

Hearts Too Young To Die:

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MEETINGS

None of us is as dumb as all of us.

Today's Discussion

- What cardiac diseases cause sudden death in young patients?
- How do we identify them?
 - What symptoms are worrisome
 - How should we screen

– ~~Genetic testing~~



Lethal Cardiac Disease in the Young

- **Congenital Heart Disease**
 - Aortic stenosis, coronary anomalies
- **Acquired Heart Disease**
 - Myocarditis, atherosclerosis, coronary artery disease
- **Arrhythmogenic syndromes**
 - Hypertrophic cardiomyopathy
 - Arrhythmogenic RV dysplasia
 - Ion channelopathies
- **Systemic syndromes (Marfan's)**



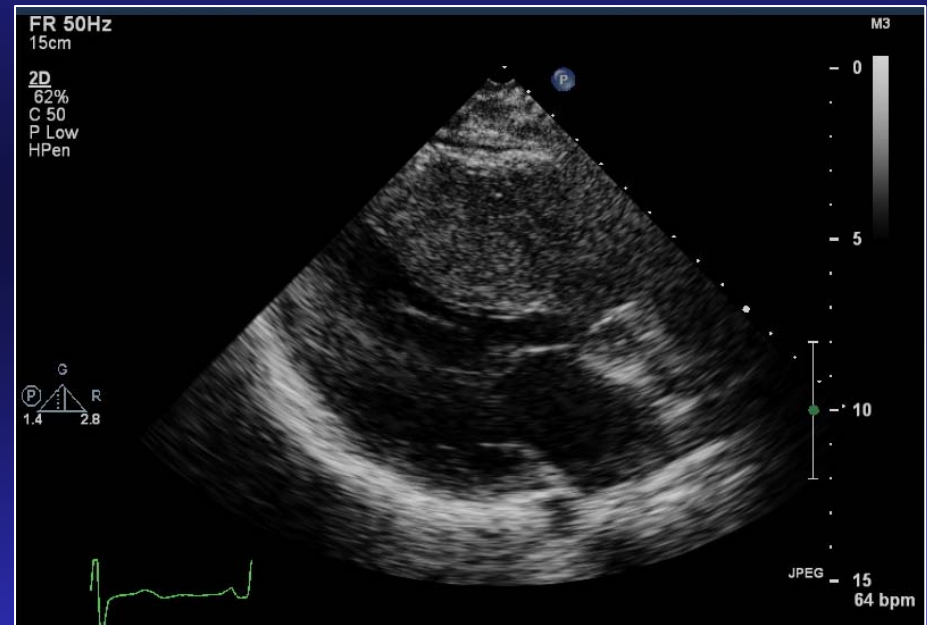
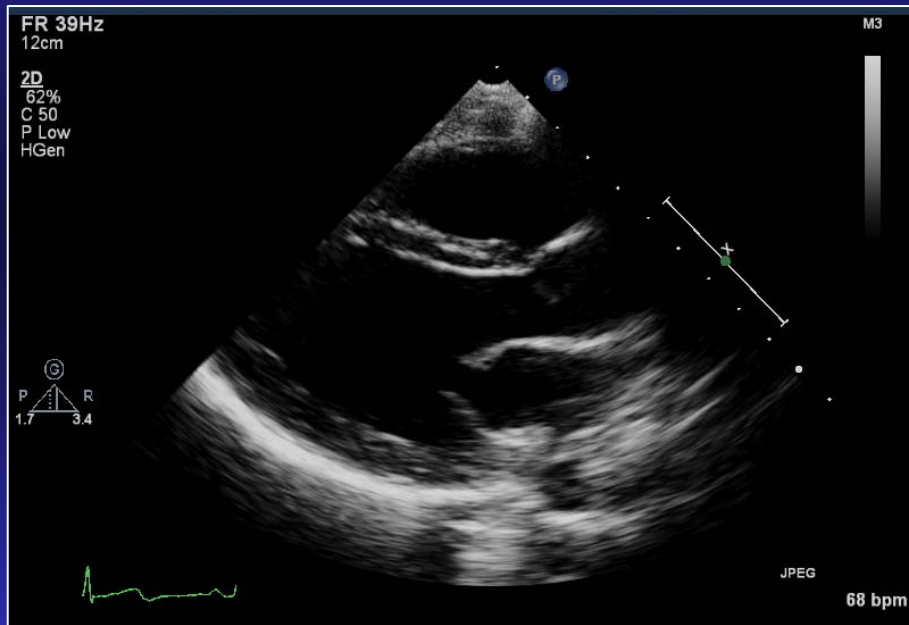
Table 1. Causes of Sudden Death in 387 Young Athletes.*

