Mort subite et ANOCOR

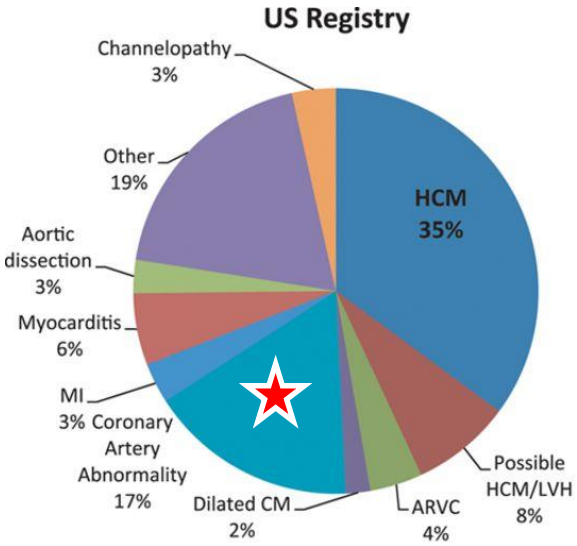
Pierre Aubry (Paris), Patrick Dupouy (Antony), Reza Farnoud (Paris), Warda Ferrag (Paris), Xavier Halna du Fretay (Aran), Fabien Hyafil (Paris), Jean-Michel Juliard (Paris), Phalla Ou (Paris), Jean-Pierre Laissy (Paris)
pour le groupe ANOCOR

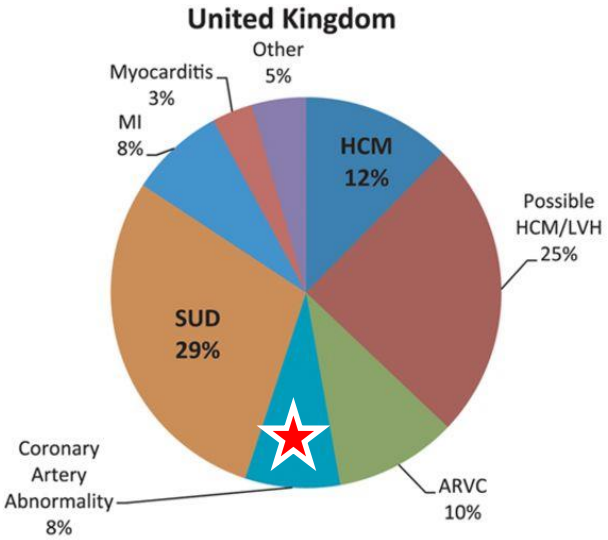
16 mars 2018

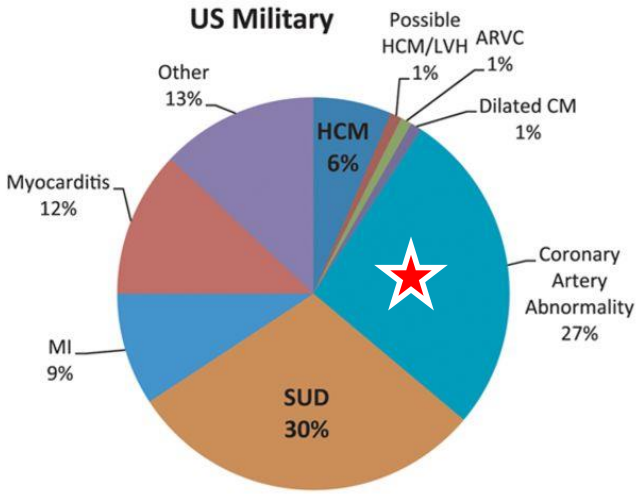
Principales questions

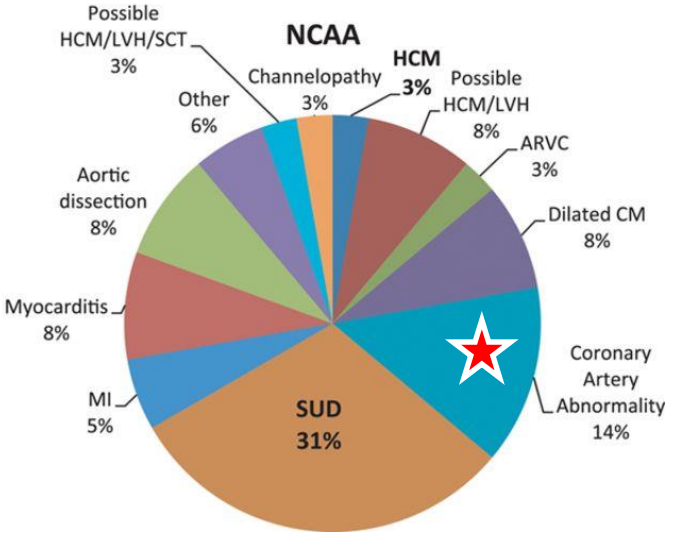
- formes anatomiques à risque de mort subite
- épidémiologie
- mécanismes de la mort subite
- évaluation du risque
- prévention du risque

sudden death in 0-35 years of age









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The Leeds Teaching Hospitals NHS
NHS Trust

Sudden Cardiac Death and Aborted SCD in Patients with Anomalous Aortic Origin of a Coronary Artery (AAOCA): A Comprehensive Review of the Literature

Dr Hunain Shiwani, BMBS
Department of Radiology
Leeds Teaching Hospitals NHS Trust

