

Pediatric Sudden Cardiac Death

Sep 19th, 2026

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Seattle Children's Tri-Cities Clinic

OUTLINE

- ❖ Epidemiology
- ❖ Pre-participation Physicals
- ❖ Causes of Sudden Cardiac Death

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Epidemiology

The incidence of SCA in the young is difficult to ascertain:

- No mandatory reporting system for juvenile sudden death
- Most data for pediatric SCD are collected postmortem, such that many cases of SCA or aborted SCD are often not reported

~ 0.5 to 20 per 100,000 person-years

Epidemiology

- ~ 2000 patients younger than 25 years will die from a sudden cardiac event each year in the United States
- The incidence is higher in athletes, with a relative risk of sudden death of 2.1 compared with nonathletes

Epidemiology

- The prevalence of sudden unexpected death, in general, and SCD, in particular, is age dependent:
 - Higher risk in early infancy (SIDS)
 - Falls in early childhood
 - Rise again in adolescence

Sudden Cardiac Death in the Young. Ackerman et al. Circulation. 2016;133:1006-1026

Epidemiology

One large study identifying 114 cases of SCD in children ages 1 to 18 in Denmark

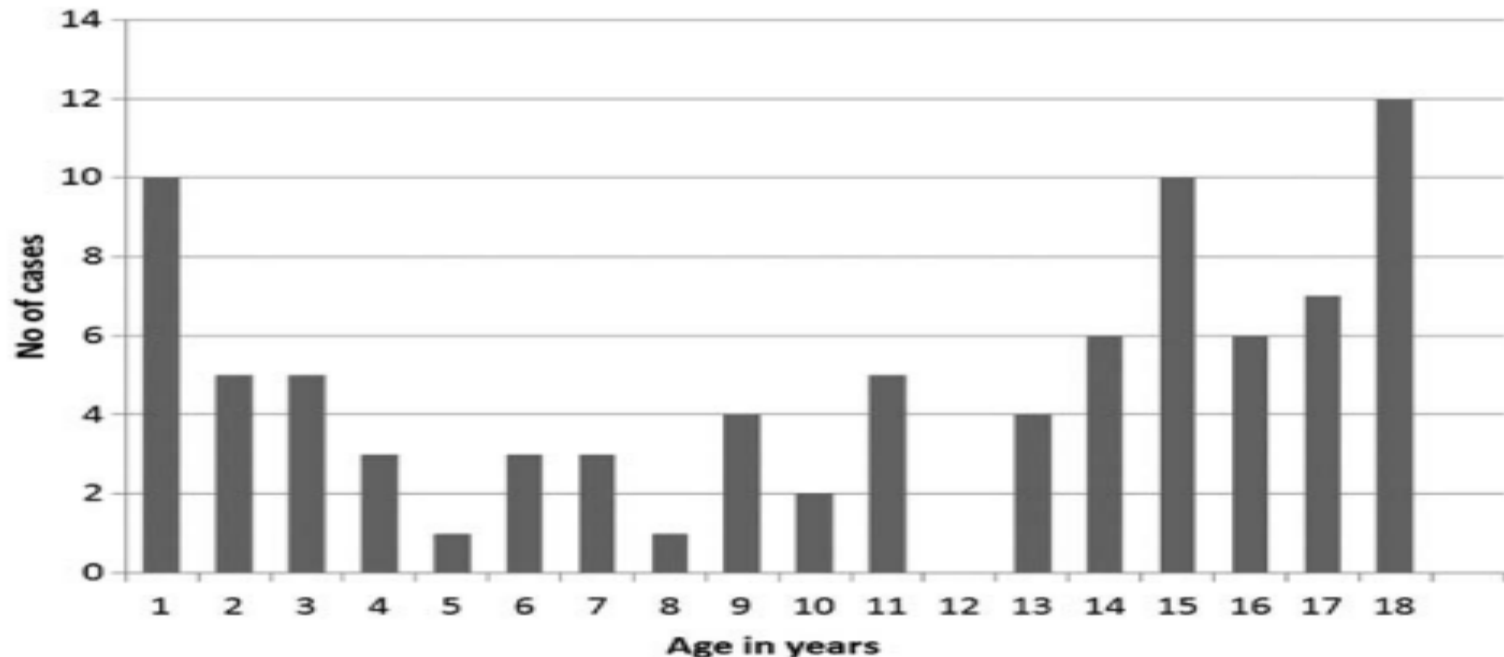


Figure 2. Age distribution of SCD in the Danish population, 2000 to 2006. SCD indicates sudden cardiac death. Reprinted from Winkel et al³³ with permission of the publisher. Copyright © 2014, European Society of Cardiology.

Epidemiology

- Sudden cardiac death (SCD) is statistically uncommon in the young, but its dramatic presentation and cascading effects in the family and community make it a newsworthy event
- High degree of concern regarding the risk of SCD in children and teenagers among the lay public, in comparison with other much more prevalent deadly risks of childhood, such as accident, injury, suicide, and violence

Sudden Cardiac Death in the Young. Ackerman et al. Circulation. 2016;133:1006-1026

Epidemiology

Mortality Rates Among Children Aged 1–14, by Selected Leading Cause and Age, 2010

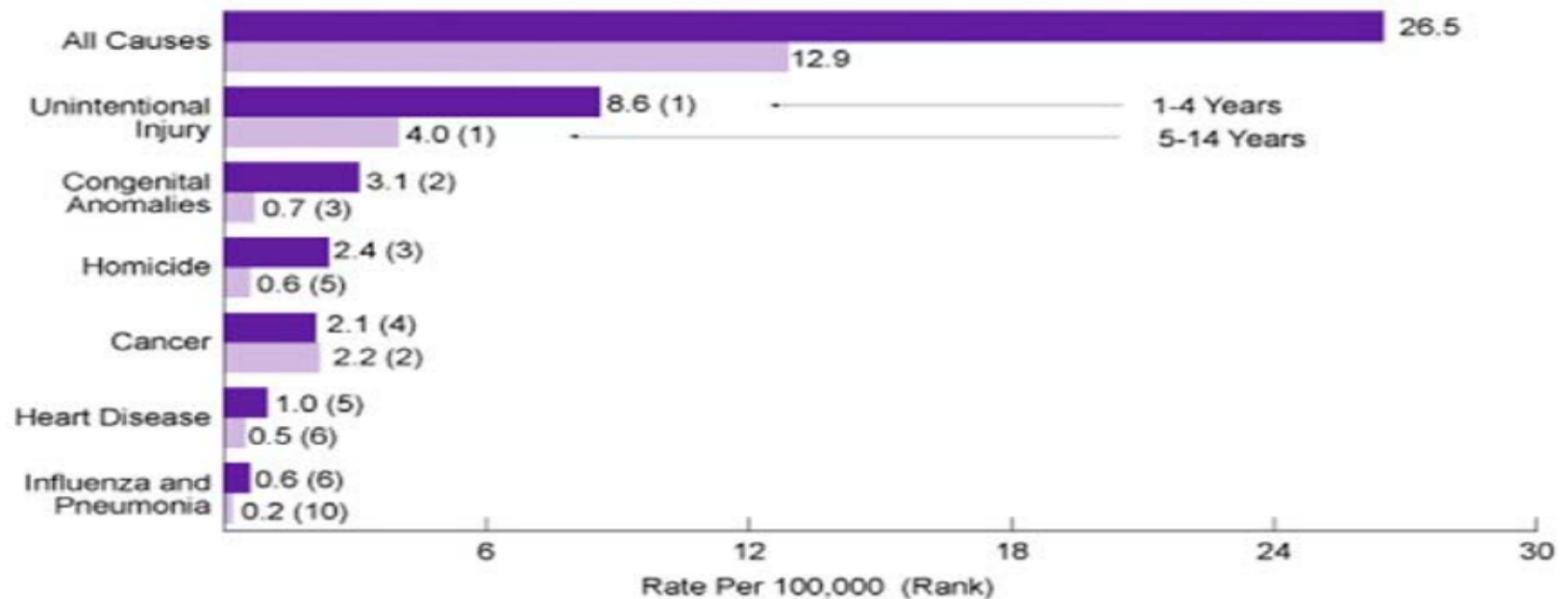


Figure 1. Causes of death in American children. Mortality rates among children 1 to 14 years of age, by selected leading cause and age, 2010.

Epidemiology

Circulation

JOURNAL OF THE AMERICAN HEART ASSOCIATION



Sudden Deaths in Young Competitive Athletes : Analysis of 1866 Deaths in the United States, 1980 –2006

Barry J. Maron, Joseph J. Doerer, Tammy S. Haas, David M. Tierney and Frederick O. Mueller

Circulation 2009, 119:1085-1092: originally published online February 16, 2009
doi: 10.1161/CIRCULATIONAHA.108.804617

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Sudden Deaths in Young Competitive Athletes. Maron et al. *Circulation* 2009, 119:1085-1092



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Epidemiology

- **U.S. Competitive Athlete Registry (N = 1,866)**
- Followed over 27 years (1980–2006); Mean age 18 ± 5 years
- **Overall Causes of Sudden Death:**
 - Cardiovascular disease (1049 [56%])
 - Blunt trauma that caused structural damage (416 [22%])
 - Commotio cordis (65 [3%])
 - Heat stroke (46 [2%])
- **Leading CV Etiologies:**
 - Hypertrophic cardiomyopathy (36%)
 - Congenital coronary artery anomalies (17%)