| Cause | No. of Athletes | Percent |
|--|-----------------|---------|
| Hypertrophic cardiomyopathy | 102 | 26.4 |
| Commotio cordis | 77 | 19.9 |
| Coronary-artery anomalies | 53 | 13.7 |
| Left ventricular hypertrophy of indeterminate causation† | 29 | 7.5 |
| Myocarditis | 20 | 5.2 |
| Ruptured aortic aneurysm (Marfan's syndrome) | 12 | 3.1 |
| Arrhythmogenic right ventricular cardiomyopathy | 11 | 2.8 |
| Tunneled (bridged) coronary artery‡ | 11 | 2.8 |
| Aortic-valve stenosis | 10 | 2.6 |
| Atherosclerotic coronary artery disease | 10 | 2.6 |
| Dilated cardiomyopathy | 9 | 2.3 |
| Myxomatous mitral-valve degeneration | 9 | 2.3 |
| Asthma (or other pulmonary condition) | 8 | 2.1 |
| Heat stroke | 6 | 1.6 |
| Drug abuse | 4 | 1.0 |
| Other cardiovascular cause | 4 | 1.0 |
| Long-QT syndrome§ | 3 | 0.8 |
| Cardiac sarcoidosis | 3 | 0.8 |
| Trauma involving structural cardiac injury | 3 | 0.8 |
| Ruptured cerebral artery | 3 | 0.8 |

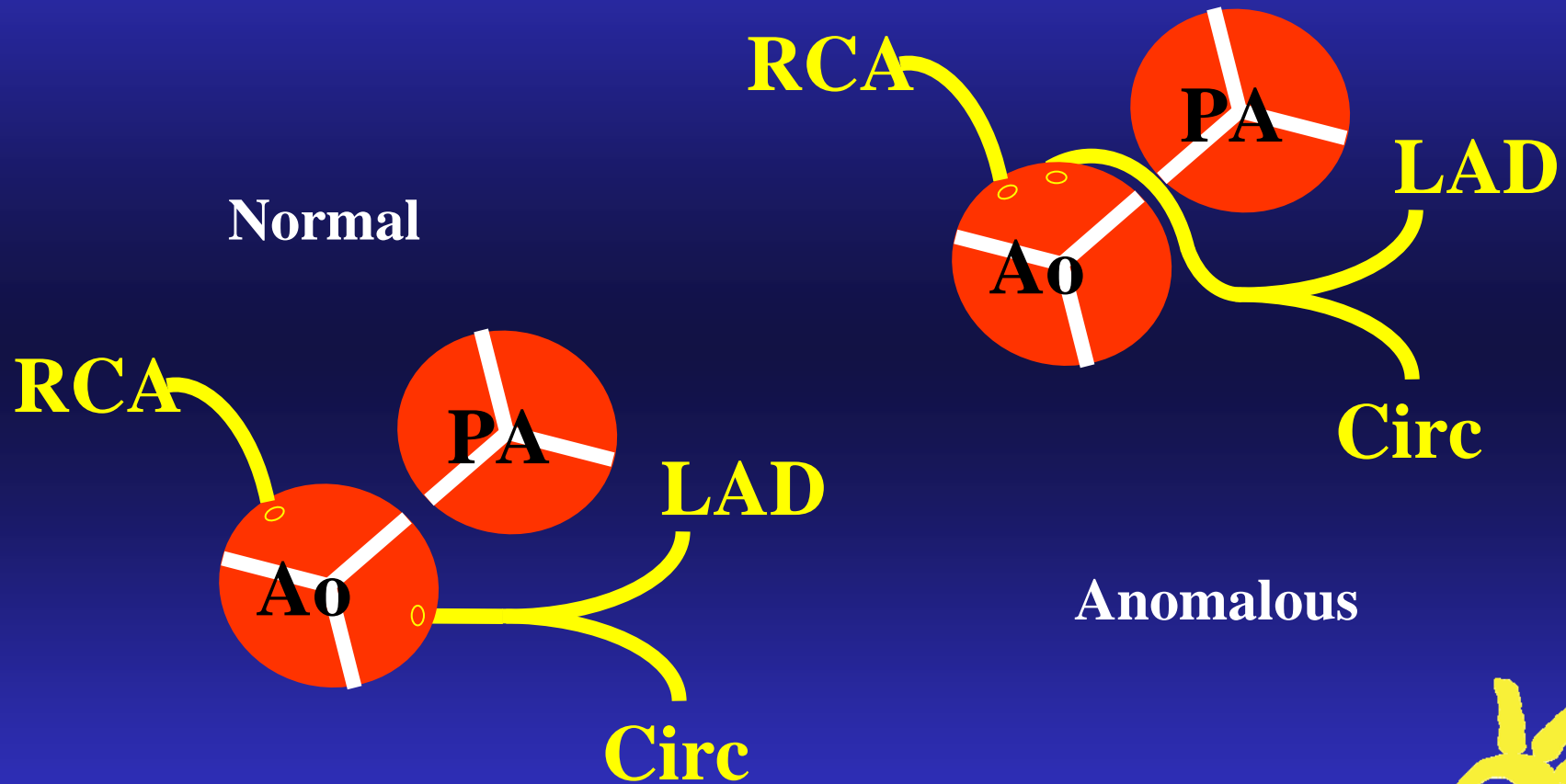
Maron BJ. Sudden Death in Young Athletes. NEJM 2003; 349: 1064-75.