12th March 2018

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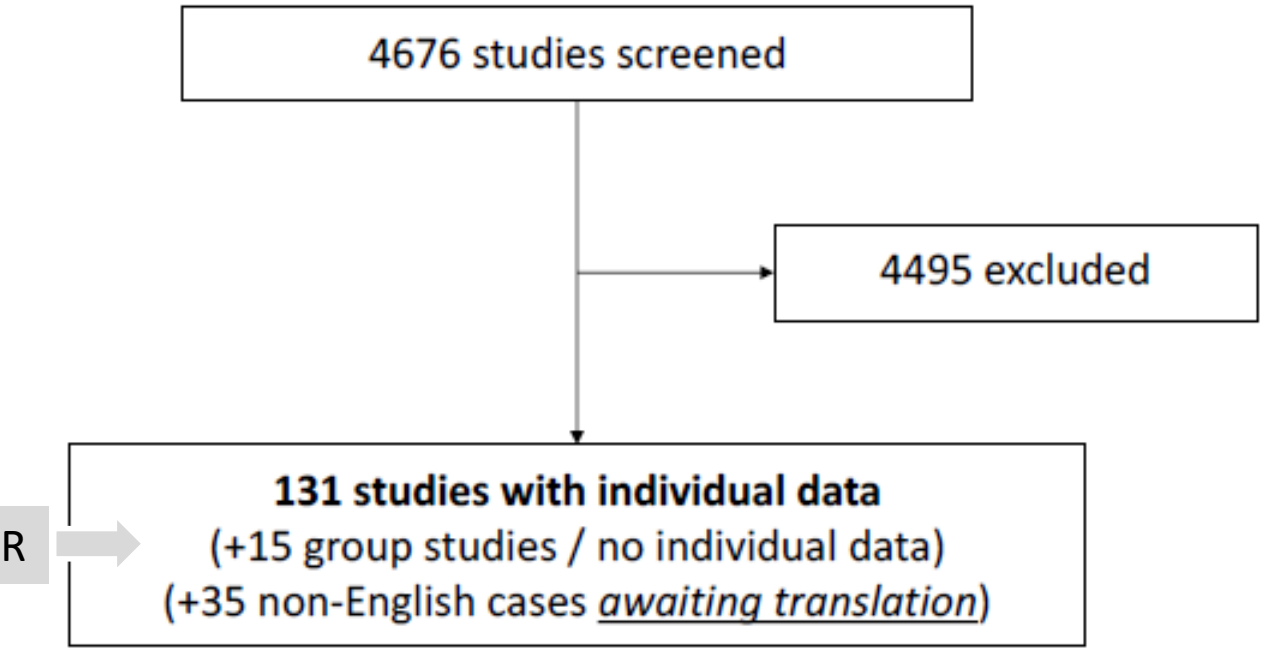
NHS

Results



Registre ANOCOR

Shiwani H. ACC 2018

sudden cardiac death and AAOCA

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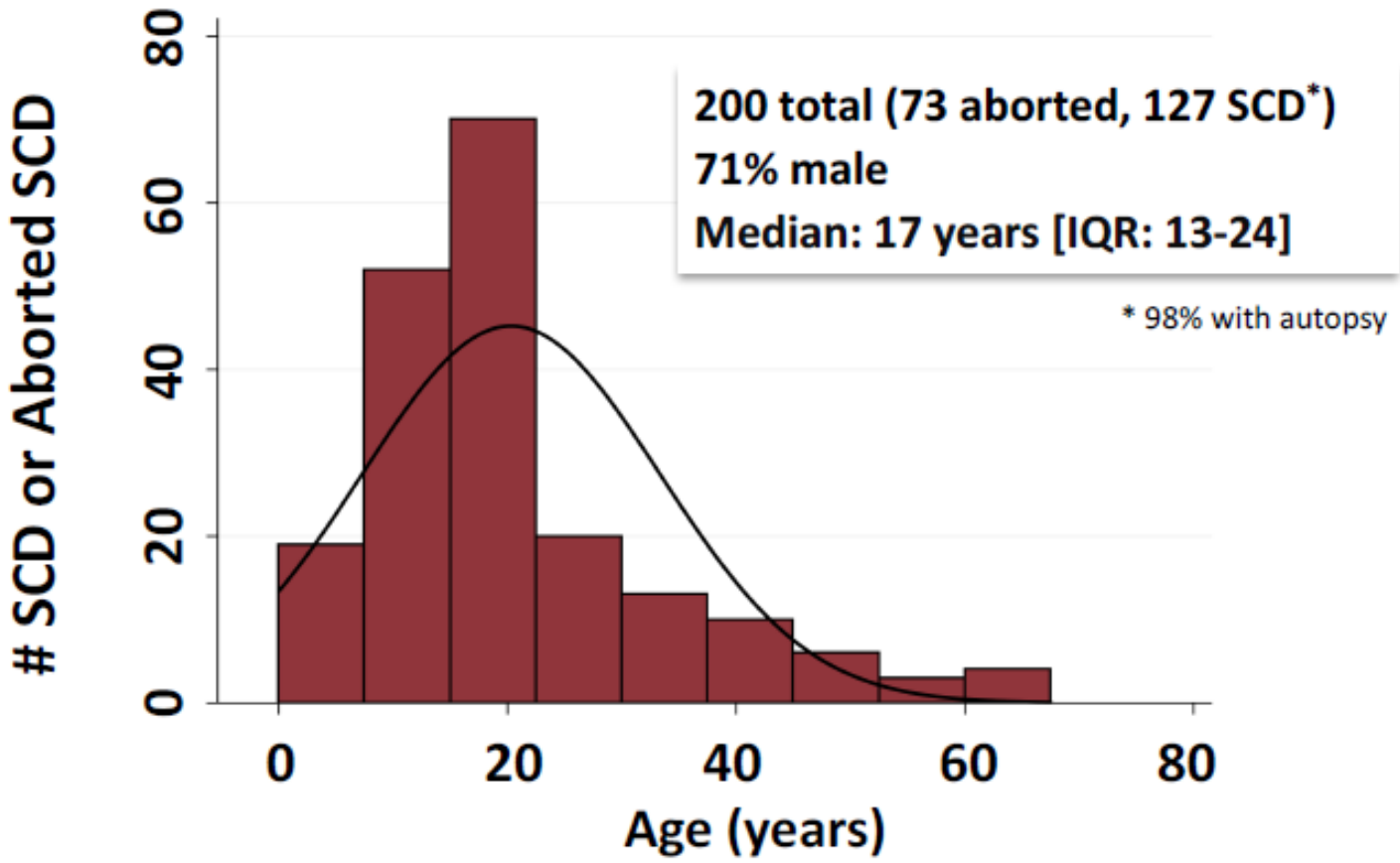
Age of SCD or Aborted SCD Attributed to AAOCA