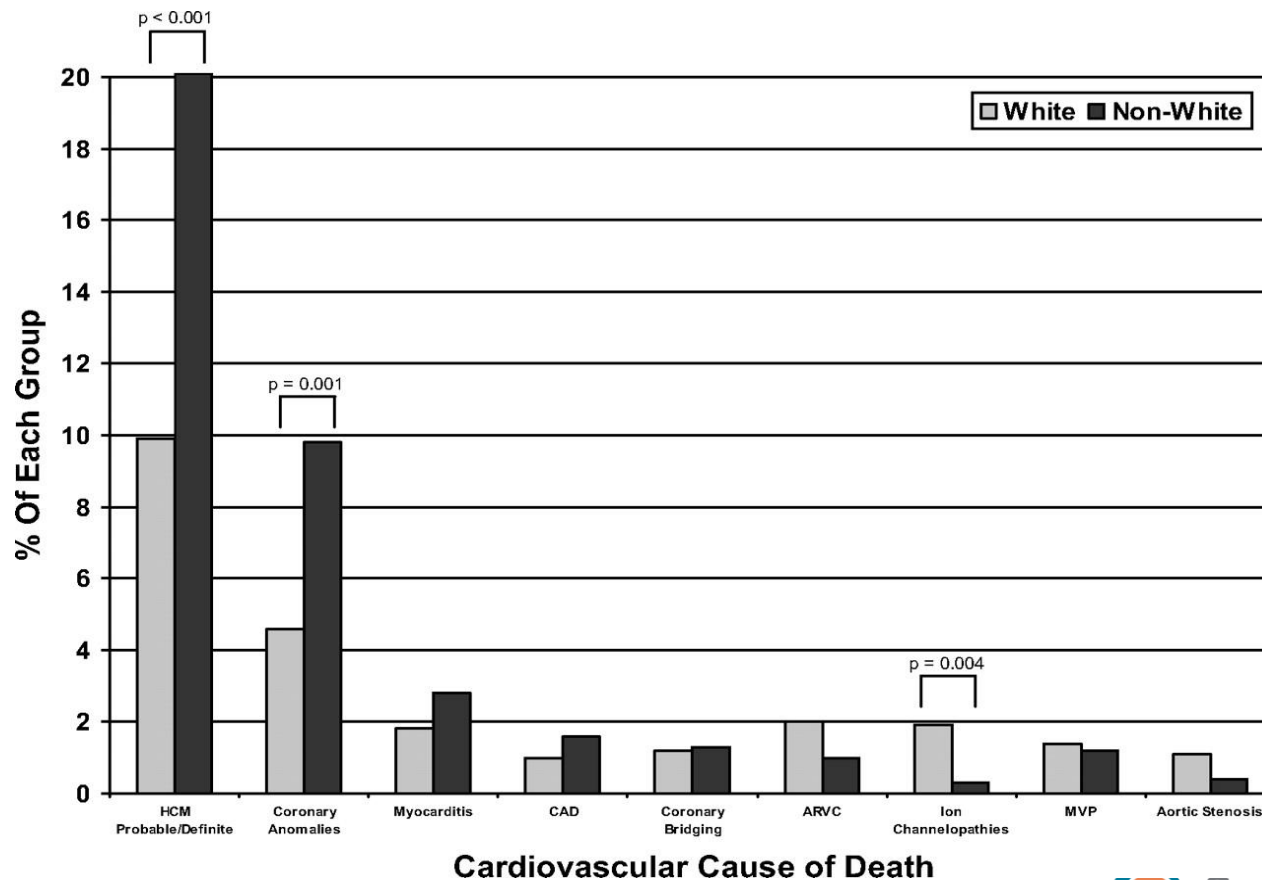
Epidemiology

❖ High-Risk Demographics:

- **Age Distribution:** Mean 18 ± 5 yrs (range 8–39 yrs); 94% ≤ 25 yrs
- **Sex Predominance:** 89% Male vs 11% Female (female proportion rising to 12% in recent years)
- **Top Sports:** Basketball (33%) & Football (25%) account for ~58% of events.
- **Exertional Onset:** 80% occurred during or immediately after physical exertion.

Epidemiology

❖ Cardiovascular deaths were significantly more common in Non-white vs White athletes (64% vs 51%, $p < 0.001$)



OUTLINE

- ❖ Epidemiology
- ❖ **Pre-participation Physicals**
- ❖ Causes of Sudden Cardiac Death

Pre-Participation Physicals

POLICY STATEMENT Organizational Principles to Guide and Define the Child Health Care System and/or Improve the Health of all Children

American Academy
of Pediatrics



DEDICATED TO THE HEALTH OF ALL CHILDREN™

Sudden Death in the Young: Information for the Primary Care Provider

Christopher C. Erickson, MD, FAAP,^{a,b} Jack C. Salerno, MD,^c Stuart Berger, MD, FAAP,^d Robert Campbell, MD, FAAP,^e Bryan Cannon, MD,^f James Christiansen, MD,^c Kody Moffatt, MD, MS, FAAP,^a Andreas Pflaumer, MD,^g Christopher S. Snyder, MD, FAAP,^h Chandra Srinivasan, MD,ⁱ Santiago O. Valdes, MD, FAAP,^j Victoria L. Vetter, MD, MPH, FAAP,^k Frank Zimmerman, MD,^l SECTION ON CARDIOLOGY AND CARDIAC SURGERY, PEDIATRIC AND CONGENITAL ELECTROPHYSIOLOGY SOCIETY (PACES) TASK FORCE ON PREVENTION OF SUDDEN DEATH IN THE YOUNG



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Pre-Participation Physicals

The Sports Preparticipation Evaluation

Shane M. Miller, MD, FAAP,* Andrew R. Peterson, MD, MSPH, FAAP[†]

**Departments of Orthopaedics and Pediatrics, UT Southwestern, and Texas Scottish Rite Hospital for Children Sports Medicine Center, Dallas, TX*

[†]Departments of Pediatrics and Orthopaedics/Rehabilitation, University of Iowa, Iowa City, IA

Preparticipation Cardiac Evaluation from the Pediatric Perspective



Andrew M. Reittinger, MD^a, Lanier B. Jackson, MD^b, Peter N. Dean, MD^{a,*}



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Pre-Participation Physicals

- 1. WHAT IS THE PURPOSE OF PREPARTICIPATION EVALUATION?**
- 2. WHO SHOULD BE EVALUATED AND WHEN SHOULD IT START?**
- 3. WHERE DO PREPARTICIPATION EVALUATIONS OCCUR?**
- 4. WHAT ARE THE COMPONENTS OF THE PREPARTICIPATION EVALUATION?**

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Pre-Participation Physicals

1. WHAT IS THE PURPOSE OF PREPARTICIPATION EVALUATION?

- To identify individuals who may be at risk for adverse health effects such as illness or injury secondary to sports participation
- To catch underlying cardiac diseases that predispose individuals to SCA and SCD

Pre-Participation Physicals

Table 1

Possible differences between pediatric and young adult/adult preparticipation cardiac screening

Pediatric Specific Screening	Young Adult and Adult Screening
More likely to have vague or unclear symptoms	Better able to communicate symptoms
Likely requires repeat screening as adolescent and/or young adult	More likely to have already expressed the phenotype of specific cardiac disease
Parents too young to demonstrate phenotype	Older parents with more detail about family history
Pediatric providers are less experienced in ECGs	Family physicians and adult providers more experienced interpreting ECGs
More likely to have undiagnosed congenital heart disease	Less likely to have undiagnosed congenital heart disease
More likely to have history of Kawasaki disease	
Suboptimal criteria for ECG abnormalities	

Pre-Participation Physicals

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Pre-Participation Physicals

2. WHO SHOULD BE EVALUATED AND WHEN SHOULD IT START?

American Academy of Pediatrics (AAP) policy statement on the topic of screening for sudden death:

- Recommend screening patients every 2- 3 years without any differentiation between athlete and nonathlete
- It also suggests that screening should be performed starting at age six

Miller SM, Peterson AR. The sports preparticipation evaluation practice gaps. Available at: <http://pedsinreview.aappublications.org>

Pre-Participation Physicals

2. WHO SHOULD BE EVALUATED AND WHEN SHOULD IT START?

- Athletes should be encouraged to complete their PPE at least 6 weeks before the first practice
- The beginning of summer or near the end of the previous school year is an ideal time
- A comprehensive PPE with a complete personal and family history and thorough physical examination should be performed at least every 2 years, with an annual review of the patient's history and a problem-focused examination as needed

Miller SM, Peterson AR. The sports preparticipation evaluation practice gaps. Pedsinreview. 2019

Pre-Participation Physicals

2. WHO SHOULD BE EVALUATED AND WHEN SHOULD IT START?

- ❖ Providers need to ask questions about physical activity, cardiac symptoms with exertion, and family history regardless of whether there is an official “school preparticipation form” to sign



Pre-Participation Physicals

1. WHAT IS THE PURPOSE OF PREPARTICIPATION EVALUATION?
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Pre-Participation Physicals

3. WHERE DO PREPARTICIPATION EVALUATIONS OCCUR?

- ❖ Ideally, these visits would occur with the patient's primary care provider, but there are other settings where evaluations occur, including mass screening events or urgent care offices
- ❖ The preferred provider is the patient's primary care provider
 - This allows for centralization of a patient's care, access to patient's past medical records, better understanding of the patient's medical, family, and social histories, and the ability to have consistent longitudinal follow-up
 - It also provides appropriate follow-up of referrals to ensure they are performed, and recommendations are followed

Pre-Participation Physicals

- ❖ **Mass screening events:** time efficient and allow for improved access to medical providers, but:
 - Difficulty maintaining patient privacy and a safe space for patient to bring concerns to physician
 - Suboptimal environment for physical examination
 - Lack of access to patient's medical records
 - Lack of time and space for appropriate counseling when an abnormality or suspected abnormality is found
 - Difficulty ensuring appropriate follow-up/referrals when abnormalities are found.

Pre-Participation Physicals

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Pre-Participation Physicals

❖ WHAT ARE THE COMPONENTS OF THE PREPARTICIPATION EVALUATION?

- Typically, PPEs are based on the PPE monograph that has been developed and endorsed by the AAP, American Academy of Family Physicians, the American College of Sports Medicine, and American Medical Society for Sports Medicine for the past 40 years (AMSSM)
- American Heart Association (AHA) 14-point questionnaire also has a similar format and questions
- The PPE has three, and possibly four, main parts: history, family history, and examination and sometimes an ECG

Pre-Participation Physicals

WHAT ARE THE COMPONENTS OF THE PREPARTICIPATION EVALUATION?

