Sudden Cardiac Death in Young Athletes
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Two Rivers, WI, (population = 12,000)
Sudden Cardiac Death in Young Athletes

Why Do We Talk About It?
Wes Leonard, 16 yo, Fennville High School
Died Minutes After Game winning shot
3/4/11

• Diagnosis:
  – Dilated Cardiomyopathy

- Diagnosis: Anomalous Left Coronary Artey
Sudden Cardiac Death in Young Athletes

- **Incidence:** 1/30,000 to 1/300,000
  - (vs. Older population > 40 is 1/15,000)

- **Mean age** = 17 – 23 years old

- **Gender** = Males:Females 9:1
Sudden Cardiac Death in Young Athletes

Predictors:

1) Family History of SCD or Heart Disease prior to age 50

2) Symptoms of Dizziness, Syncope, Exertional Chest Pain
Sudden Cardiac Death in Young Athletes

- SCD is related to *Intensity* of Exercise
- 90% of SCD occurs During or Shortly After Exercise
- Basketball and Soccer are most common sports, then Football and Track
  - Swanson W.; Seattle Children’s Hospital Research Foundation, 3/23/12
Sudden Cardiac Death in Young Athletes

Causes:

Hypertrophic Cardiomyopathy (HCM)
Coronary Artery Anomalies
Long QT Syndrome
Arrhythmogenic Right Ventricular Hypertrophy
Brugada Syndrome
Marfan Syndrome
Commotio Cordis
Others...

All Causes Lead to Ventricular Fibrillation!
The 12-Element AHA Recommendations for Preparticipation Cardiovascular Screening of Competitive Athletes

Medical history*

**Personal history**
1. Exertional chest pain/discomfort
2. Unexplained syncope/near-syncope†
3. Excessive exertional and unexplained dyspnea/fatigue, associated with exercise
4. Prior recognition of a heart murmur
5. Elevated systemic blood pressure

**Family history**
6. Premature death (sudden and unexpected, or otherwise) before age 50 years due to heart disease, in 1 relative
7. Disability from heart disease in a close relative 50 years of age
8. Specific knowledge of certain cardiac conditions in family members: hypertrophic or dilated cardiomyopathy, long-QT syndrome or other ion channelopathies, Marfan syndrome, or clinically important arrhythmias
The 12-Element AHA Recommendations for Preparticipation Cardiovascular Screening of Competitive Athletes

Physical examination

9. Heart murmur‡
10. Femoral pulses to exclude aortic coarctation
11. Physical stigmata of Marfan syndrome
12. Brachial artery blood pressure (sitting position)§

*Parental verification is recommended for high school and middle school athletes.
†Judged not to be neurocardiogenic (vasovagal); of particular concern when related to exertion.
‡Auscultation should be performed in both supine and standing positions (or with Valsalva maneuver), specifically to identify murmurs of dynamic left ventricular outflow tract obstruction.
§Preferably taken in both arms.
Case 1 “A close call”

“Buddy” 13 year old male 8th grade school and club basketball player

Chief Complaint: chest tightness, shortness of breath, dizziness,

? Asthma
Buddy – age 13
**Buddy**

**History:**
- 13 y.o. male became symptomatic playing basketball after 3rd game in a tournament.

- Had chest tightness, shortness of breath and dizziness. Looked pale and sweaty.

- A registered nurse in attendance stated he had a normal pulse. Did not return to play.

- He drank fluids, went home and Sxs resolved after 4 hrs.

- Parents concerned he may have “sports asthma” (+ family Hx)
Buddy

PMH: ADHD; Very Rapid growth spurt in previous 6 months

Medications: Vyvanse 30 mg, Multivitamins, Vitamin D 10,000 units daily

FH: Positive for ADHD, Asthma; Negative for cardiac disease. No cigarette smoke exposure.

Note: Patient had no symptoms whatsoever during the previous fall football season
**PE:**

- Tall thin 13 yo male in no acute distress.

- Ht. 176cm, wt. 56kg, BP 100/64, pulse 72

- Heart = regular, without murmur

- Lungs = Clear, normal resp effort

- Neuro/extremities = normal
Buddy

**Labs** = normal chemistry panel, Hgb=12.3

**CXR** normal

**EKG** = prominent mid-precordial voltage, possible biventricular hypertrophy.

  Over-read = “Normal for teen athlete”.
**Pediatric ECG analysis**

Sinus bradycardia
Prominent mid precordial voltage, Possible Biventricular hypertrophy

**Normal**

Referred by: SMITH

Unconfirmed
Buddy

Assessment:
Exercise induced chest tightness, dyspnea, dizziness with no true syncope. Sxs triggered only by extreme exertion.