NHS



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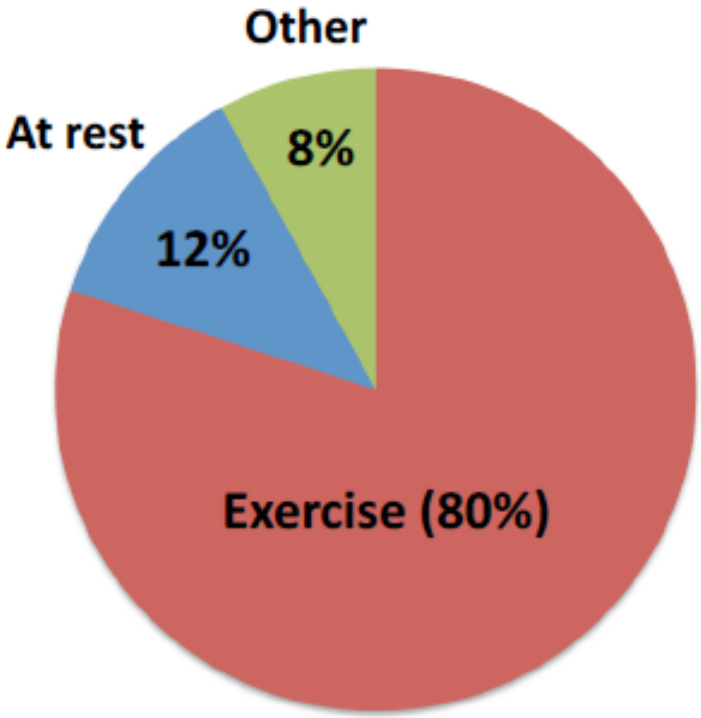
SCD/Aborted SCD Related to Exercise

NHS



- Exercise (n=142, 80%)
- At rest (n=21, 12%)
- Other (n=14, 8%):
  - 12 babies (in crib, crying spell)
  - 2 adults w/ emotional distress

* Data not available in 23 cases

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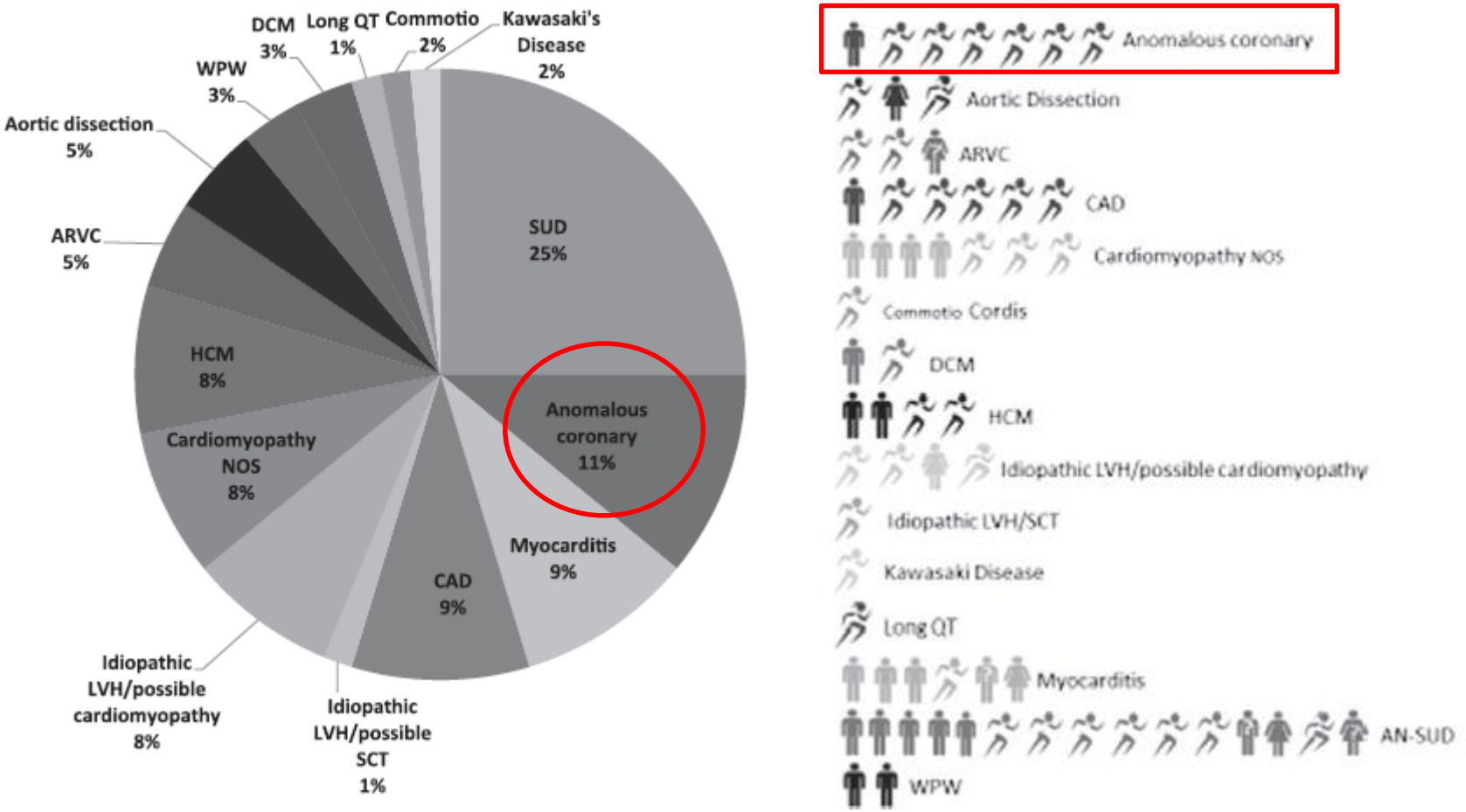
Incidence, Cause, and Comparative Frequency of Sudden Cardiac Death in National Collegiate Athletic Association Athletes