1. History:

- Past medical history
 - Cardiac symptoms with exercise
 - Odd or suspicious symptoms: seizures, syncope
 - Any known or previously diagnosed cardiac pathology, including congenital heart disease, cardiac arrhythmia or channelopathies, cardiomyopathy, history of myocarditis, or coronary artery anomalies, including those caused by KD, should be evaluated by a cardiologist before clearance.
- Studies have shown that complete agreement between athlete and parental reports of medical history on the PPE is less than 20%. This highlights the importance of involving parents or primary caregivers in the history

Pre-Participation Physicals

The AAP Policy statement recommended 4 questions directed toward SCA and SCD detection:

1. Have you ever fainted, passed out, or had an unexplained seizure suddenly and without warning, especially during exercise or in response to sudden loud noises, such as doorbells, alarm clocks, and ringing telephones?
2. Have you ever had exercise-related chest pain or shortness of breath?
3. Has anyone in your immediate family (parents, grandparents, siblings) or other, more distant relatives (aunts, uncles, cousins) died of heart problems or had an unexpected sudden death before age 50? This would include unexpected drownings, unexplained auto crashes in which the relative was driving, or sudden infant death syndrome (SIDS).
4. Are you related to anyone with HCM or hypertrophic obstructive cardiomyopathy, Marfan syndrome, arrhythmogenic cardiomyopathy, LQTS, short QT syndrome, Brugada Syndrome, or catecholaminergic polymorphic ventricular tachycardia or anyone younger than 50 years with a pacemaker or implantable defibrillator?

Miller SM, Peterson AR. The sports preparticipation evaluation practice gaps. Available at: <http://pedsinreview.aappublications.org>



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Pre-Participation Physicals

WHAT ARE THE COMPONENTS OF THE PREPARTICIPATION EVALUATION?

2. Physical Examination:

Vital signs:

- Height, weight, and BMI
- Blood pressure
 - Elevated blood pressure is common. It can be a sign of underlying cardiac disease (eg, coarctation of the aorta) or undiagnosed hypertension
 - If a patient has upper extremity hypertension, careful attention should be paid to femoral pulses and blood pressure gradient between right arm and leg.

Pre-Participation Physicals

2. Physical Examination:

General:

General examination is important to identify features that could be suspicious for certain syndromes:

- Abnormalities of spinal curvature, pectus deformities, hyperextensible joints, arm span to height ratio, myopia, and other characteristic facial features: Marfan syndrome
- Neck webbing, short stature, a low hairline, or low-set ears: Turner syndrome

Pre-Participation Physicals

2. Physical Examination:

Cardiovascular exam:

- There should be a focus on identifying pathologic heart murmurs, abnormal or extra heart sounds, or rhythm irregularity
- Benign murmurs are common in pediatrics, and it can be a challenge to differentiate between benign and pathologic

Pre-Participation Physicals

❖ **Electrocardiogram Screening:**

- There continues to be debate regarding whether or not ECGs should be added to the typical PPE
- The European Society for Cardiology recommends including ECG for competitive athletes and some countries mandate ECGs at various ages
- The most recent guidelines from the American College of Cardiology (ACC), AHA, and AMSSM suggest that ECG may be performed but did not recommend universal ECG screening

Pre-Participation Physicals

TABLE 1. **The American Heart Association
14-Element Cardiovascular Screening
Checklist for Congenital and Genetic
Heart Disease**

MEDICAL HISTORY^a

Personal history

1. Chest pain/discomfort/tightness/pressure related to exertion
2. Unexplained syncope/near-syncope^b
3. Excessive and unexplained dyspnea/fatigue or palpitations associated with exercise
4. Previous recognition of a heart murmur
5. Elevated systemic blood pressure
6. Previous restriction from participation in sports
7. Previous testing for the heart, ordered by a physician



Pre-Participation Physicals

TABLE 1. **The American Heart Association
14-Element Cardiovascular Screening
Checklist for Congenital and Genetic
Heart Disease**

Family history

8. Premature death (sudden and unexpected or otherwise) before 50 y of age attributable to heart disease in ≥ 1 relative

9. Disability from heart disease in a close relative <50 y of age

10. Hypertrophic or dilated cardiomyopathy, long-QT syndrome, or other ion channelopathies, Marfan syndrome, or clinically significant arrhythmias; specific knowledge of genetic cardiac conditions in family members



Pre-Participation Physicals

TABLE 1. **The American Heart Association
14-Element Cardiovascular Screening
Checklist for Congenital and Genetic
Heart Disease**

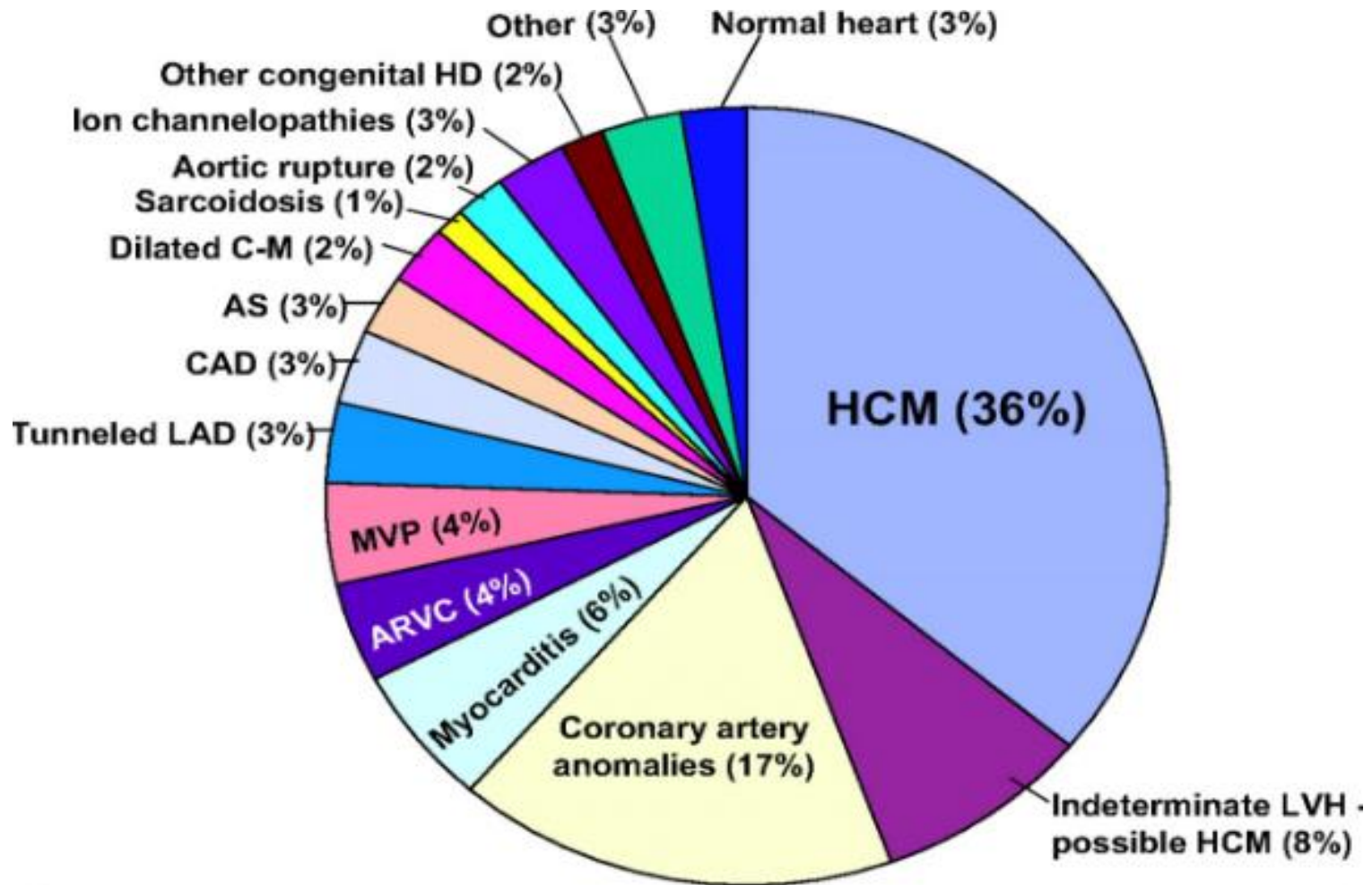
Physical examination
11. Heart murmur ^c
12. Femoral pulses to exclude aortic coarctation
13. Physical stigmata of Marfan syndrome
14. Brachial artery blood pressure (sitting position) ^d



OUTLINE

- ❖ Epidemiology
- ❖ Pre-participation Physicals
- ❖ Causes of Sudden Cardiac Death

Etiology based on largest US data set

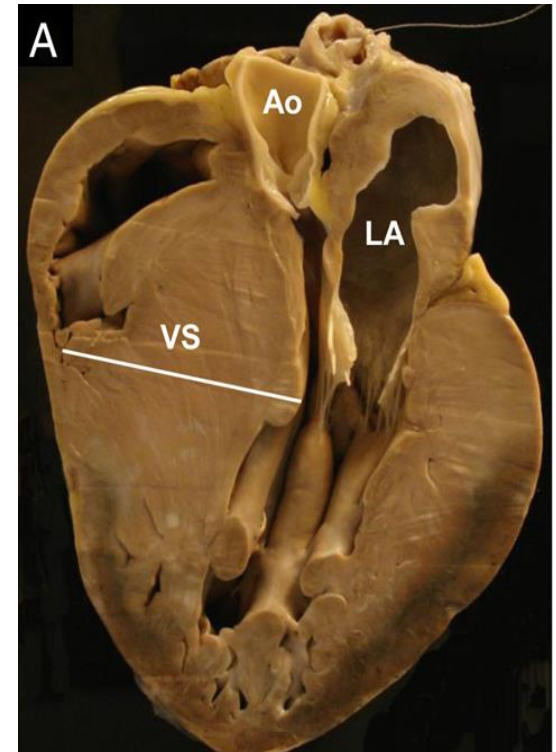


Causes of Sudden Death

- ❖ **Hypertrophic Cardiomyopathy**
- ❖ **Coronary artery anomalies**

Hypertrophic Cardiomyopathy

“A disease state characterized by unexplained LV hypertrophy associated with non-dilated ventricular chambers, in the absence of another cardiac or systemic disease that could produce this hypertrophy”



