Sudden Cardiac Arrest in Children and Adolescents

Other causes of SCA
- Coronary artery abnormalities
  - Myocarditis
  - Commotio Cordis

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Coronary Artery Abnormalities
Coronary artery abnormalities

- Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA)
- Origin of the coronary artery from the opposite sinus of Valsalva
- Coronary artery fistulae
- Kawasaki disease
- Atherosclerotic coronary artery disease

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Coronary artery from the opposite sinus of Valsalva

- Which coronary artery has the anomalous origin?
  - Right or left
- What is it’s course?
  - Intramural, intrarterial
  - Intramyocardial
- What are the patient’s symptoms if any?
- What to do and when?
Coronary artery anomalies

• Byron H
  • “Healthy” 14 year old
  • March 2001, sudden cardiac arrest while playing basketball at community center
  • CPR performed, defibrillation 15 minutes later (paramedics)
  • Transfer to CHW, severe DCM

Coronary artery anomalies

• Byron’s severe LV dysfunction:
  • Primary?
    • History suggests that he was asymptomatic
  • Secondary to the prolonged arrest and fibrillation?
    • Most likely scenario
Coronary artery anomalies

- Byron H.
  - Balloon pump then mechanical cardiac support with ECMO for 72 hours
  - Past history: This was the 4th episode of syncope in the last 2 years
  - Cardiac work-up in Racine 1 year previous with echocardiogram, tilt, EP – all “normal”

Coronary artery anomalies

- Echocardiogram
  - Transthoracic
  - TEE
Coronary artery anomalies

Anomalous origin of LCA from right sinus of Valsalva
Coronary artery anomalies

• Byron H.
  • Weaned from ECMO after 72 hours with full recovery of LV function
  • Taken to OR for ECMO decannulation and “unroofing” of LCA into left sinus of Valsalva
  • Full neurologic recovery, discharged 3 weeks after admission
AOCA from the Opposite Sinus with an Interarterial Course—Anatomic Features

A
LCA from right sinus

B
RCA from left sinus

AOCA: Sudden Death Risk

- LCA from right sinus of Valsalva is rare (0.03%) but clearly associated with sudden cardiac death
  - 29/38\(^1\) and 36/49\(^2\) cardiac death
  - 23/23 <20 yrs died suddenly with exercise\(^1\)
  - Majority of exercise-induced SD were asymptomatic \(^1,3\)

- RCA from the left sinus of Valsalva is more common (0.1%) but also associated with sudden cardiac death
  - 15/52 cardiac death; 13 sudden cardiac death\(^2\)
  - All 13 SD were asymptomatic
  - 8/25 sudden cardiac death; 6/8 were 16-33 yrs\(^1\)

From: \(^1\)Roberts WC, Am Heart J 1986; 111
\(^2\)Taylor AJ et al, J Am Coll Cardiol 1992; 20
\(^3\)Frescura C et al, Hum Pathol 1998; 29
AOCA: Which is Riskiest?

- Largest retrospective review of SCD in military recruits from 1977-2001
  - 64 sudden deaths classified as cardiac among 6.3 million recruits
  - 39/64 died with coronary pathology at autopsy
  - 54% had AOCA (left from right sinus)
  - 26% had obstructive atherosclerotic CAD
  - 0% had AOCA (right from left sinus)
  - 50% had symptoms

Anatomy of CCAA Associated with SCD

- Certain anatomic subtypes of anomalous coronary origin carry higher risk of acute myocardial complications
  - Anomalous origin of a coronary artery (AOCA) from the opposite sinus with an interarterial course
    - Interarterial:
      - coursing between the great arteries
    - Intramural:
      - coursing within the wall of the aorta
    - Intramyocardial:
      - coursing through the myocardium
Anomalous origin of the LMCA from the right sinus can have a variable course

A) Anterior to PA
B) Between Ao and PA
   - interarterial
C) In ventricular septum
   - intramyocardial
D) Posterior to Ao

From Roberts and Shirani, Am J Cardiol 1992; 70

AOCA with an intramural course: runs within the wall of the aorta

Features
- separate orifice
- acute angle take-off
- may have ostial stenosis
  - exits aortic wall from appropriate sinus then courses normally
- surgical unroofing technique ideal

LCA from right sinus
AOCA from the Opposite Sinus with an Intramural Course - Anatomic Features

The LMCA arises from the right aortic sinus passing between the great arteries through a long intramural segment before exiting the aortic wall from the left sinus.