A Decade in Review



Harmon KG. Circulation 2015

Sports-Related Sudden Death in the General Population

Eloi Marijon, MD; Muriel Tafflet, PhD; David S. Celermajer, PhD, FRACP; Florence Dumas, MD; Marie-Cécile Perier, MSc; Hazrije Mustafic, MD; Jean-François Toussaint, MD, PhD; Michel Desnos, MD; Michel Rieu, MD; Nordine Benameur, MD; Jean-Yves Le Heuzey, MD; Jean-Philippe Empana, MD, PhD; Xavier Jouven, MD, PhD

- 10 -75 ans
- population avec activité sportive
- ≈ 2 cas/100 000 pratiquants/an
- ≈ 1 000 morts subites/an en France
- **≈ 60 (6%) morts subites chez le sportif compétiteur/an**
- âge moyen : 46 ± 15 ans
- hommes : 95%
- présence d'un témoin : 93%

Circulation 2015

mort subite et ANOCOR

étiologies de la mort subite chez le sportif

Etudes	âges	MS	ANOCOR	CMH	MC
Maron et al.	8-39 ans	1049	119 (17%)	251 (36%)	23 (2%)
Corrado et al.	12-35 ans	55	6 (13%)	14 (25%)	11 (20%)
Harmon et al.	17-24 ans	64	7 (11%)	13 (20%)	6 (10%)
Bohm et al.	10-79 ans	64	3 (5%)	2 (3%)	34 (53%)
Suarez-Mier et al.	9-69 ans	168	7 (4%)	19 (11%)	85 (51%)
Risgaard et al.	17-49 ans	35	1 (2%)	6 (17%)	15 (43%)
Marijon et al.	10-75 ans	199	2 (1%)	13 (7%)	152 (75%)

ANOCOR : anomalies coronaires congénitales
CMH : cardiomyopathie hypertrophique
MC : maladie coronaire
MS : mort subite

Aubry et al. Ann Angeio Cardio 2017

registry
n=472

12 aborted SCD (2.5%)

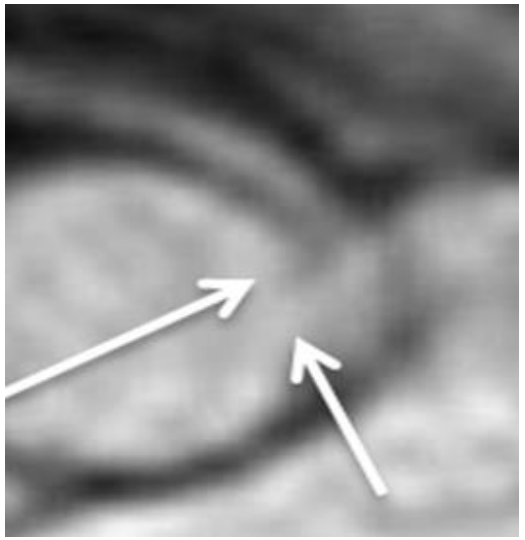
3 ANOCOR-related SCD (0.6%)

number	age	artery	connection	course	significant CAD
1	50	Cx	contralateral artery	retroaortic	present
2	75	Cx	contralateral artery	retroaortic	present
3	72	Cx	contralateral artery	retroaortic	present
4	16	LM	pulmonary artery	normal	absent
5	53	Cx	contralateral artery	retroaortic	present
6	48	Cx	contralateral artery	retroaortic	absent
7	57	CX	contralateral artery	retroaortic	present
8	60	RCA	ascending aorta	preaortic	present
9	31	RCA	contralateral sinus	preaortic	absent
10	60	RCA	contralateral sinus	preaortic	present
11	30	RCA	contralateral sinus	preaortic	absent
12	44	CX	contralateral sinus	retroaortic	absent

Aubry P. Arch Cardiovasc Dis 2016

ACAOS prevalence in general population

MRI-based study (n = 5.255)
middle school children (mean age 13 years)
2010-2017



L-ACAOS-IM = 2
R-ACAOS-IM= 17
Total ACAOS = 19

Prevalence L-ACAOS = 0.04%
Prevalence R-ACAOS = 0.32%
Total prevalence = 0.35% ≈ 3.5/1000

AMERICAN COLLEGE of CARDIOLOGY

Angelini P. ACC 2017

CHD with risk of sudden cardiac death (estimation)