Hypertrophic Cardiomyopathy - Epidemiology

❖ Global disease

- reported in over 50 countries on all continents, all ethnic/racial groups, both genders

❖ Prevalence of 0.2% (1 in 500)

Scope of the Disease

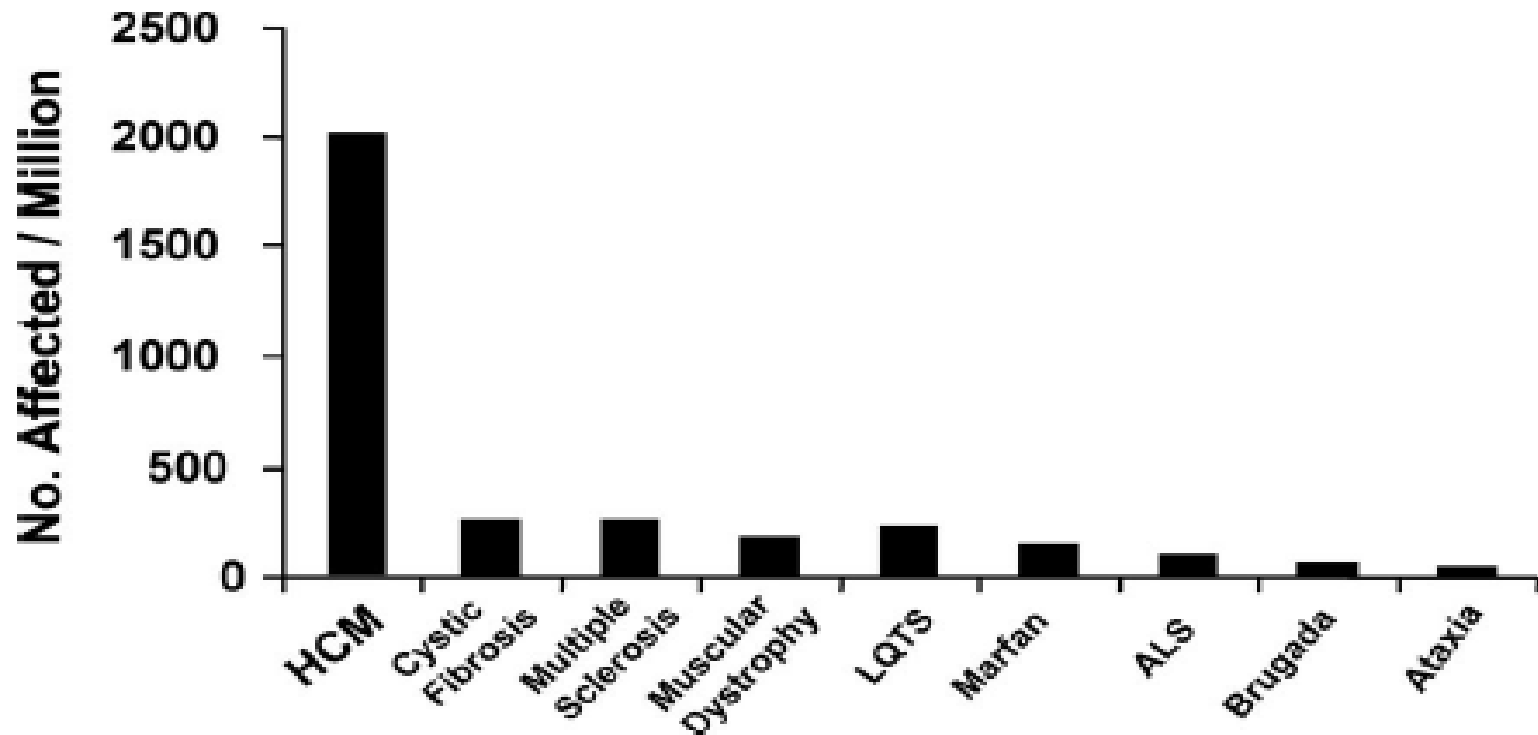


Figure 2. The prevalence of hypertrophic cardiomyopathy in the general population substantially exceeds that of several other cardiac and noncardiac diseases, which paradoxically have achieved greater recognition in the public sphere.

Genetics

- ❖ Transmission is autosomal dominant, 50% chance of inheriting
- ❖ De novo /sporadic mutations do arise
- ❖ Phenotypic heterogeneity is present within and between families
- ❖ Most will demonstrate LVH by early adulthood, accelerated during adolescent growth spurt

Genetics

TABLE 1. Distribution of Genes in HCM-Genotyped Index Cases According to Familial or Sporadic Cases

Gene	Total*	Familial HCM	Sporadic	Mutations (Novel)
Total	n=124	n=109	n=15	97 (60)
MYBPC3	52 (42%)	45 (41%)	7 (47%)	39 (25)
MYH7	50 (40%)	45 (41%)	5 (33%)	40 (24)
TNNT2	8 (6.5%)	5 (4.5%)	3 (20%)	7 (2)
TNNI3	8 (6.5%)	8 (7%)	0	7 (6)
MYL2	5 (4%)	5 (4.5%)	0	4 (2)
MYL3	1 (<1%)	1 (<1%)		1 (1)

*There were 120 initial index cases, but 2 different mutations within the same family were identified in 4 families. The distribution was therefore performed on 124 index cases.

Phenotype and Morphologic Features

❖ Left ventricular hypertrophy

- No single pattern
- Diffuse disease more common, but 1/3rd have localized hypertrophy
- Asymmetric- anterior septum predominant
- Other markers: hypercontractile LV, dynamic subaortic obstruction produced by systolic anterior motion of the mitral valve (SAM)

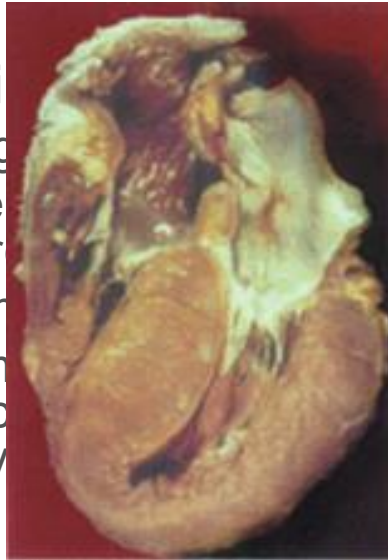
❖ Cellular components

- LV myocardial architecture is disorganized
- Hypertrophied cardiac muscle cells with bizarre shapes, intercellular connections in chaotic alignment
- Degree of cellular disarray variable- more extensive in patients who die young
- Abnormal intramural coronary arteries

Phenotype and Morphologic Features

❖ Left ventricle

- No single
- Diffuse hypertrophy
- Asymmetric
- Other morphologic obstruction of the mitral valve

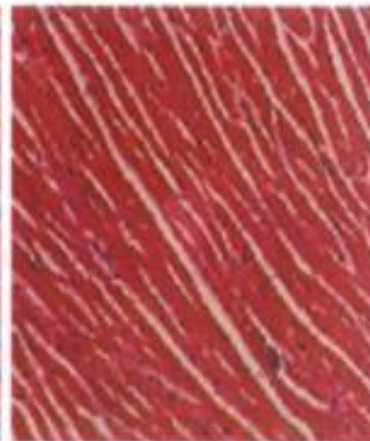
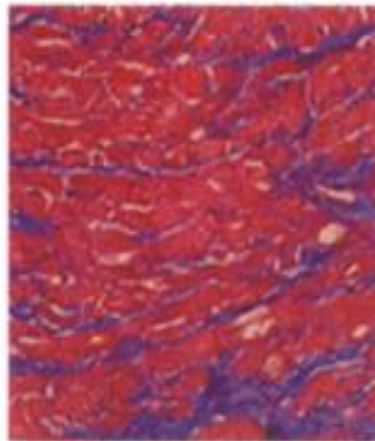


are localized

c subaortic stenosis of the

❖ Cellular composition

- LV myocardium
- Hypertrophy of intercalated discs
- Degree of fibrosis in patients
- Abnormal



irregular shapes,
t
extensive in

HCM

Normal



Pathophysiology

- ❖ LVOT obstruction
- ❖ Diastolic dysfunction
- ❖ Myocardial ischemia
- ❖ Autonomic dysfunction
- ❖ Mitral regurgitation

Pathophysiology

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Pathophysiology

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- ❖ Diastolic dysfunction
- ❖ **Myocardial ischemia**
- ❖ Autonomic dysfunction
- ❖ Mitral regurgitation

Pathophysiology

❖ Myocardial Ischemia

- Demand Supply mismatch
- Myocardial bridging

Demand

Hypertrophy
Afterload



Supply

Abnormal CAs

Pathophysiology

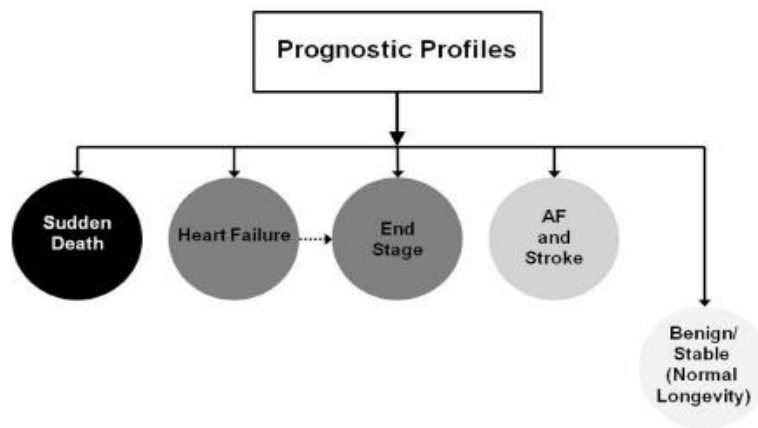
- ❖ LVOT obstruction
- ❖ Diastolic dysfunction
- ❖ Myocardial ischemia
- ❖ **Autonomic dysfunction**
- ❖ Mitral regurgitation

