Pre-Participation Athletic Cardiac Screening

Kimberly A Krabill, MD
Pediatric and Fetal Cardiologist
Northwest Congenital Heart Care, Division of MedNax

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Case one

14 year old female teen presents with a murmur and history of tachypnea and left midsternal chest pressure with exercise. The pain radiates to her left shoulder. She will have to stop and the symptoms fade with rest.

Family history positive for hypertrophic obstructive cardiomyopathy in mother; cardiomyopathy in maternal uncle and maternal grandmother had heart transplant due to “heart failure”.

PE: BP is 118/76. HR is 62. Grade 2-3/6 harsh SEM at LLSB radiating to URSB.

ECG shows BVH with strain

Echo: Hypertrophic cardiomyopathy with hyperdynamic LV function; mild mitral insufficiency and a left ventricular outflow track gradient of 35mmHg.

What do you not recommend:

1. Holter and Exercise test
2. Let patient play sports
3. Genetic testing
4. Medication, AICD
Sudden Cardiac Death (SCD) in Young Competitive Athletes (YCA)

- “Abrupt and unexpected natural death of cardiac cause, typically within one hour of onset of symptoms and often without an immediately recognizable life-threatening condition”
  Zipes DP and Wellens HJJ, Circ 1998
- When it happens, the “stories” are big ones, they get tremendous press coverage
- They exist at both a national as well as local level
- They are devastating events - Death of a healthy, asymptomatic youth

- SCD is NOT a disease – it is an OUTCOME.
- SCD is caused by multiple, rare underlying heart conditions, usually present from birth.
  - The first symptom is often SCD.
- Traditional forms of structural congenital heart disease (ASD, VSD, aortic stenosis, Tetralogy of Fallot, aortic coarctation) are rarely associated with sudden cardiac death; whether or not they have been repaired.
Importance and Goals of Screening

Use a preparticipation sports form (wiaa.com/ConDocs/Con395/Physical%20Form%202011.pdf)

- Prevent Sudden Cardiac Death or Near Death Events
- An Excellent History and Physical Examination is your key to success
- Personal History:
  - Exertional chest pain/discomfort
  - Unexplained syncope/near-syncope
  - Judged not to be neurocardiogenic (vasovagal); of particular concern when related to exertion
  - Excessive exertional and unexplained dyspnea/fatigue, associated with exercise
  - Prior recognition of a heart murmur
  - Elevated systemic blood pressure
- Family History:
  - Premature death (sudden and unexpected, or otherwise) before age 50 years due to heart disease, in 1 relative
  - Disability from heart disease in a close relative 50 years of age
  - 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
- Physical Exam:
  - Vital signs including BP
  - Heart murmur; pulses; Physical traits (syndromic)
Causes of Sudden Cardiac Death

“Other”
- Commotio cordis
- LQTS
- WPW
- Brugada syndrome
- Catecholaminergic PMVT
- Drugs
- Sickle cell trait
- Blunt neck trauma
- Intracranial artery rupture
- Sarcoidosis
- Heat stroke
- Asthma
- Sinus node artery narrowing
Hypertrophic Cardiomyopathy

- Incidence in general population 0.2 - 0.5%
  - Annual mortality 2-3%
  - More common in African Americans
- Idiopathic subaortic stenosis (IHSS) in 25%
- Half present with sudden death
- Other half present with syncope, presyncope, chest pain, palpitations, dyspnea on exertion, fatigue, heart murmur, or positive family history.

- Subaortic murmur at lower to mid-left sternal border that may radiate to URSB or neck.
- Murmur increases with Valsalva maneuver or sitting up. (decreased venous return and increased sympathetic tone worsen the dynamic muscular obstruction)
- May hear an S3, S4, or find a laterally displaced PMI (fairly nonspecific)
- May see cardiomegaly on CXR (nonspecific and not always present)
Hypertrophic Cardiomyopathy

• ECG: Abnormal ECG in 90%
  • Abnormal ECG in 96% with SCD
  • LVH with high voltage QRS and T waves
  • Septal Q waves
  • Deeply inverted precordial T waves
  • A normal ECG does not rule out HCM.