Hypertrophic Cardiomyopathy



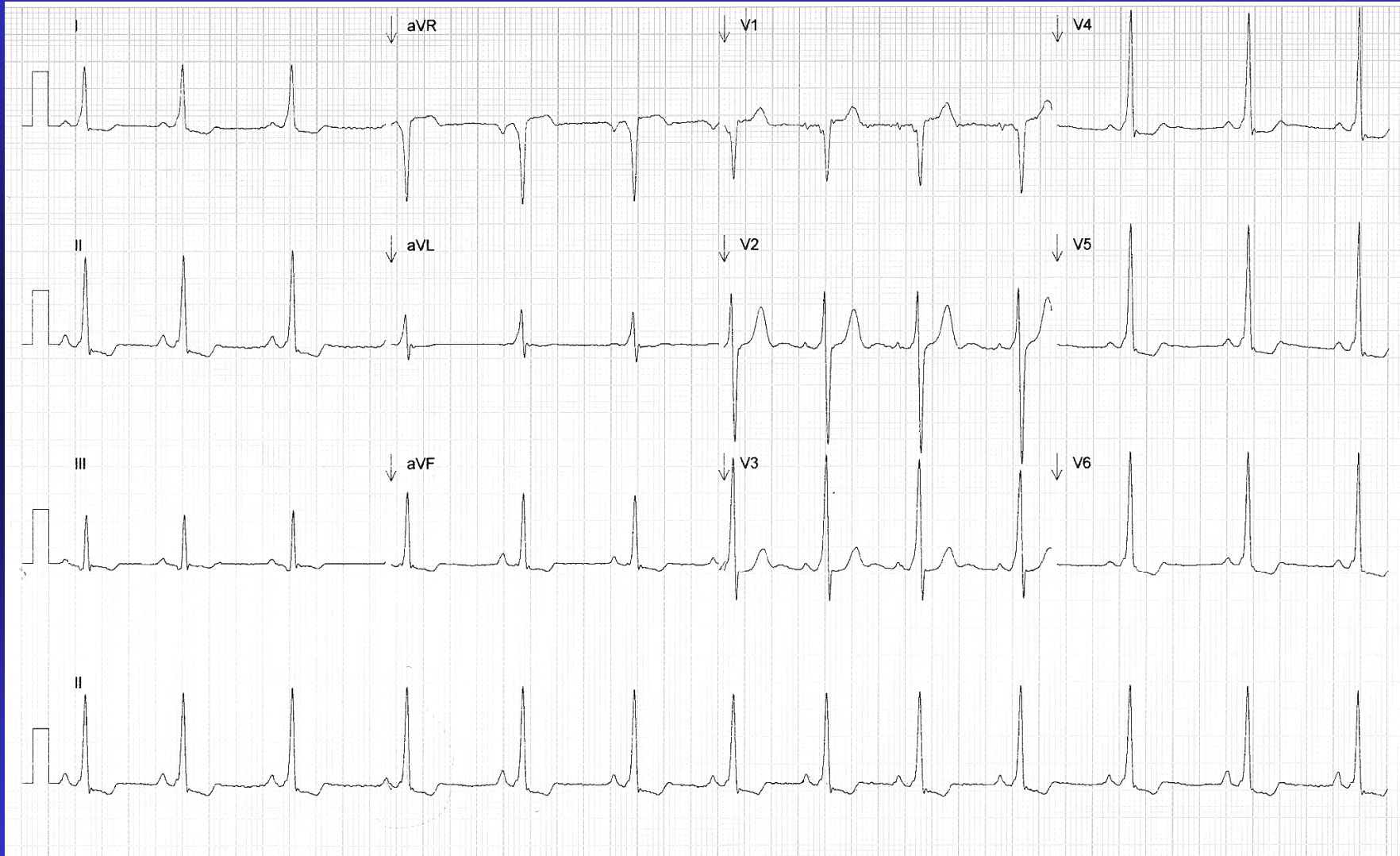
Anomalous Coronary Origins



Long QT syndrome



WPW



Other arrhythmogenic syndromes

- Brugada syndrome
- Polymorphic catecholaminergic VT
- Idiopathic ventricular fibrillation



Common features

- Rare
 - Inheritable
- Often silent
- Normal exam
- Predisposition to sudden cardiac death
 - Sudden death may be 1st symptom
- Risk exacerbated by exertion



Common Symptoms

- Syncope, Seizure, Sudden Cardiac Death
- Palpitations, Dyspnea
- Chest pain



Risk stratification of SCD

- Retrospective review of 134 athletes with sudden death
 - 3% Had suspected disease based on history and physical
 - Symptoms were predominantly exertional.
 - Syncope was chief among them

Maron BJ et al. Sudden death in young competitive athletes. Clinical, demographic, and pathological profiles. JAMA 1996; 276: 199-204.



Worry Index

- Very worrisome
 - Exertional syncope / seizure
 - Syncope / seizure while in water
- Moderately worrisome
 - Other exertional symptoms (CP, palps, dyspnea)
 - Syncope at rest, but with other cardiovascular symptoms
- Possibly worrisome
 - Unexplained syncope, palpitations at rest
- Not worrisome
 - Isolated CP, Vasovagal syncope



Arrhythmogenic Syndromes

- How do we screen?
- Current standard set by AHA for athletic screens.
 - Family history
 - Personal history
 - Examination
 - Additional testing is discretionary



Are athletes at higher risk?

YES!

- **Relative risk of sudden death ~ 2.5 for athletes vs non-athletes**

Corrado et al. Does Sports Activity Enhance the Risk of Sudden Death in Adolescents and Young Athletes? JACC 2003; 1959-63.



Stimulant Risk?

????? !

- Relative risk unknown
 - May be slightly higher
- Consensus statement 2008 recommended EKG for all patients starting stimulants
- **PAPER HAS BEEN DISCREDITED !!!**



Do Stimulants Require an EKG?

- NO
- Perform routine “athletic screen”
 - If unremarkable, you are well with in standard of care to begin stimulants without EKG



Effect of Current Screen

- Screening policies in the U.S. appear to be ineffective at reducing sudden cardiac death in adolescents and young adults.
-



Athletic Screening: Echo or EKG?