CONGENITAL HEART DISEASE	PREVALENCE cases per 100 000
Anomalous connection of right coronary artery	**300 (0.30%)**
Hypertrophic cardiomyopathy	200 (0.20%)
Wolf-Parkinson-White syndrome	150 (0.15%)
Long QT syndrome	50 (0.05%)
Anomalous connection of left coronary artery	40 (0.04%)
Idiopatic dilated cardiomyopathy	40 (0.04%)
Arrhythmogenic right ventricular cardiomyopathy	40 (0.04%)
Brugada syndrome	20 (0.02%)
Catecholaminergic polymorphic ventricular tachycardia	10 (0.01%)

prévalence angiographique des anomalies à risque

connexion ectopique avec trajet interartériel

cohorte ANOCOR*

472 patients ≥ 15 ans - 496 ANOCOR

2010 - 2013

135 ANOCOR à risque

ANOCOR gauches	=	6	5%
ANOCOR droites	=	129	95%

*Aubry P et al. Anomalous connections of the coronary arteries: a prospective observational cohort of 472 adults. The ANOCOR registry. Eur Heart J 2015;36 suppl 1:1138.

mort subite et ANOCOR

MRI-based study
n = 5 255
mean age 13 years

1 L-ACAOS

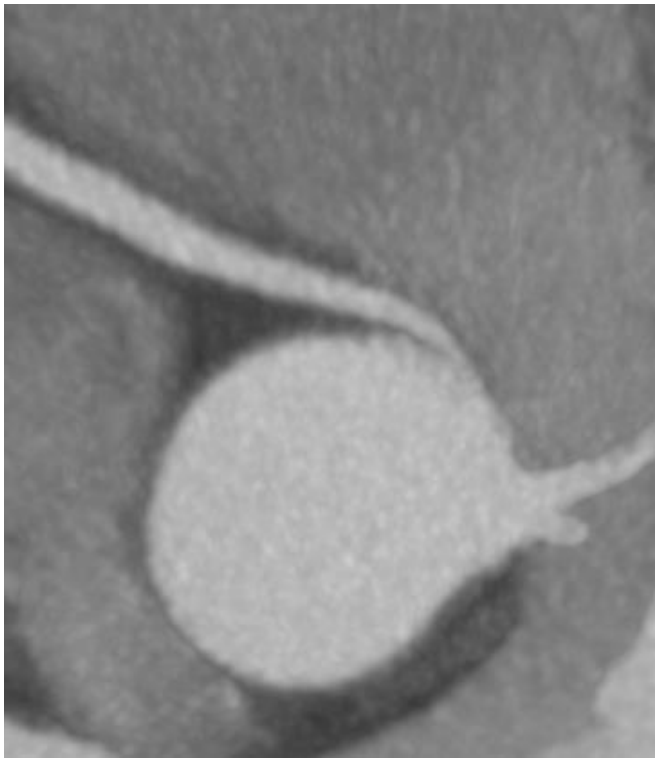
8 R-ACAOS

Registre ANOCOR
n = 472
âge moyen 63 ans

1 ANOCOR-G

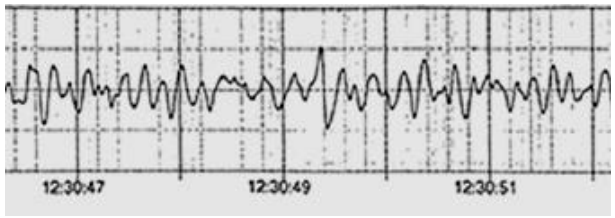
21 ANOCOR-D

mécanisme(s) de la fibrillation ventriculaire

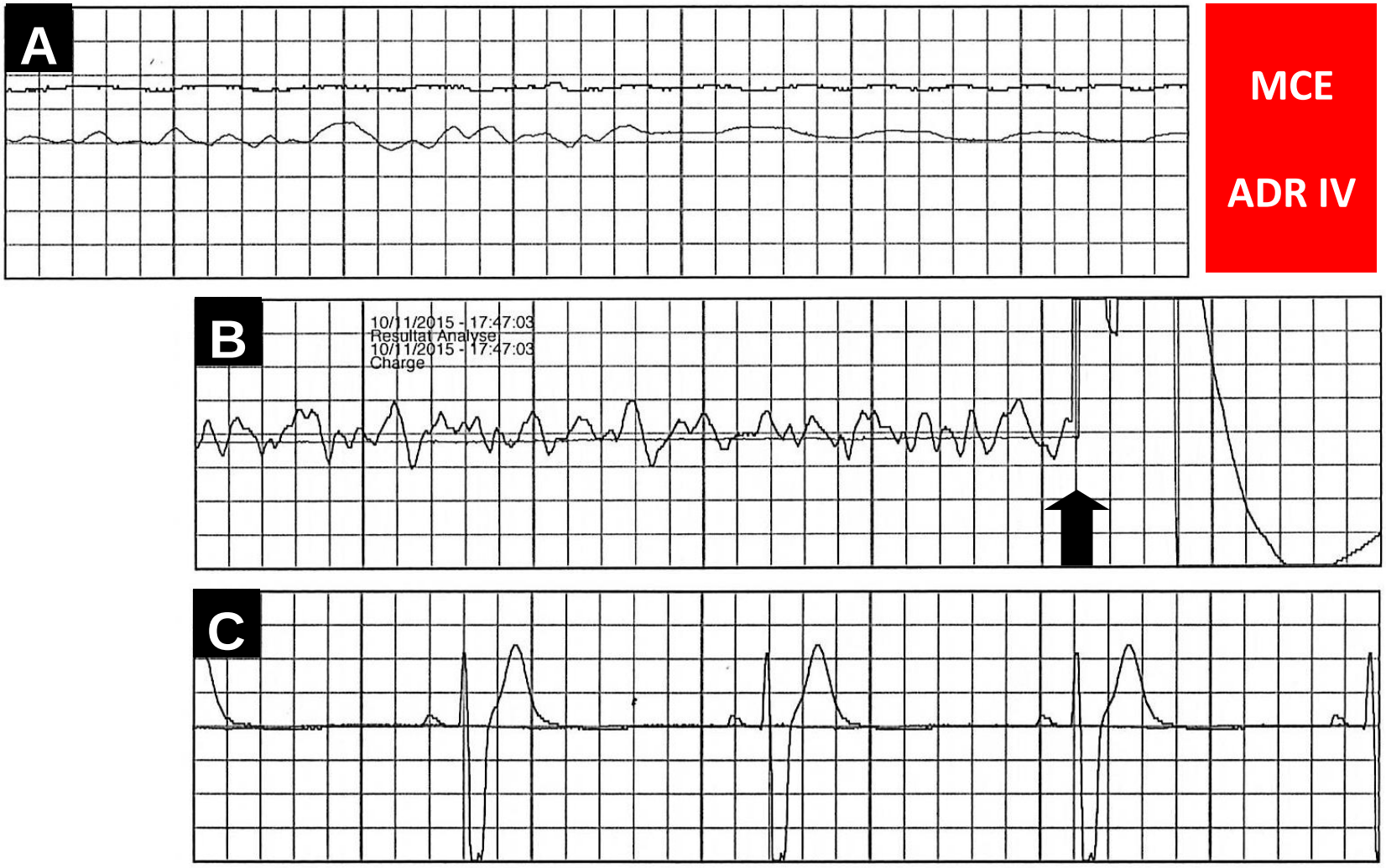


?

24/07/2012
vers 12.30



mort subite et ANOCOR



jeune garçon de 15 ans - activité sportive scolaire - arrêt cardiaque récupéré
ANOCOR gauche

mécanisme(s) de la fibrillation ventriculaire

- ischémie myocardique
- zones de fibrose myocardique
- seuil arythmogène bas
- hypotension post-effort
- association de plusieurs mécanismes
- substrat-gâchette-modulateurs (triangle de Coumel)
- association fortuite
- …

sudden cardiac death and AAOCA

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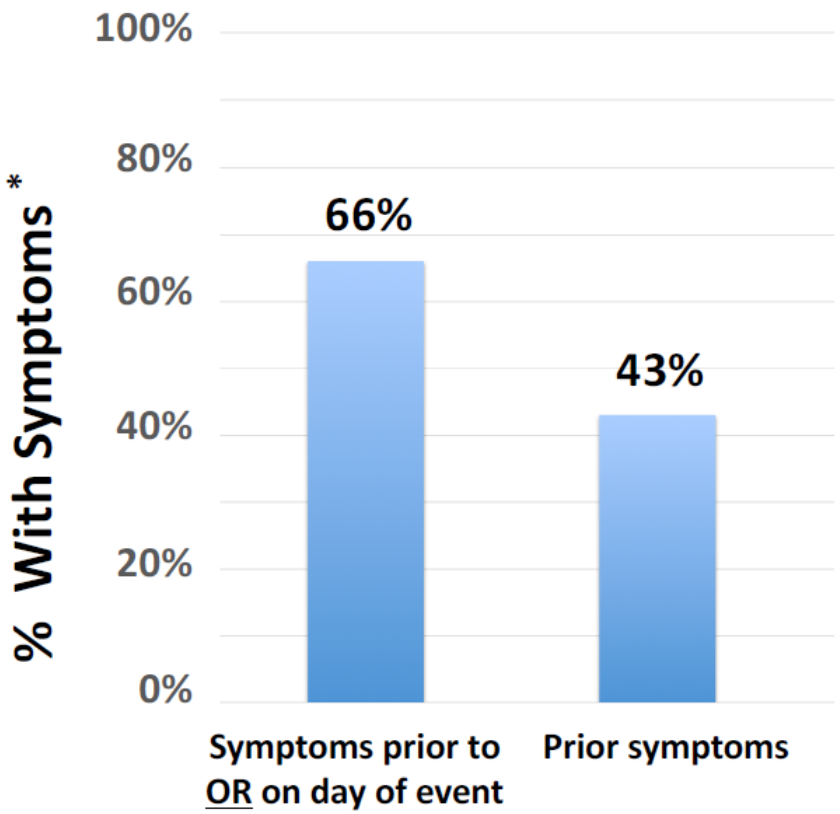
Cardiac Symptoms