Pathophysiology

- ❖ LVOT obstruction
- ❖ Diastolic dysfunction
- ❖ Myocardial ischemia
- ❖ Autonomic dysfunction
- ❖ **Mitral regurgitation**

Clinical Course/Natural history

- ❖ Presented with murmur, family history, abnormal EKG and arrhythmia
- ❖ From asymptomatic to obstructive disease to aborted sudden cardiac death
- ❖ Mortality rate 1% per year, much lower than older studies



Asymptomatic/mildly Progressive HF
Sudden death
Atrial fibrillation and embolic stroke

Sudden Death

- ❖ Most devastating and unpredictable consequence
- ❖ Can be the presenting manifestation
- ❖ Most common cause of sudden death in young adults
- ❖ Most frequent cause of SCD in US competitive athletes
- ❖ Sudden death risk decreases with increasing age
- ❖ Mechanism: Primary VT and Vfib; substrate:
 - abnormal cellular architecture
 - myocardial fibrosis from silent ischemia

Diagnosis: EKG

- ❖ As screening for sports physical/murmur/family history
- ❖ 12 lead EKG is abnormal in %75-95 of the patients with HCM
- ❖ Falsely negative in 10%
- ❖ Evidence of WPW
- ❖ Holter: evidence of VT

TABLE 1. *Frequency of Common Electrocardiographic Abnormalities in Patients with Hypertrophic Cardiomyopathy*

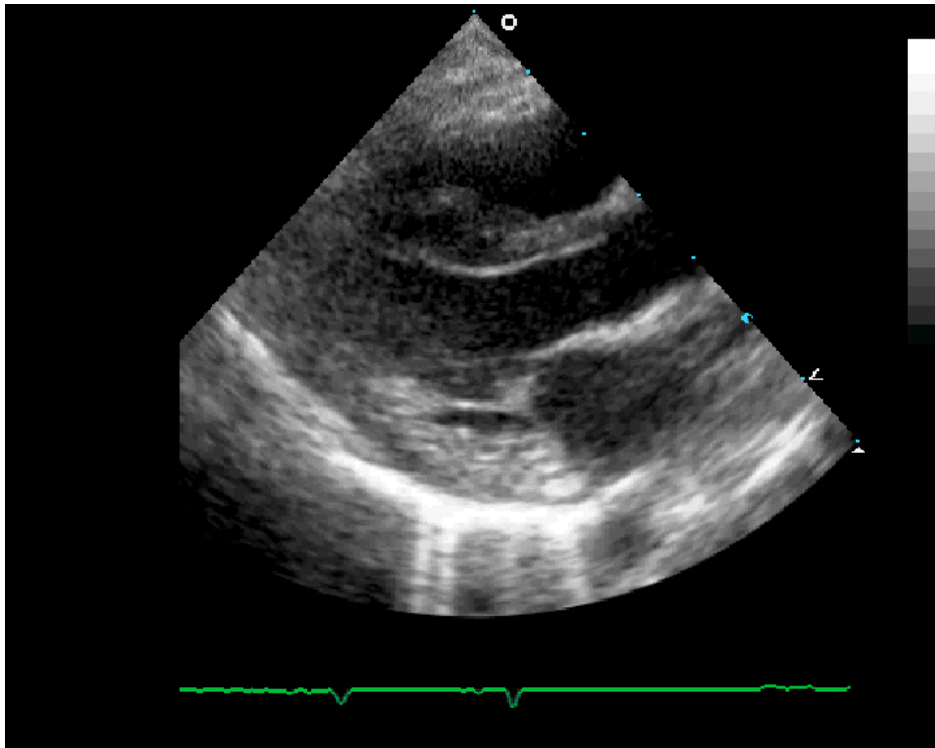
ECG Finding	Obstructed		Nonobstructed	
	Symptomatic	Asymptomatic	Symptomatic	Asymptomatic
Abnormal ECG	54/55 (98)	13/13 (100)	37/40 (93)	19/26 (73)*
Repolarization abnormalities	48/55 (87)	11/13 (85)	34/40 (85)	15/26 (58)†
Left ventricular hypertrophy	31/38 (82)	8/10 (80)	19/33 (58)	7/21 (33)‡
Left atrial abnormality	37/50 (74)	5/12 (42)	14/38 (37)*	5/26 (19)*
Left axis deviation (0 to -90°)	25/53 (47)	8/13 (62)	12/40 (30)	5/26 (19)‡
Abnormal Q waves	23/55 (42)	5/13 (38)	8/40 (20)	8/26 (31)
Right atrial enlargement	9/50 (18)	3/13 (23)	3/38 (8)	1/26 (4)
Right atrial enlargement and left ventricular hypertrophy	7/38 (18)	2/10 (20)	3/33 (9)	0/21 (0)
P-R interval >0.20 sec	5/50 (10)	2/13 (15)	4/38 (11)	1/26 (4)

Diagnosis- Imaging: TTE

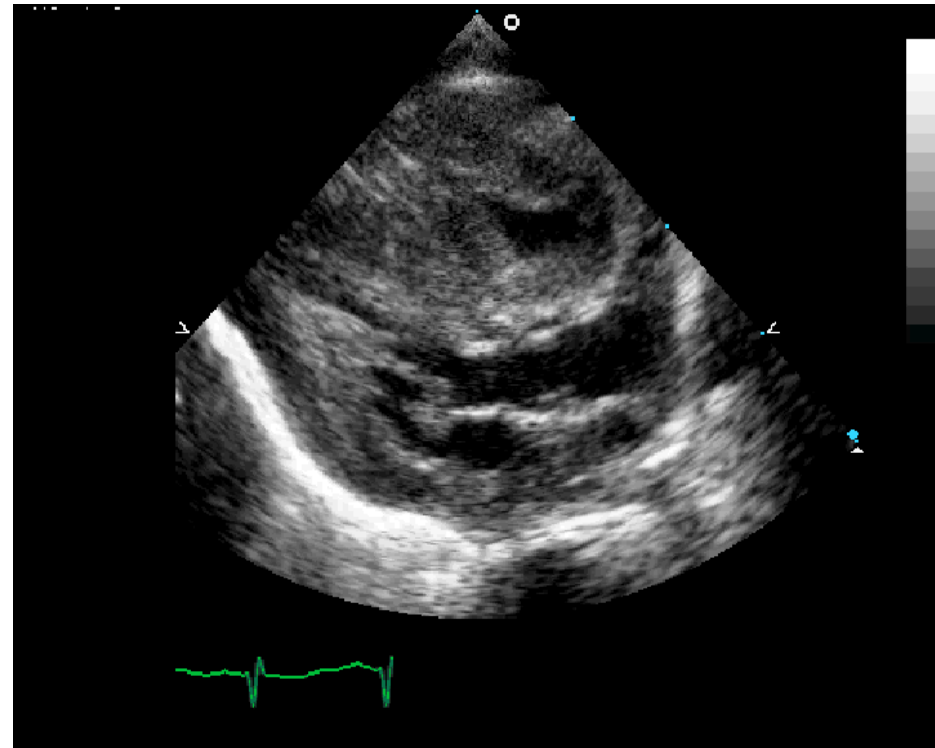
- ❖ Suspicion arises with a heart murmur or abnormal EKG
- ❖ Diagnosis confirmed with 2D echocardiography
- ❖ Echocardiogram:
 - LV/RV wall thickness
 - LV mass
 - Symmetric/asymmetric
 - Systolic anterior motion of the MV
 - Hyperdynamic LV
 - Subaortic obstruction and degree (CW Doppler)
 - LV function- systolic and diastolic