• Echo: Definitive diagnosis
  • Patients with an abnormal ECG or
  • Suggestive history, family history, or examination
  • Must have echocardiography to diagnose
Coronary Artery Anomalies:

- Rare (0.06%-0.3%) structural abnormalities leading to compromised coronary blood flow:
  - Single coronary
  - LCA from right sinus or RCA from left sinus
  - Anomalous LCA from the pulmonary artery (ALCAPA)
  - Bridging muscle bands causing coronary compression (tunneled LCA or myocardial bridge)
  - Coronary ostial stenosis: Rare cases of SCD as infants

- LCA from right sinus of Valsalva is rare (0.03%)
  - 29/381 and 36/492 cardiac death
  - 23/23 <20 yrs died suddenly with exercise
  - Majority exercise-induced SCD were asymptomatic

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

1Roberts WC, Am Heart J 1986; 111
2Taylor AJ et al , J Am Coll Cardiol 1992; 20
3Frescura C et al, Hum Pathol 1998; 29
Coronary Artery Abnormalities; Possible mechanism

Roberts WC, Am J Cardiol 1982; 49: 863-868
Coronary Artery Anomalies: Clinical Presentation/ Management

• Usually victims are asymptomatic and present with SCD.
  • Some will complain of chest pain.
• Rarely ischemic changes seen on ECG.
• Most (90%) can be diagnosed by echo.
  • Often an incidental finding
• Require surgical correction?

• Most experts agree that anomalous LCA has greater risk of SCD and should be operated on.
• No data to demonstrate that surgery alters the natural history of anomalous RCA.
• Hard to determine efficacy with uncommon events over long periods of time
• The question is: what is your threshold of comfort? Error of commission or error of omission?
Long QT Syndrome

• Prevalence of about 1:2,000
• Prolongation of QT interval is hallmark of LQTS
• Between 10% (LQT3) and 37% (LQT1) of genotype-positive patients have a QT interval within normal limits at rest
• Arrhythmic events are due to runs of Torsades de Pointes VT which, according to its duration, produces syncope, cardiac arrest, and – when it deteriorates into VF – sudden death.

• 13 genetic forms of congenital LQTS caused by mutations in genes encoding potassium-channel proteins, sodium-channel proteins, calcium channel-related factors, and membrane adaptor proteins.
• Patients with LQT1, LQT2, and LQT3 genotypes with mutations involving KCNQ1, KCNH2, and SCN5A make up over 92% of patients with genetically confirmed LQTS.
• 15-20% of patients with LQTS remain genetically elusive
Long QT Syndrome

- Abnormal ventricular depolarization
- Syncope from Torsades
- Inherited or acquired

Tx:
- Beta-blockers
- Pacing
- AICDs
Long QT Syndrome:

- Schwartz criteria

LQTS IS DIAGNOSED:
- In the presence of an LQTS risk score > 3.5 in the absence of a secondary cause for QT prolongation, and/or
- In the presence of an unequivocally pathogenic mutation in one of the LQTS genes, or
- In the presence of a QTc > 500 ms in repeated 12-lead ECG and in the absence of a secondary cause for QT prolongation.

LQTS can be diagnosed in the presence of a QTc between 480-499 ms in repeated 12-lead ECGs in a patient with unexplained syncope in the absence of a secondary cause for QT prolongation and in the absence of a pathogenic mutation.

New criteria HRS ‘13

Circulation. 1993; 88: 782-784
Case Two

10 year old boy presents with exertional chest pain and he has a history of Job syndrome.

Pain is squeezing in quality, occurs with running and wrestling. He stops, holds his chest for 1-2 minutes and pain resolves. No associated palpitations or syncope.

PE is unremarkable.

ECG shows sinus rhythm and is normal for age.

ECHO shows anomalous origin of RCA from left sinus with a narrowed orifice and likely intramural course

Thallium CPX: Normal

Coronary CTA: anomalous origin of the right coronary artery from the left aortic sinus at and takes an interarterial course between the main pulmonary artery and ascending aorta near the level of the pulmonic valve plane. The angle of origin is acute. The proximal 10 mm of the right coronary artery has features consistent with an intramural course, and is diminutive in size with lateral compression of the lumen

What would you do with this patient?

1. Let exercise and follow conservatively
2. Operate
3. Restrict exercise
4. Both 2 and 3