The RCA arises from the left aortic sinus and passes between the great arteries through a shorter intramural segment before exiting the aortic wall from the right sinus.

AOCA with an intramyocardial course: runs within the crista supraventricularis muscle before reaching the AV groove

Other features:
- single Y-shaped orifice, normal angle take-off
- ostial stenosis uncommon
- runs through muscle distant from sinus
- surgical options poor
Pathophysiology of SCD with AOCA

- Possible mechanisms of sudden death
  - Compression of intramural segment
  - Compression of the interarterial segment
  - Acute angle at take-off
  - Ostial stenosis
  - Coronary spasm
  - Previous ischemic injury

The wall tension maintaining vessel geometry is directly related to pressure times radius.

Pressure within the aorta and normally arising perpendicular coronary arteries are similar.

The smaller diameter coronary artery is at risk for compression when intramural distortion ischemia.
Imaging modalities: echo

- Likely to be the first modality used in the evaluation of coronary artery abnormalities
  - available
  - affordable
  - portable
  - interpretable
  - risk-free
  - Diagnostic
  - TEE if TTE equivocal

CCAA—Additional Imaging Techniques

Magnetic Resonance Imaging

Courtesy of Mark Fogel, MD CHOP
Indications for coronary MRI/CT

- When echo is incapable of providing the required diagnostic information
- When there are inconsistent findings
- As an alternative to cardiac cath
  - need a “go-to” technique when echo fails
- To obtain information for which MRI/CT may offer unique advantages
  - assessment of myocardial perfusion
AOCA—Who is at risk?

- Problems with prediction of SCD risk
  - Typically normal ECG, exercise testing
  - Sudden death may be first manifestation of disease
  - Myocardial ischemia likely occurs in infrequent bursts; autopsy studies may reveal acute injury, chronic changes
  - How do we reliably predict a sporadic event?
  - How can we risk stratify each patient?

AOCA—Who is at risk?

- Reviewed sudden death cases in athletes from two large registries (Minneapolis Heart Institute; University of Padova, Italy); only patients with AOCA/ no other cause of death/ age < 35 years.

- 27 patients (22 male)
  - Age range 9-32 years (mean 16 ± 5 yrs)
  - LMCA from R sinus (n=23); RCA from L sinus (n=4); all interarterial

- Clinical profile
  - Each athlete died during (n=25) or after (n=2) intense exertion
  - Clustered between 3pm and 9pm
  - 10 had prior symptoms (syncope, chest pain, dizziness with exertion)
  - ECG’s: available in only 10 pts (all normal)
  - Stress test: available in 6 pts (all normal)
  - ECHO: 2 pts (reported as normal)

From: Basso et al, JACC 2000, 35.
It is not clear that this data helps us risk stratify!!!!