Diagnosis:
Exercise-Induced Asthma.

Plan:
Albuterol inhaler 2 puffs 15 minutes prior to exertion. Recheck in 2 weeks. Call sooner if recurrent Sxs or if true syncope occurs. Take multivitamin with iron for mild anemia.
Follow-up: mother calls and states he had another episode, more severe, almost “passed out”.

Action: ECHO Cardiogram ordered same week; athlete held out of sports!

Where to get the ECHO…?
Kim Jankovsky, board certified in ECHOCardiography
RTR, RDMS, RVT, RDCS (Adult) (Pediatric)
Years of Echo Experience  29 years
ECHO results:

“Normal heart structure and function except Left Coronary Artery origin appeared to arise from the Right sinus of Valsalva with an intramural course.

DX: Anomalous Left Coronary Artery
New Plan:

- Stay out of sports

- Discontinue Vyvanse

- Pediatric Cardiology consult at Milwaukee Children’s Hospital
Cardiology Eval:

“Anomalous origin of the left coronary artery from the right sinus of Valsalva with an intramural course through the wall of the aorta before exiting through the left sinus.”

So what does this look like?...
NORMAL Coronary Artery Diagram
Anomalous LEFT Coronary Artery
Anomalous LCA

- Rare (0.047%) and dangerous

- Anomalous LCA originates from the Right Sinus of Valsalva and travels between the Aortic root and trunk of the Pulmonary Artery before supplying the Left Ventrical with its blood supply

- During or immediately after exercise the LCA is compressed by the enlarged Aorta and PA

- LCA may travel within the outer layer of muscle of the aorta ("Intramural Course") causing the angle of takeoff to be pinched, predisposing to obstruction.

- Either scenario results in reduction of blood flow during exercise that can result in cardiac ischemia, arrhythmia and sudden death

- This lesion should be repaired!
Proposed mechanism of ischemia of anomalous left coronary arteries. With exercise, the aorta and pulmonary trunk dilate, producing torsion and additional narrowing of the ostium of the anomalous artery.
“This particular coronary artery anomaly is highly associated with sudden cardiac death in athletes, as the blood supply to the left ventricle is pinched off by the wall of the aorta as it expands during vigorous exercise.”

Surgical “Unroofing” of the LCA is recommended in the near future.
3/18/11 Cardiac Surgery performed: Unroofing of the intramural portion of the LCA, creating a perpendicular origin of the LCA from the aorta.

• Unroofing Procedure: A new coronary ostium is created in the correct sinus and the course between the PA and Aorta is eliminated. The intramural course is no longer present.

(Modified unroofing procedure in anomalous origin of left main coronary artery (LCA) from the right sinus of Valsalva Ann Thorac Surg 1997;64:568-569)
Buddy 3/21/11 Discharged home in good condition on Aspirin 81 mg
March 31, 2011 Outpatient F/U visit: doing well, resume Vyvanse for ADHD, normal activities except sports; continue Aspirin.

April 2011 Spring Break Family Vacation to Mexico!
7/7/11 Follow up visit and stress test/ECHO: Doing well, went 14 minutes 56 secs on treadmill, max HR 203, max BP 130/62 (resting BP=98/40). ECHO portion= normal.

Plan= discontinue Aspirin,

...Return to sports!

Now High School Sophomore in Football and Basketball doing well!
Case 2 – Lightening Strikes Twice!

“Ally”
– 12 year old female 7th grade school and club basketball player in the same school as Buddy from Case 1, presents 2 months after him
Ally

**Chief Complaint:**

- “Asthma symptoms” – chest tightness with exertion.
- …also with follow up of knee pain, hip pain and vomiting – (I *do* run a **primary care** clinic you know!)