NHS

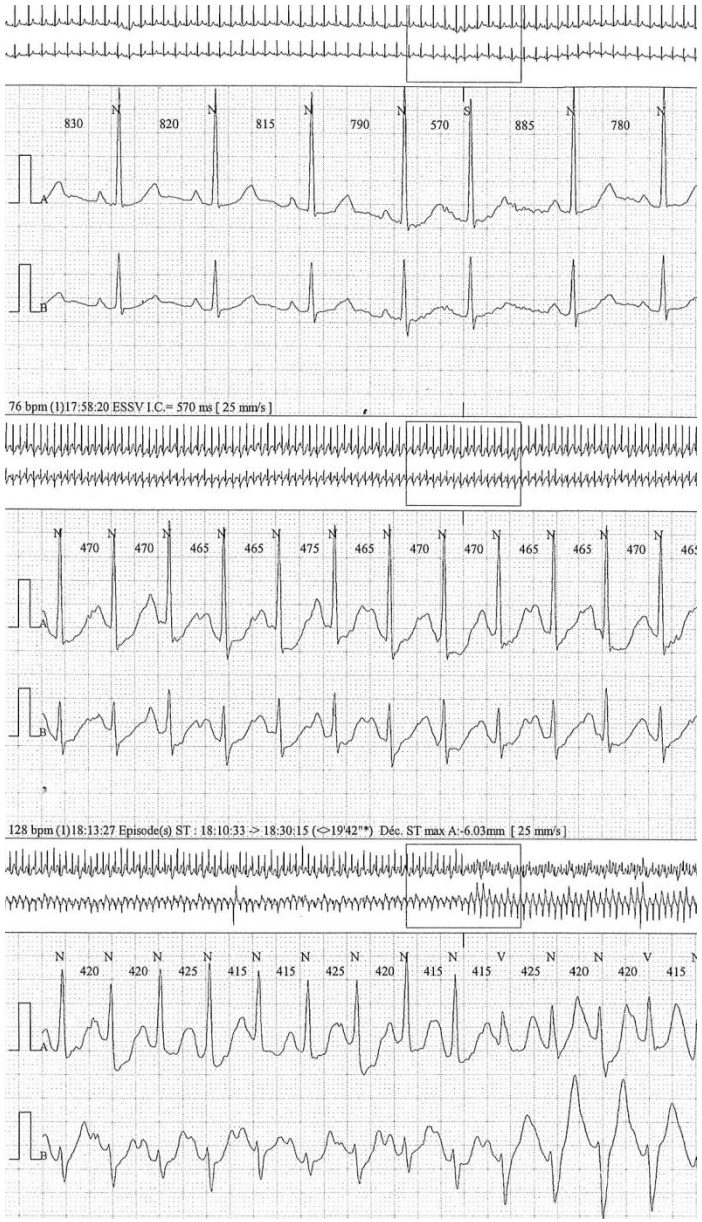


In patients with symptoms^:

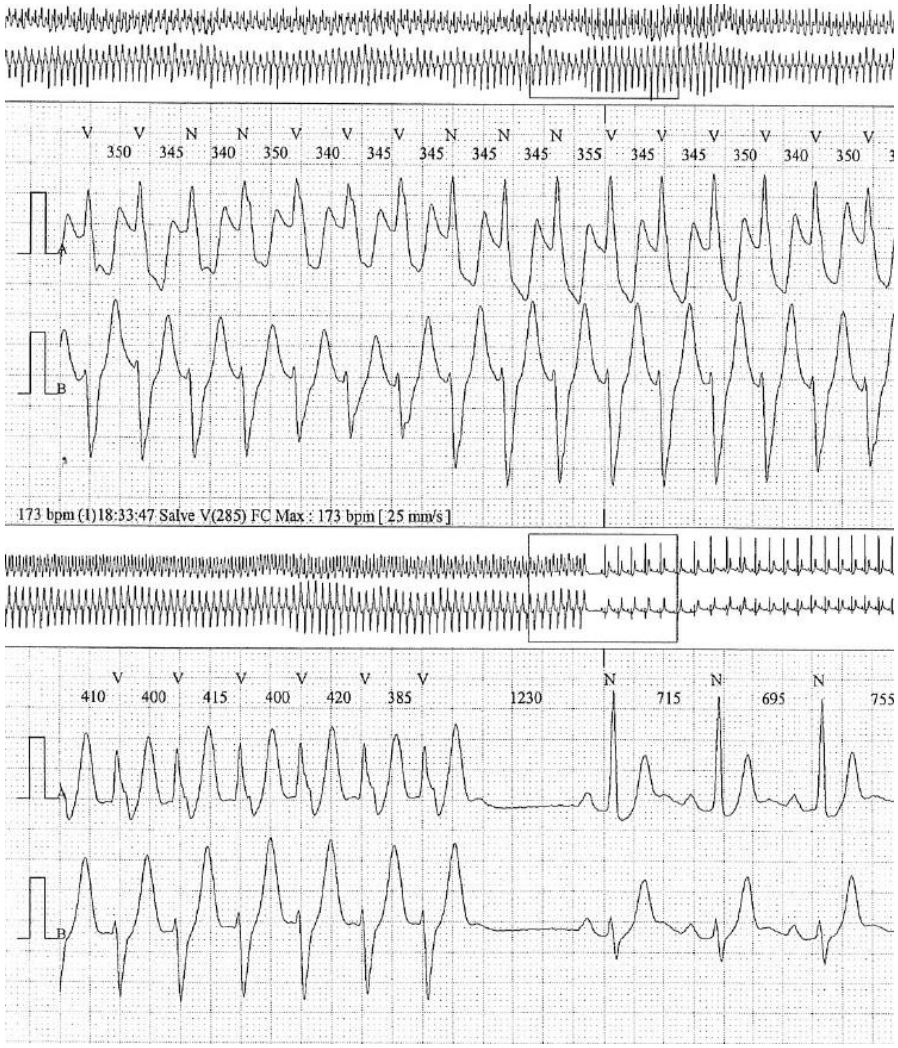
- 45% syncope/presyncope
- 41% chest pain
- 10% dyspnea
- 9% palpitations

^ 8 reported multiple symptoms; * 41 omitted (no data)

Shiwani H. ACC 2018



femme de 53 ans - palpitations (jogging)
ANOCOR droite



2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC)

Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC)

anomalies de connexion coronaire : **non citées**

Priori SG. Eur Heart J 2015

2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death: Executive Summary

4.3. Surgery and Revascularization Procedures in Patients With Ischemic Heart Disease

Recommendations for Surgery and Revascularization Procedures in Patients With Ischemic Heart Disease

References that support the recommendations are summarized in Online Data Supplement 11.