- United States: ~75 sports related cardiovascular deaths / year
- 70% of the cardiovascular conditions are identifiable by EKG or Echo
- US: estimated 12.5 million athletes
 - Could potentially save 50 lives
- Indiana: estimated 255,000 athletes
 - Could potentially save 1 live

Maron, BJ et al. *Circulation* 2009;119:1085-1092

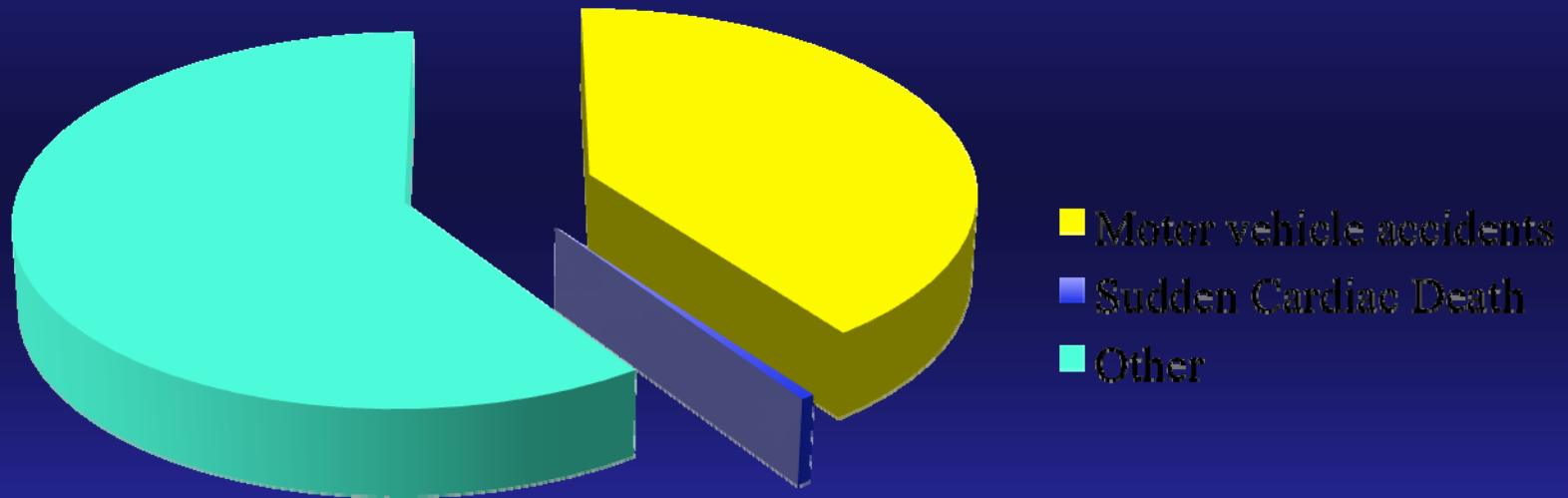


Question of the Day

- What is the leading cause of death in athletes?
 - Hypertrophic cardiomyopathy
 - Arrhythmogenic right ventricular dysplasia
 - Coronary anomalies
 - Other



Causes of Teenage Deaths, 2006



2006 and 2007: 76 Sudden Cardiac Deaths each year



AHA Conclusion

- “... it is not prudent to recommend routine use of such tests as [EKG], [echo], or graded exercise testing...”
- AAP
- ACP
- ACSM



Len's Laws for Identifying Children at Risk

1. You can't identify all children at risk
2. The use of elaborate, expensive CV testing cannot change rule #1
3. Test all family members of index cases
4. Maintain high index of suspicion
 - In the right circumstances
5. Go ahead, get that echo / EKG.



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Risk Stratification: Additional Eval

- Family may have to pay out of pocket for testing in asymptomatic patients
- Consider
 - EKG instead of echo for additional screening of asymptomatic athletes
 - Listening VERY CAREFULLY for a murmur
- Children's Heart Center at the Care Group offers affordable pre-participation cardiac evaluation
 - H&P
 - EKG
 - Complete echocardiogram