**HPI:**

- Long history of asthma and allergic rhinitis.
- Has been experiencing chest tightness with exertion, worsening over last couple months.
- CP seems to resolve with continued activity and has not kept her from continuing in exercise.
- Uses her Albuterol inhaler before games and now *during* the game too.
- When *specifically questioned* she admits to significant **dizziness with exercise**, getting worse in recent weeks. No syncope.
Ally

**PMH:** Allergic Rhinitis, Chronic Sinusitis, Mild Asthma, Migraine Headaches and Cyclic Vomiting Syndrome.

**PSH:** Sinus surgery, Tonsillectomy/Adenoidectomy

**FH:** Negative for heart disease. Parents and 2 siblings well. No cigarette exposure.

**Meds:** Albuterol inhaler PRN

**PE:** Healthy, athletic 12 year old girl. Wt. 47.5kg (71%), Ht. 162cm (92%), P 92, BP 98/60 COR- RRR no murmurs. Lungs – clear EKG – NSR
Ally

**Assessment:** Chest pain and dizziness with exercise

*What did I do with such a case …2 months after my first diagnosis of anomalous coronary arteries?*

**Plan:**

- ECHO Cardiogram ordered.
- Hold out of sports!
Dr. Smith: “We’re ordering the ECHO just to be safe”

Ally: “I don’t have the same thing Buddy has, do I?”

Dr. Smith: “Probably not…but it’s possible”

Ally: “Cool!”

Ally’s Mom: “Now you’re making me scared!”
Ally

Ally with Dr. Smith, a few days after diagnosis
ECHO results:

“Anomalous Right Coronary Artery, originating from Left Sinus of Valsalva with intramural course.”

What?? A second case in the same school in 2 months!

Ally with Buddy, right after Ally’s diagnosis
Diagnosis confirmed at Milwaukee Children’s Hospital by ECHO.

Anomalous Right Coronary Artery

The RCA originates from the **Left** Sinus of Valsalva
Anomalous Right Coronary Artery

RCA arising from the left coronary sinus and taking an interarterial course in a 44-year-old man.

(RadioGraphics March-April 2006 vol. 26 no. 2 317-333Coronary Artery Anomalies: Classification and ECG-gated Multi–Detector Row CT Findings with Angiographic Correlation)
Ally

Exercise Stress Testing: 14 mins, 15 secs (90% for age) with drop in BP and developing chest pain at peak exercise. No ischemic changes; no arrhythmias. Normal pulmonary function studies.

Assessment: Anomalous RCA with hemodynamic changes and symptoms at peak exercise. Likely not as high risk as an Anomalous LCA.

Plan:
   Option 1: No surgery, No high intensity exertional activities (i.e., No Sports!)

   *Note – non surgical option was offered to patient because some evidence suggests that Anomalous Right Coronary Arteries may not be as prone to ischemia – and hence sudden death – as Anomalous Left Coronaries

   Option 2: Surgical Unroofing of the anomalous RCA

Which option did she choose?
Ally - 7/28/11 Surgical Unroofing of Anomalous RCA
Surgical Unroofing of Anomalous Right Coronary Artery
Ally

7/30/11 Post op ECHO = unobstructed RCA segment; Discharged home on Aspirin 81mg. Avoid exertion or chest trauma type activities.

3 weeks post-op
Ally

10/21/11 Exercise Stress Test: 12 mins 2 secs (normal for age, but 2 minutes less than her original). Normal HR and BP response. No CP, no dizziness.

Full Return to Sports!

Ally’s First Shot after surgery!
Ally Tackling Her First Post-Op Wisconsin Winter
Florida Vacation – A Happy Family!
Ally

8 Months Post-op
Anomalous Coronary Arteries – A Discussion

- Coronary arteries finish development at 32 weeks gestation

- The complex embryology can result in abnormal developments of the arteries and their origins

- Some anomalies are of no clinical significance, while others may be lethal (i.e. aLCA)

- Anomalous coronary arteries are a major cause of sudden death in athletes under age 35 = 13% of all sudden death cases, 2nd only to HCM (36%)

Anomalous Coronary Arteries – A Discussion

- Another study found anomalous coronary arteries to be the most common cause of sudden cardiac death in military recruits (61% of sudden cardiac death cases)
  

- Overall incidence abnormal aortic origin of coronary arteries is 0.64% of births

- Abnormal Left Coronary Artery origin from Right Sinus of Valsalva = 0.047%

- Abnormal RCA from Left Sinus of Valsalva = 0.17%

- 2 cases in Two Rivers, WI, pop=11,799: 0.017% There are more out there!


**Conclusion**

**Suspect Coronary Artery Anomalies when:**

- Chest pain or tightness with exercise
- Young Athlete
- Vigorous competitive sports (Basketball, Football, Soccer, Track)
- Symptoms of *dizziness* during or right after exercise
- Symptoms following rapid growth spurt

**Diagnose CAA:**

- ECHO with color flow Doppler in the hands of an *experienced* technician
- Coronary CT Angiography if needed to confirm
- Stress testing to assess level of cardiac function

**Treatment of CAA:**

- Withhold from sports participation
- Surgery to correct anomalies if applicable
Thank You!