Echocardiogram

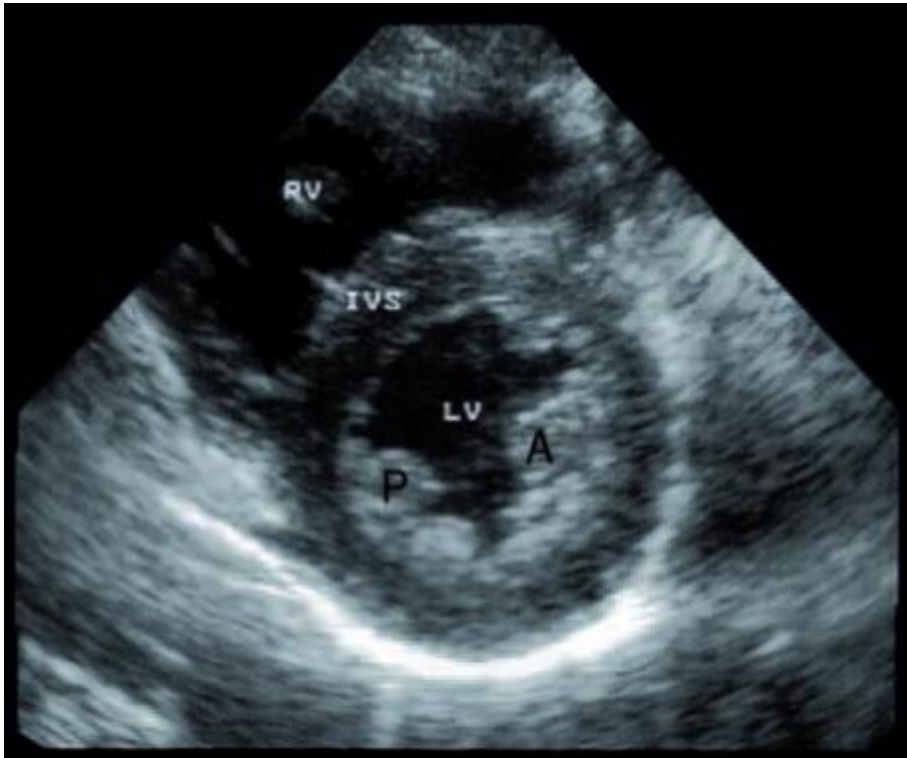
Normal



HCM



Echocardiogram



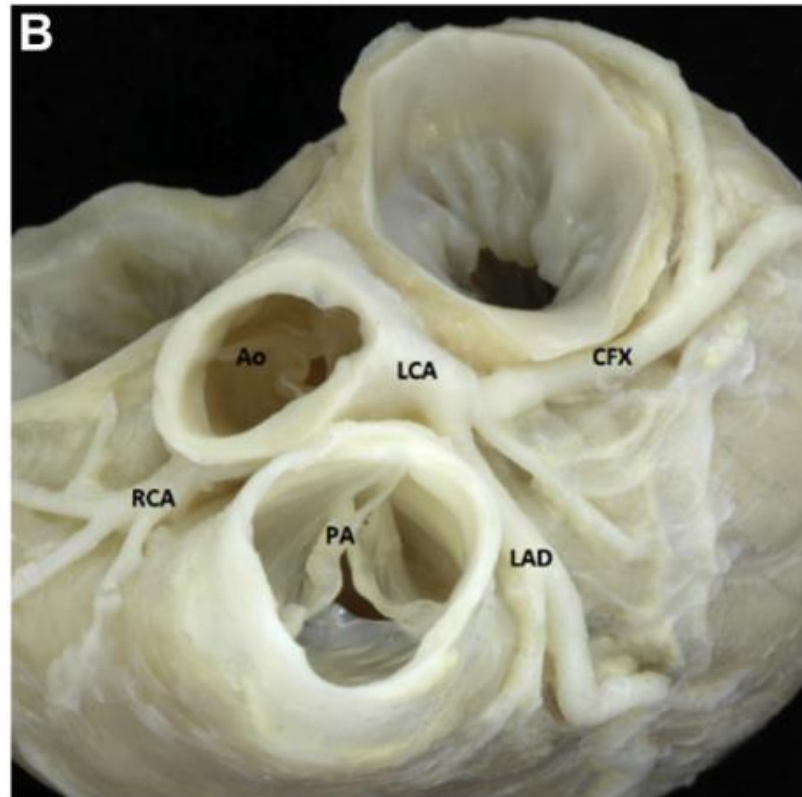
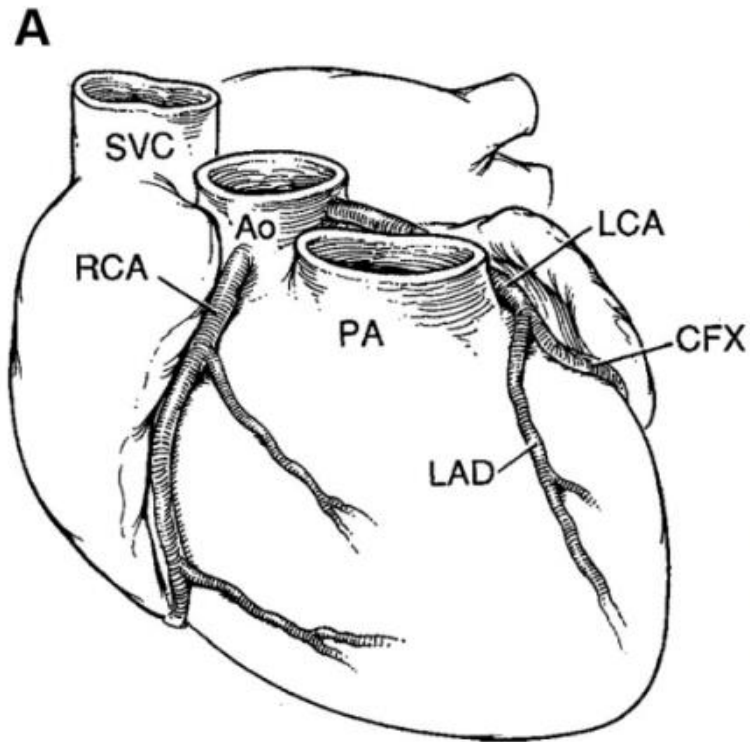
(a)

Causes of Sudden Death

❖ Hypertrophic Cardiomyopathy

❖ Coronary artery anomalies

Coronary artery abnormalities



Frommelt et al. Journal of the American Society of Echocardiography. March 2020



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Coronary artery abnormalities

Table 1. Simplified Nomenclature of CAAs

Type of anomaly	Variant	Subvariants
Anomalies of origin	Anomalous pulmonary origin of the coronaries	Origin of left main coronary artery from the pulmonary artery Origin of right coronary artery from the pulmonary artery Origin of circumflex coronary artery from the pulmonary artery Origin of left and right coronary arteries from the pulmonary artery
	Anomalous aortic origin of the coronaries	Origin of left main coronary artery from the right aortic sinus of Valsalva Origin of right coronary artery from the left aortic sinus of Valsalva Origin of left anterior descending coronary artery from the right aortic sinus of Valsalva Origin of left anterior descending coronary artery from the right coronary artery Origin of circumflex coronary artery from the right aortic sinus of Valsalva Origin of circumflex coronary artery from the right coronary artery Single coronary artery Inverted coronary arteries Others
	Congenital atresia of the left main artery	
Anomalies of course	Myocardial (or coronary) bridging	Symptomatic Asymptomatic
	Coronary aneurysm	Congenital Acquired
Anomalies of termination	Coronary arteriovenous fistula	Congenital Acquired
	Coronary stenosis	Congenital Acquired

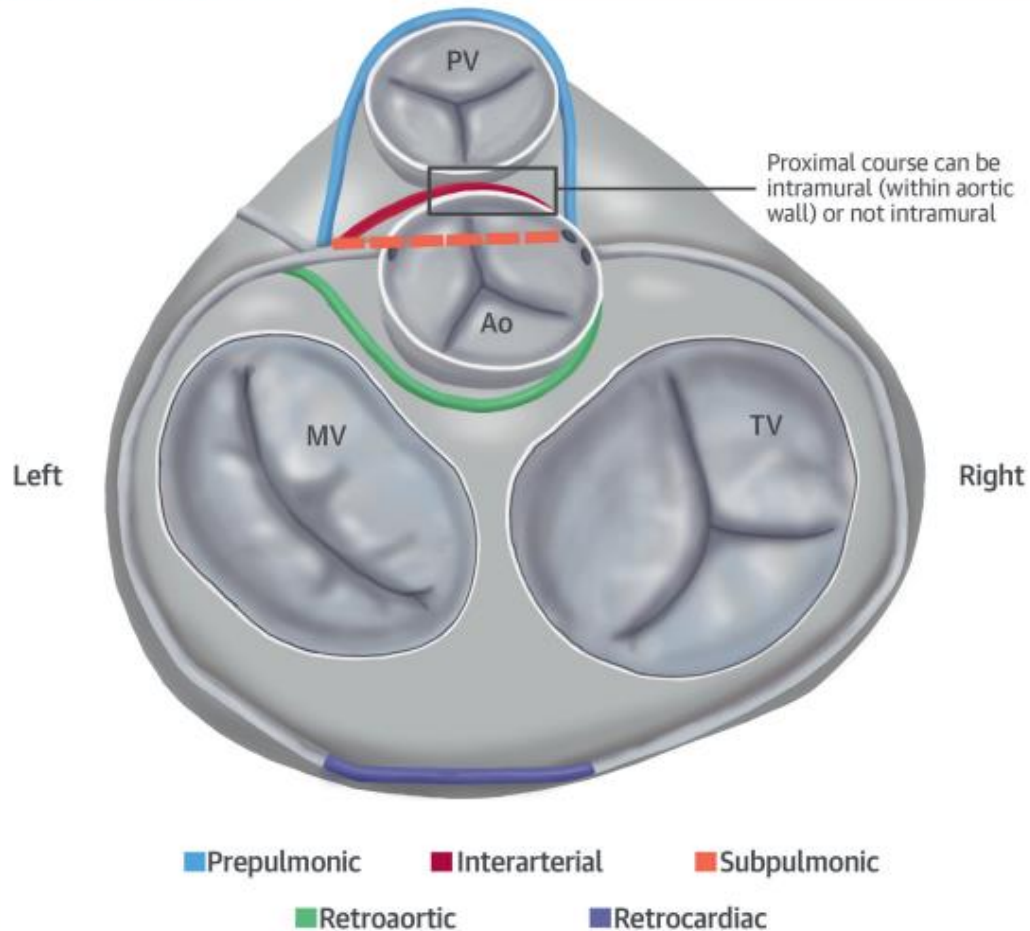
CAA indicates coronary artery anomaly.

AAOCA Variations

- ❖ Origin: at or above the inappropriate sinus of Valsalva
- ❖ Ostium Morphology: Slit-like, ovale, round
- ❖ Course:
 - Interarterial
 - pre-pulmonic (0.04%)
 - sub-pulmonic (0.07%)
 - Retrocardiac (0.03%)
 - Retroaortic (0.28%)

AAOCA Variations

CENTRAL ILLUSTRATION AAOCA: Course Subtypes



Cheezum, M.K. et al. *J Am Coll Cardiol.* 2017;69(12):1592-608.

The 5 main course subtypes of anomalous aortic origin of a coronary artery (AAOCA) arising from the inappropriate sinus are shown: **blue** – pre-pulmonic; **red** – interarterial; **orange** – subpulmonic; **green** – retroaortic; **purple** – retrocardiac. Figure prepared by Robert Cheezum and Chris Shearin (DesignVis Studios Inc., Indianapolis, Indiana), and adapted with permission from Angelini et al. (80). Ao – aorta; MV – mitral valve; PV – pulmonic valve; TV – tricuspid valve.

Cheezum et al. JACC 2017

Anomalous Aortic Origin of a Coronary Artery (AAOCA)

- ❖ Which coronary artery has the anomalous origin?
- ❖ What is its course?
- ❖ What are the patient's symptoms if any?
- ❖ What is the management?