AOCA—Methods of Intervention

- Multiple surgical techniques utilized
  - Coronary reimplantation
    - May be difficult when course is intramyocardial
    - May develop neo-ostial stenosis
  - Bypass graft placement
    - Subjects a young pt to a bypass graft/need for reintervention
    - Theoretical risk of decreased patency of graft related to competitive flow
  - Patch enlargement of the stenotic origin
    - Does not address intramural segment
    - May develop neo-ostial stenosis
    - Unroofing of the intramural segment
Methods of Intervention—Unroofing Technique

- Unroofing is technique of choice for intramural AOCA at CHW
  - Relieves ostial stenosis
  - Eliminates the intramural segment
  - Creates large neo-orifice in the appropriate sinus perpendicular to the aortic root

Conclusions - The Symptomatic Patient

- With either left from the right or right from the left presenting symptoms might include
  - Dizziness
  - Chest pain (with or without exercise)
  - Syncope
  - SCA
- Is it clear that the symptoms are a result of the coronary artery anomaly?
- Would you take a chance that they are not?
Conclusions - The Asymptomatic Patient

- The anomaly might be serendipitously discovered (bicuspid aortic valve, functional heart murmur, etc.)
- Then what – Is intervention required?
- If so, when?
- Does it matter which coronary artery is anomalous (right or left)?
- Does the specific course of the coronary artery matter (intramural and intrarterial vs. intramyocardial)?

Summary

- Although rare, AOCA is clearly associated with sudden cardiac death, particularly in young athletes
- Origin of the LMCA from the right sinus with an intramural course appears to be the highest risk subgroup
- In the asymptomatic child with an anomalous RCA from the left sinus, surgical therapy remains controversial
- A national or international registry of these patients is critical to improve risk stratification
Isolated myocardial fibrosis as a cause of sudden cardiac death and its possible relation to myocarditis.

Lecomte D, Fornes P, Fouret P, Nicolas G. Department of Forensic Medicine, Institut Médico-Légal de Paris, France.

In performing medicolegal autopsies on sudden deaths, there occur a number of cases in which no cause of death can be found. In particular, no evidence of macroscopic cardiac abnormalities can be observed. However, extensive histological screening may reveal isolated areas of myocardial fibrosis. The five cases presented discuss the etiology of this fibrosis and its possible relation to myocarditis. The cases involve white women between the ages of 19 and 25 with no previous medical history. The weight of the heart in all five cases was normal. Macroscopic evidence of fibrosis was visible in four out of five cases. No other macroscopic abnormalities were observed. Histologically, there was evidence of scarring or interstitial fibrosis in all five cases. In four of the cases, additional screening permitted the observation of dispersed inflammatory foci consisting of lymphocytes, plasmocytes and macrophages. Two of the cases demonstrated eosinophil and neutrophil aggregates in the center of necrotic foci. No evidence of vascular inflammatory phenomena was observed in any of the five cases. According to the Dallas criteria, three of the five cases fulfill the requirements for myocarditis and one of the five cases for borderline myocarditis. The Dallas criteria, however, do not take into consideration the possible association between inflammation and myocardial fibrosis since many of the reported series of myocarditis have been from hospital autopsies or endomyocardial biopsies and have not taken into account sudden death from fibrotic sequelae of myocarditis.

Incidence of Fatal Myocarditis: A Population-based Study in Finland

Ville Kytö, Antti Saraste, Liisa-Maria Voipio-Pulkki and Pekka Saukko

pp 570-74.

