3. Annual incidence rates of sudden cardiac death in the young population of the German-speaking region of Switzerland. Young population included individuals aged 10 to 39 years. Sudden cardiac deaths (SCDs) occurring between 1999 and 2010 were evaluated. Incidence rates are presented for NONE, REC and COMP categories classified based on relation of the SCD to sports. COMP, SCD related to competitions; NONE, SCD not related to physical activities; REC, SCD associated with physical activities other than competitions.

¿Qué deportes?



11-14 años

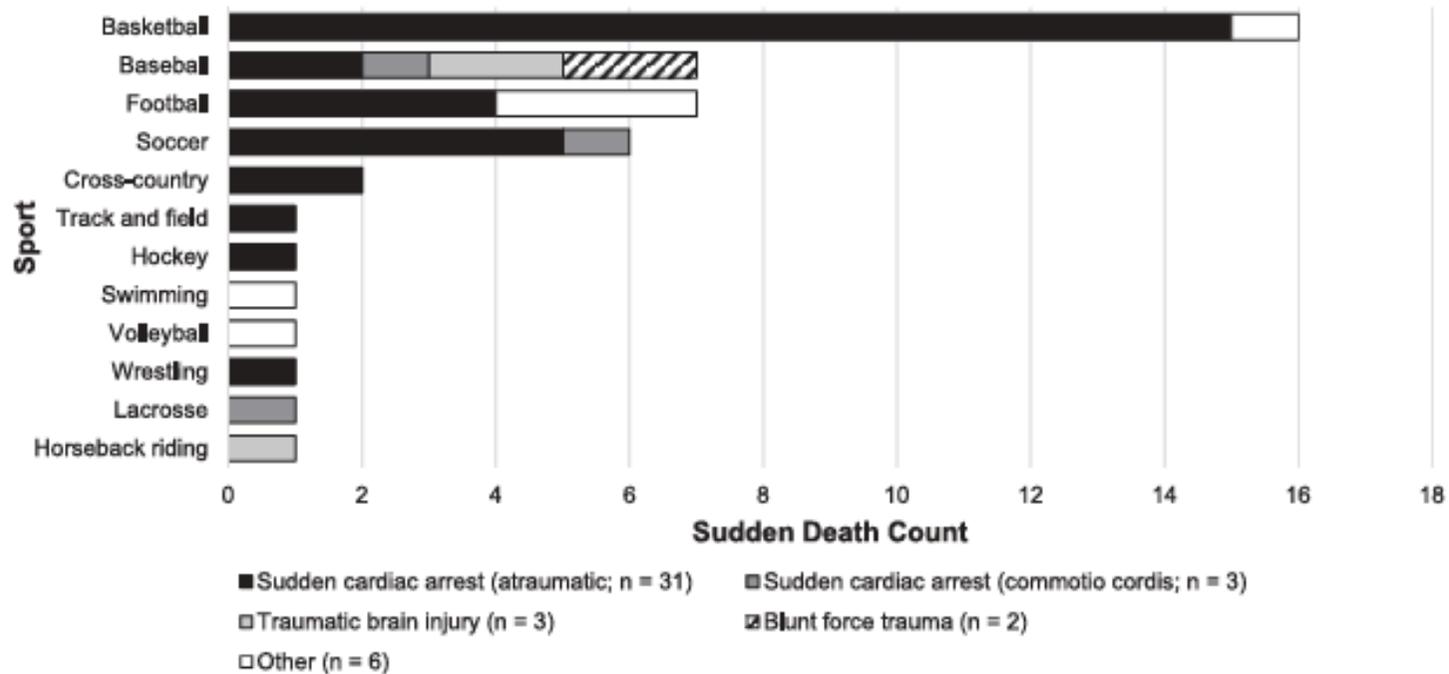
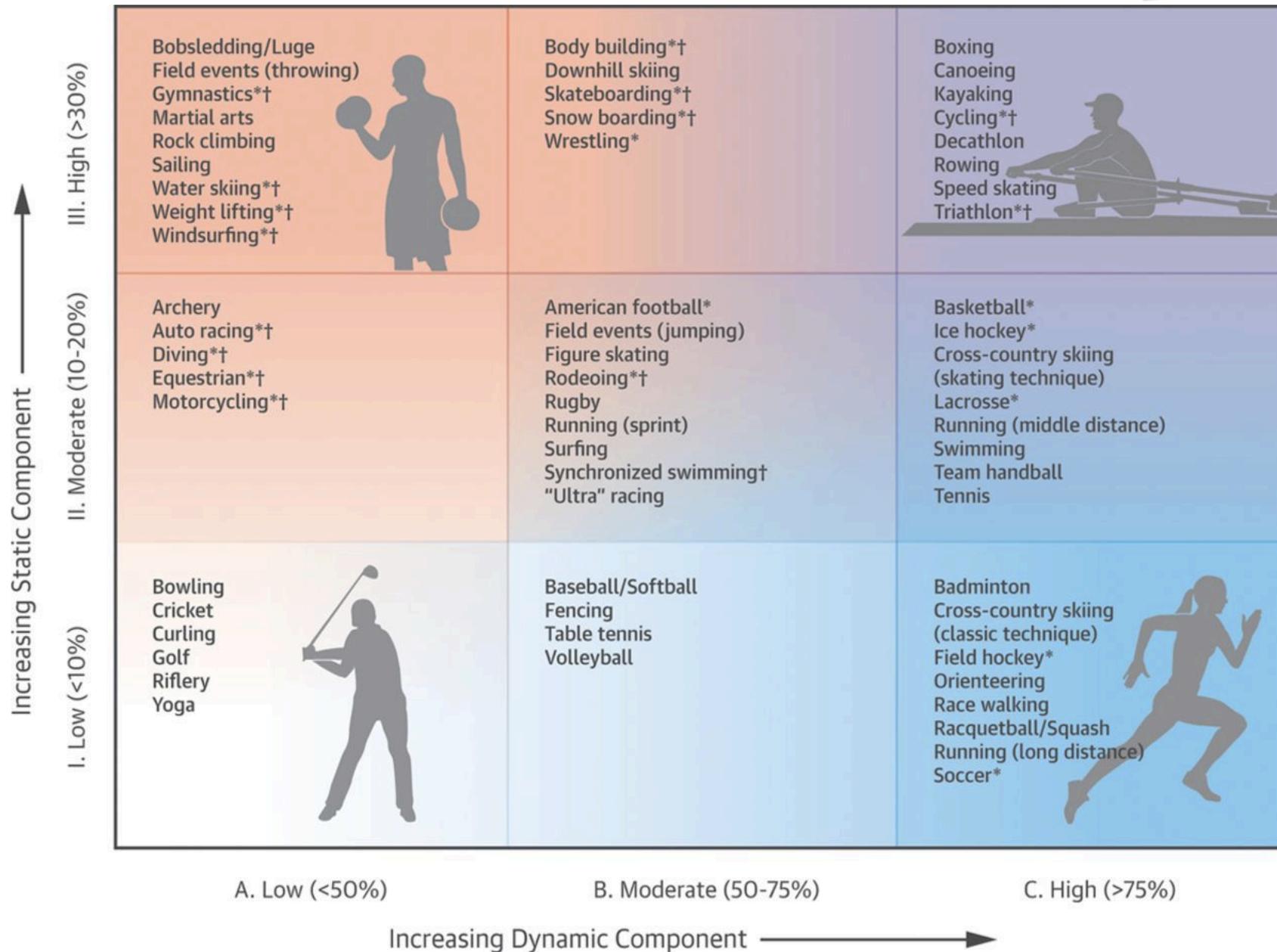
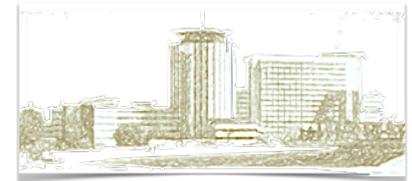