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Anomalous Aortic Origin of a Coronary Artery (AAOCA)

❖ Estimated Prevalence:

- Interarterial AAOLCA: 0.03%
- Interarterial AAORCA: 0.23 %

AAOCA – Sudden Cardiac Death

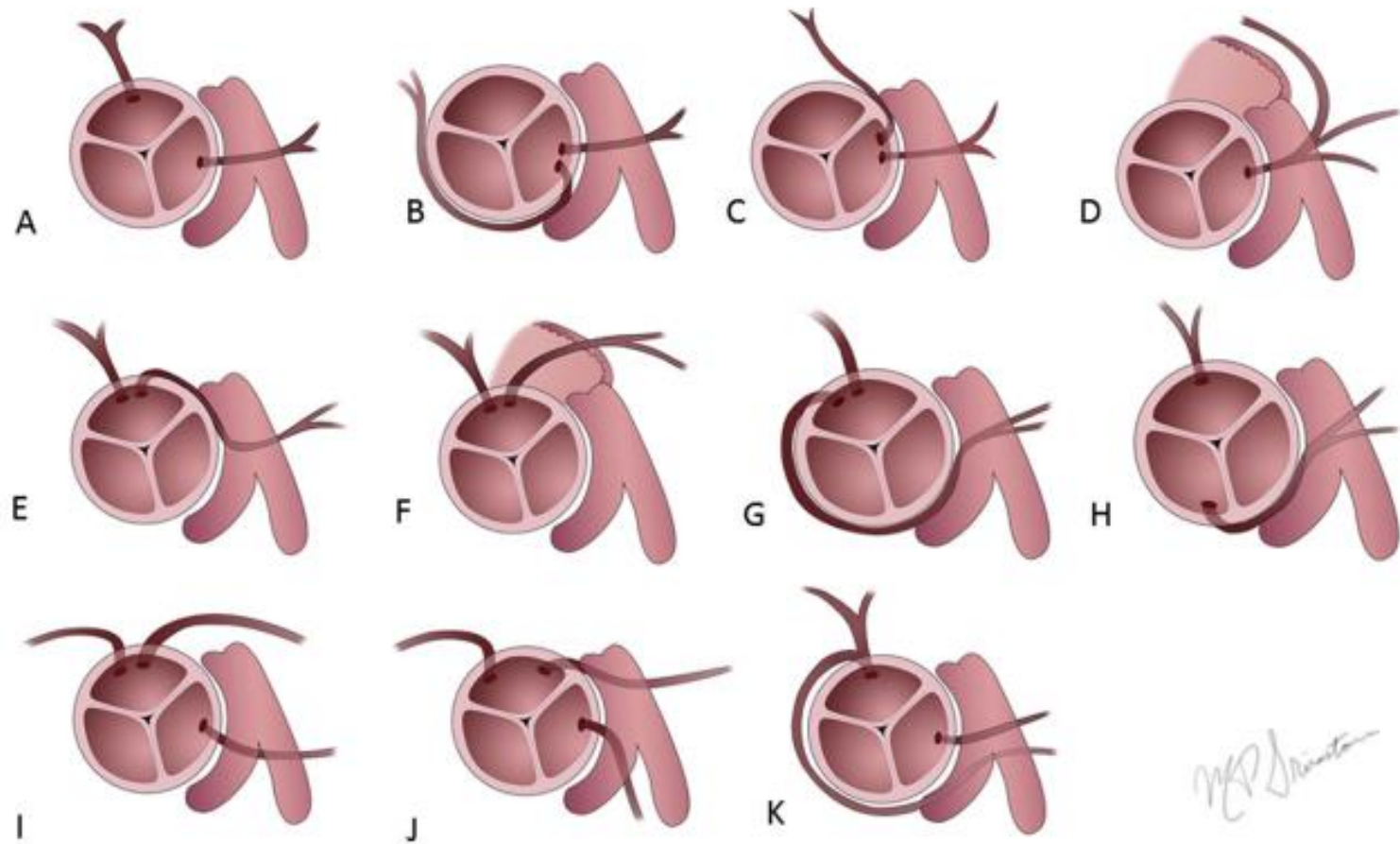
- ❖ Cumulative risk of death with AAOCA (15-35 years) participating in competitive sports:
 - 6.3% AAOLCA
 - 0.2% AAORCA

Case reports of SCD with AAOCA at rest/recreational activities

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AAOCA Variations



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Anomalous Aortic Origin of a Coronary Artery (AAOCA)

Clinical presentation:

- Asymptomatic
- Exertional chest pain
- Exertional syncope
- Sudden death

Pathophysiology of SCD with AAOCA

Possible mechanisms of sudden death

- Compression of intramural segment
- Compression of the interarterial segment
- Acute angle at take-off
- Ostial stenosis
- Coronary spasm

Anomalous Aortic Origin of a Coronary Artery (AAOCA)

Imaging modalities: Echo

❖ Pros:

- available
- affordable
- Portable
- Noninvasive nature



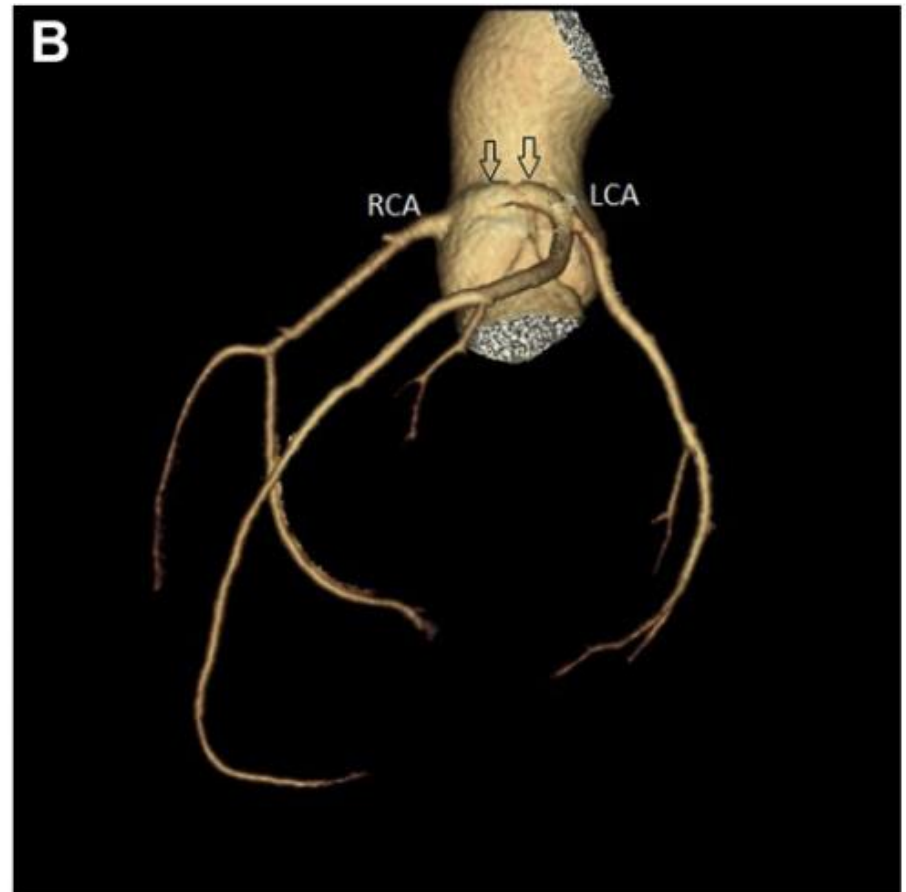
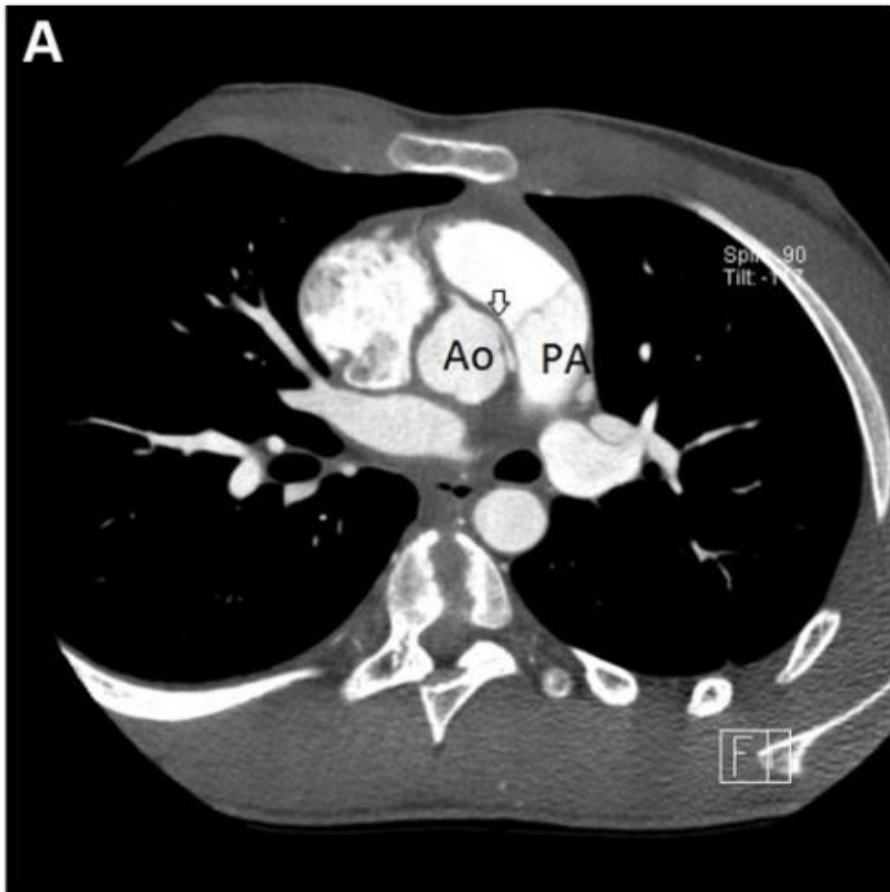
Cons:

- Spatial resolution
- Body habitus and sonographer experience

Anomalous Aortic Origin of a Coronary Artery (AAOCA)