Abstract

To study the incidence of fatal myocarditis in the general population, the authors retrospectively collected all death certificates recording myocarditis as the underlying cause of death in Finland in 1970–1998. The incidence of myocarditis and its proportion of all deaths were calculated from 141.4 million person-years and 1.35 million deaths. Myocarditis was recorded as the underlying cause of death in 639 cases. Thus, its death certificate-based incidence was 0.46 (95% confidence interval (CI): 0.43, 0.49) per 100,000 person-years, and it caused 0.47 (95% CI: 0.44, 0.51) of 1,000 deaths. The incidence of 0.51 (95% CI: 0.46, 0.56) in males was higher than the incidence of 0.42 (95% CI: 0.37, 0.47) in females, the odds ratio being 1.34 (95% CI: 1.15, 1.58) (p < 0.001). The proportion of deaths caused by myocarditis was highest (up to six of 1,000 deaths) in children and adults aged less than 45 years. Because previous histopathologic reanalysis showed that only 32% of cases fulfilled the Dallas criteria, the authors estimated the incidence of histopathologically certain fatal myocarditis to be 0.15 (95% CI: 0.13, 0.17) per 100,000. The death certificate-based incidence of fatal myocarditis was found to be 0.46 per 100,000, and the histopathologically corrected incidence was 0.15 per 100,000.
We appreciated the outstanding review by Drs Zipes and Wellens on sudden cardiac death. However, the authors did not mention myocarditis as a cause of sudden death. In our experience at the Forensic Institute of Paris, myocarditis accounts for 5% of sudden, out-of-hospital cardiac deaths. Two patterns are common. Acute myocarditis, the first type, occurs most frequently in children or young adults. The diagnosis is likely to be overlooked because of the frequent lack of symptoms. When present, symptoms are not specific; they suggest a bad cold in most instances. The heart is grossly normal. In particular, it is not enlarged. Myocyte necrosis and inflammatory infiltrates, predominantly composed of lymphocytes, are present, but they are most frequently patchy rather than diffuse (American Academy of Forensic Sciences, Orlando, Fla, personal communication, 1999). Therefore, multiple myocardial samples are required for the diagnosis.
The second pattern is chronic myocarditis, which also involves young adults. The heart is again grossly normal. Histological examination of both ventricles shows patchy areas of fibrosis. In some sections, small clusters of lymphocytes may be present. These victims have no history of drug abuse, and toxicological analyses are negative. Viral infection is likely to be involved in most cases of both acute and chronic myocarditis. Idiopathic giant cell myocarditis, granulomatous myocarditis, and eosinophilic myocarditis are exceptional in our experience.

Fibrosis is known to cause slow conduction in cardiomyocytes, resulting in arrhythmogenic substrate reentry circuits and subsequent ventricular desynchronization.

Even more important is the possible role of acute inflammation of the myocardium, which could, by itself, promote ventricular arrhythmias or act as a trigger if this inflammation occurs in patients with heart disease who are prone to arrhythmia, such as those with concealed forms of cardiomyopathies. Hoffman et al studied reperfusion arrhythmia and demonstrated, for the first time, the role played by the activation of neutrophils in producing early after-depolarization. These results demonstrate a direct link between acute inflammation and an electrophysiological phenomenon that is a possible trigger of arrhythmias. From a personal correspondence with Dr Brian Hoffman (October 1997), we learned that this mechanism is also likely to be involved in patients with advanced congestive heart failure in which tumor necrosis factor- is present and that it is also an important cause of early after-depolarizations.
Response

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Hein J.J. Wellens, MD
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Fontaine et al correctly called our attention to the role of myocarditis as a cause of sudden death. Indeed, in our article, we did not comment on inflammatory and infiltrative causes of sudden arrhythmic death for 2 reasons: restrictions regarding the size of the article and difficulty in obtaining reliable information on the incidence of these abnormalities as causes of sudden death.

Fontaine et al refer to their findings at the Forensic Institute of Paris, where myocarditis was found in 5% of sudden, out-of-hospital cardiac deaths. However, we would like to know the true incidence of myocarditis in an unselected population of sudden death victims. Myocarditis continues to be a very difficult diagnosis to make when the patient is still alive, both in the acute and the chronic state. We hope that new molecular biological and genetic techniques will provide better insight into the incidence of myocarditis and its contribution to sudden cardiac death. It will be quite difficult to unravel the arrhythmogenic mechanism(s) in this setting.
Amber

- 11 y.o. healthy girl running around the track at gym and collapsed
- Pulseless and non-breathing
- CPR begun immediately
- School (Project ADAM) AED delivered a single shock
- Woke up, transfer to CHW
Work-up

- History negative
- Family history negative
- Physical exam normal
- ECG normal
- Echo normal anatomy AND function
Amber

- Presumptive diagnosis = myocarditis
  - Delayed enhancement on MRI
  - Review history – Flu-like illness 2 weeks ago
  - ESR 25
- Now what?
  - Is this a self-limited illness?
  - When does the risk go away?
  - ICD vs. vest vs. AED
What would you do?