COR	LOE	Recommendations
I	B-NR	1. Patients with sustained VA and survivors of SCA should be evaluated for ischemic heart disease, and should be revascularized as appropriate (1-4).
I	C-EO	2. In patients with anomalous origin of a coronary artery suspected to be the cause of SCA, repair or revascularization is recommended.

SCA: sudden cardiac arrest

Al-Khatib et al. Circulation 2017

question du patient : à quel risque suis-je exposé ?

$$\text{fréquence annuelle} = \frac{\text{n événements}}{\text{100 patients}^*}$$

*patients ayant une anomalie de connexion coronaire identifiée à risque de mort subite

mort subite et ANOCOR

- 12-35 years athletic population
- follow-up period of 26 years
- 2.938.270 person-years of observation
- 55 deaths
- 91% during sports activity

- 1.9 deaths/100.000 person-years
- 0.48 deaths/100.000 person-years (cardiomyopathy)
- 0.24 deaths/100.000 person-years (coronary anomaly)

Corrado D. JAMA 2006

sudden cardiac death and AAOCA

annual risk of sudden cardiac death (estimation)
in population with ages 12-35 years

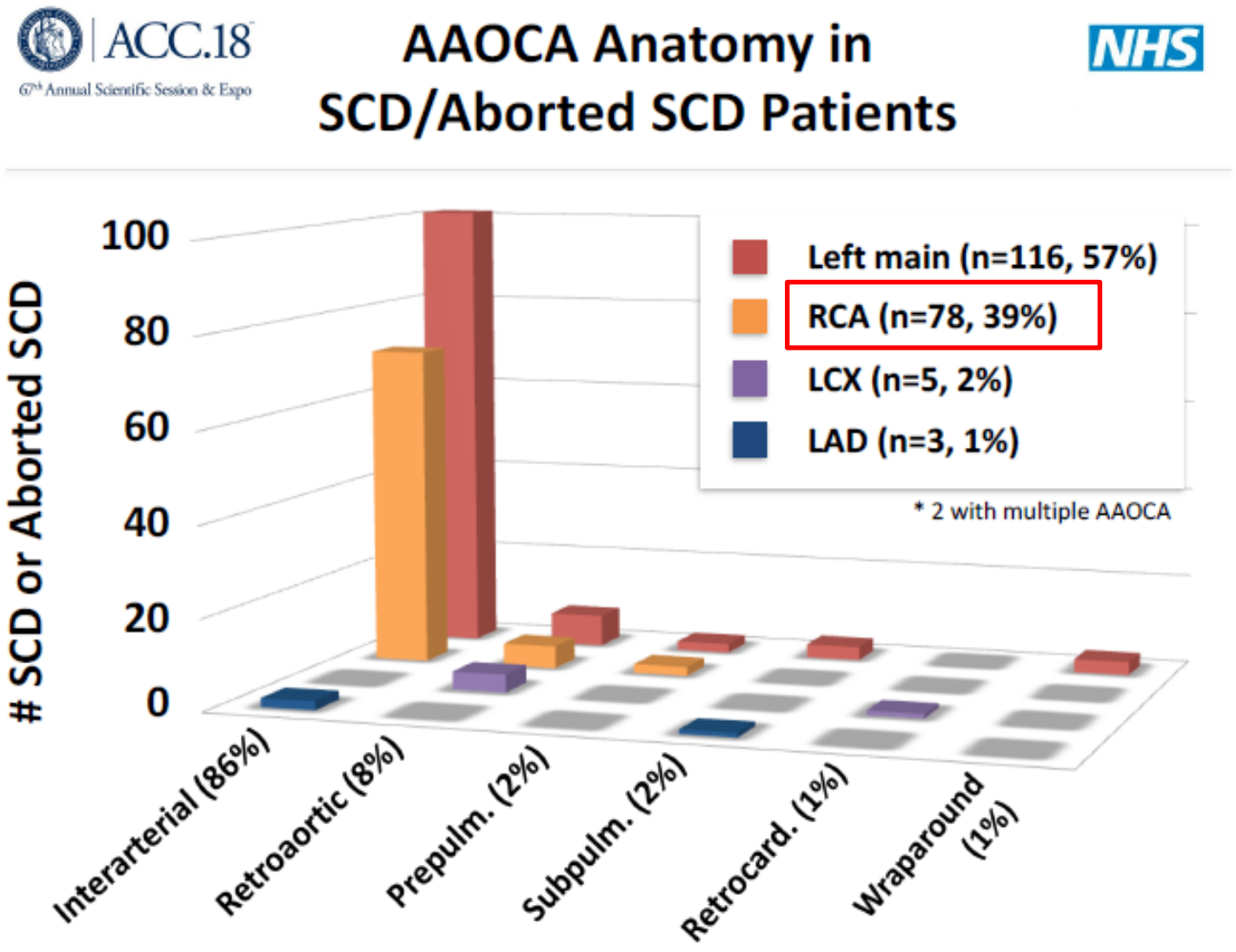
L-ACAOS	**0.3%**
R-ACAOS	**0.01%**

Brothers J. J Thorac Cardiovasc Surg 2010

CHD with risk of sudden cardiac death (estimation)

CONGENITAL HEART DISEASE	SCD cases per 100 000/year
Catecholaminergic polymorphic ventricular tachycardia	1500
Hypertrophic cardiomyopathy	1000
Brugada syndrome	1000
Long QT syndrome	500-1000
Idiopatic dilated cardiomyopathy	500-1000
Arrhythmogenic right ventricular cardiomyopathy	500-1000
Wolf-Parkinson-White syndrome	100
Anomalous connection of left coronary artery	100
Anomalous connection of right coronary artery	5

sudden cardiac death and AAOCA



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Une question

- mort subite ischémique ?
- mort subite rythmique ?