Imaging modalities: Coronary CTA

AAOLCA with an interarterial, intramural course



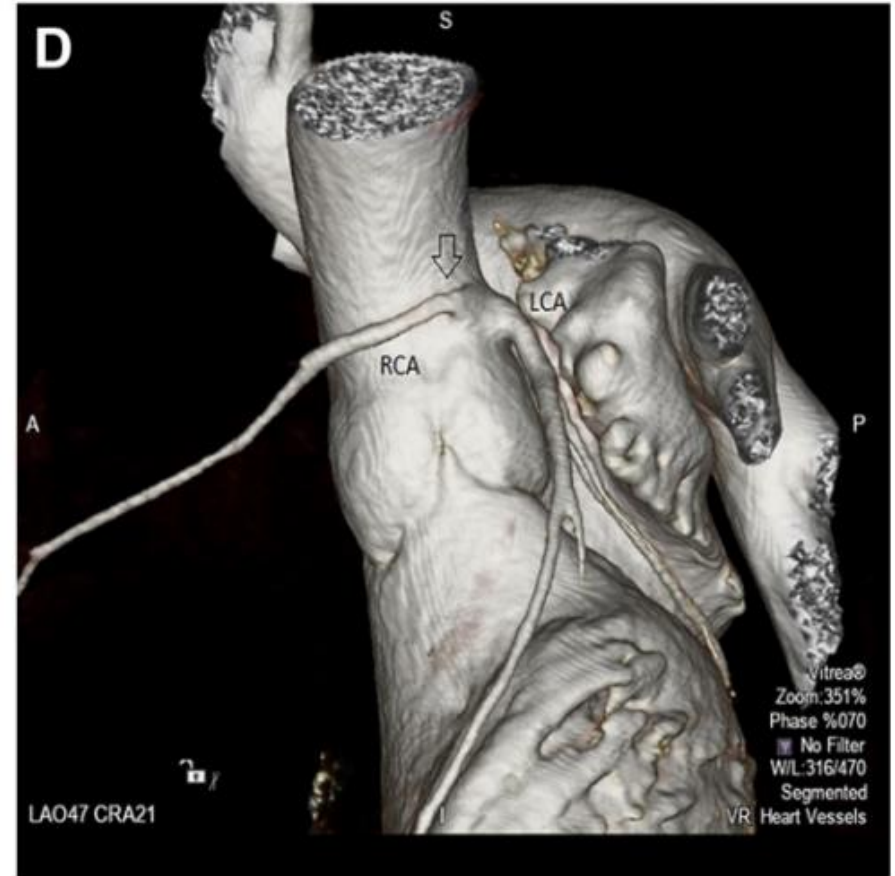
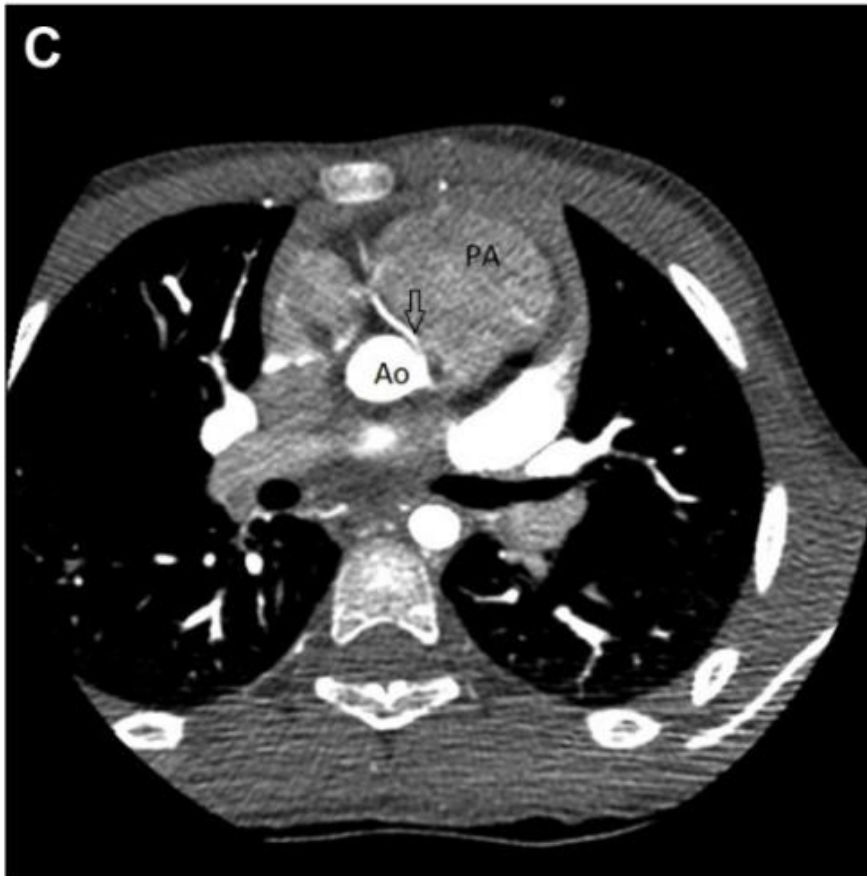
Frommelt et al. Journal of the American Society of Echocardiography. March 2020



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Anomalous Aortic Origin of a Coronary Artery (AAOCA) Imaging modalities: Coronary CTA

AAORCA with an interarterial, intramural course



Anomalous Aortic Origin of a Coronary Artery (AAOCA)

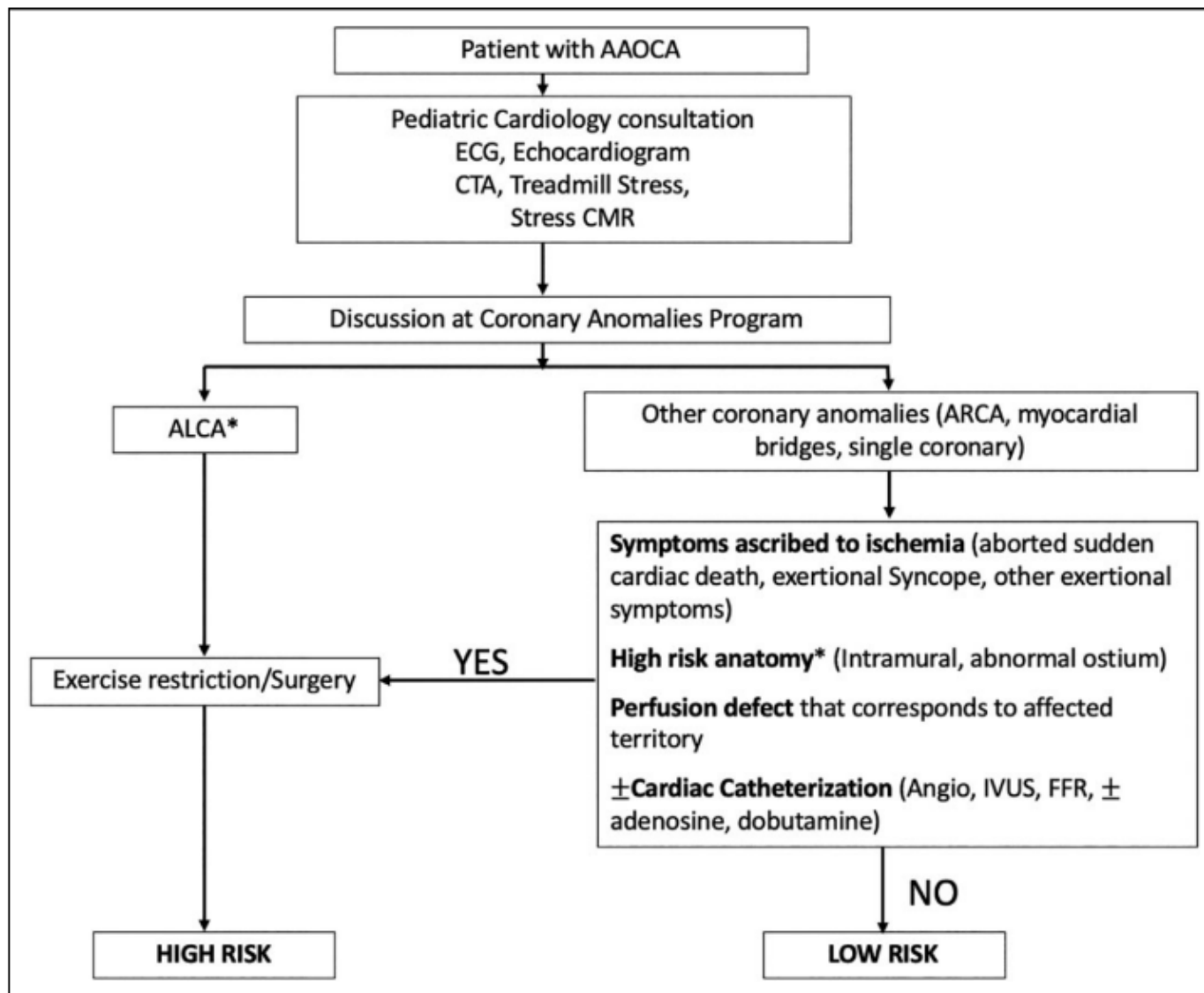
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Anomalous Aortic Origin of a Coronary Artery (AAOCA)

❖ Treatment:

- The indications for intervention in asymptomatic patients with AAOCA with an intramural course are debated, especially with AAOCA RCA from the left sinus of Valsalva
- **Surgical intervention is recommended:**
 - Signs or symptoms of myocardial ischemia (eg, true angina, findings on provocative testing, aborted sudden cardiac death or arrest, or non-vagally-mediated arrhythmia)
 - Anomalous origin of the left coronary artery from the right sinus with an interarterial/intramural course, even in the absence of symptoms (due to the higher calculated risk of sudden cardiac death)
 - Asymptomatic patients with an intramural RCA from the left sinus of Valsalva (AAORCA), provocative testing is recommended

Anomalous Aortic Origin of a Coronary Artery (AAOCA)



Pediatric Sudden Cardiac Death

THANK YOU!