Amber

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  • Delayed enhancement on MRI
  • Review history – Flu-like illness 2 weeks ago
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• Now what?
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  • ICD vs. vest vs. AED
So what does this all mean?
Commotio Cordis

19th century Latin
“agitation of the heart”
Pediatric Pre-hospital SCA

- Sean Morley, 13 years old
- Struck in chest with baseball, collapsed
- Unresponsive, eyes “glassy,” agonal gasps
Sean M, 13 year old, collapsed

- What is his most likely problem?
  *Commotio cordis with VF*

- What treatment does he require?
  *Immediate bystander CPR*
  *Defibrillation within 3-5 minutes*

Sean M, 13 year old, collapsed

- Immediate bystander CPR provided
- A mother flagged down passing police car that carried a trained officer with an AED
A. AED attached, analyzing
B. VF then PEA
C. Shock advised, delivered
C. PEA $\rightarrow$ CPR (1 min)
E. Perfusing rhythm returns

**Successful Resuscitation**

- Emergency recognition
- Immediate bystander CPR
- Defibrillation within minutes
- 1 shock + CPR $\rightarrow$ ROSC and survival to hospital discharge

*Source: NEJM, 10/2002*
Commotio cordis

Maron BJ, Estes NA 3rd. Hypertrophic Cardiomyopathy Center Minneapolis Heart Institute Foundation, Minneapolis, MN 55407.


Commotio Cordis

- Ventricular fibrillation and sudden death
- Triggered by blunt, non-penetrating unintentional blow to the chest
- Without damage to the heart, sternum, ribs
- In the absence of underlying cardiovascular disease
Commotio Cordis

- First described in the 19th century
- Different from cardiac contusion because of the absence of structural cardiac injury
- Through the 1990s described in occasional case reports
- Increasing awareness and interest
- Precise incidence is unknown
- National commotio cordis registry (Maron)
Outcome

- Usually but not always fatal
- Maron registry 25% survival
- Outcome related to speed at which intervention/resuscitation occurs; recognition
- Survival rates have increased over time; better recognition, chain of survival, AEDs available
- Documented cases of the events aborting spontaneously
Mechanisms

- A primary arrhythmic event
- Mechanical blow confined to a small area of the precordium
- Profoundly alters the electrical stability of the myocardium, results in VF
- Excessive vagal reflex, coronary vasospasm, R on T?
Mechanisms

• Mark Link experimental model; projectile blows delivered at a wide range of velocities to anesthetized pigs
• Two critically important mechanical determinants
  • Location of the blow must occur directly over the heart
  • Timing of the blow: must occur within 10-20 msec. on the upstroke of the T wave

Mechanisms

• Variable factors in Link’s experimental model
  • Speed of the impact
  • Projectile sizes, shapes and weights
  • Hardness of the object
  • Adult vs. children (mature and fully developed rib cage)
  • Propensity in some patients based upon underlying QT interval – no evidence for this
  • Cellular mechanisms – Similar to ion channelopathies?
Prevention

- **Primary Prevention**
  - Avoid precordial blows – Don’t use chest to block pucks, balls (lacrosse), etc.
  - Improved commercial sports equipment
    - Softer balls
      - May change the game
      - Don’t always work
  - Chest protection
    - Don’t always work
Prevention

• Secondary Prevention
  • Wide dissemination of AEDs at sporting events
  • Teach CPR
  • Recognize commotio cordis
  • Early institution of the chain of survival

Summary Commotio Cordis

• An important cause of SCD
• Occurs in healthy and active young people
• A random yet very specific event: chest blow in right place at right time in right person, etc.
• Education, better-designed athletic equipment and early recognition can result in prevention of death
Thank you very much