Figure 1. Frequency of sudden death in youth sport by sport and cause of death: 2007–2015. *Other* includes anaphylactic shock, lightning, drowning, exertional heat stroke, and inconclusive.





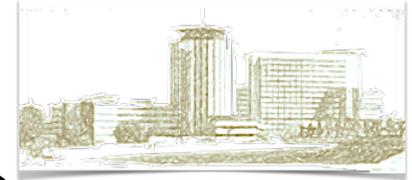
III. High (>50% MVC)	Gymnastics = 2 Sport climbing = 6 Weight lifting = 3 Total = 11 (15.9%) <i>Incidence*:</i> 0.25/100'000 (95% CI: 0.16 - 0.37)	Downhill skiing = 3 Wrestling = 3 Total = 6 (8.7%) <i>Incidence*:</i> 0.18/100'000 (95% CI: 0.10 - 0.31)	Cycling = 9 Rowing = 2 Total = 11 (15.9%) <i>Incidence*:</i> 0.25/100'000 (95% CI: 0.16 - 0.37)	
	II. Moderate (20%–50% MVC)	Auto racing = 1 Diving = 2 Equestrian = 1 Total = 4 (5.8%) <i>Incidence*:</i> 0.94/100'000 (95% CI: 0.41 - 1.84)	Total = 0 (0%) <i>Incidence*:</i> N/A	Basketball = 1 Ice hockey = 2 Cross-country skiing = 1 Swimming = 7 Team handball = 2 Total = 13 (18.8%) <i>Incidence*:</i> 0.33/100'000 (95% CI: 0.22 - 0.48)
		I. Low (<20% MVC)	Total = 0 (0%) <i>Incidence*:</i> N/A	Volleyball = 1 Total = 1 (1.5%) <i>Incidence*:</i> 0.25/100'000 (95% CI: 0.03 - 0.90)
	(A) (<40% MaxO ₂) I. Low	(B) (40%–70% MaxO ₂) II. Moderate	(C) (>70% MaxO ₂) III. High	

Figure 3. Numbers and incidences of sports-related sudden cardiac deaths in different sport categories. Classification of sports-related sudden cardiac deaths in recreational sport and competitive sport athletes based on peak static (isometric) and dynamic (isotonic) components according to the Task Force 8 of the American College of Cardiology.⁷ I to III represent increasing static components of sports; A to C represent increasing dynamic components of sports.

*Incidences are shown in “athlete person-years” and calculated based on the average autopsy rate in Switzerland (47.5%)¹³ and adjusted ($\times 2.1$) sports-related sudden cardiac death numbers.

Max O₂: maximal oxygen uptake; MVC: maximal voluntary contraction; CI: confidence interval.

Características de riesgo



- ☞ Ejercicio con **picos de actividad** (*burst*): aceleraciones y deceleraciones rápidas (sprints, baloncesto, tenis, fútbol)
- ☞ **Condiciones ambientales extremas** de temperatura humedad y altitud que afectan la volemia y los electrolitos
- ☞ Cargas de entrenamiento sistemáticas y **progresivas** para conseguir niveles altos de acondicionamiento

Fin y principio



El ejercicio es un desencadenante de ARRITMIAS MORTALES en niños predispuestos AUNQUE

☞ no sólo son las arritmias las que producen problemas durante el ejercicio

☞ muchas arritmias suceden sin ejercicio en individuos predispuestos (a veces difíciles de identificar)

☞ la mayoría de individuos predispuestos no tienen eventos durante el ejercicio

☞ no todos los ejercicios son iguales, pero a veces se parecen en el riesgo arrítmico