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Why Texas High School Athletes Die Suddenly: Unexpected Cardiac Death in Childhood

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Key Points

- The sudden death of athletes during competition is quite rare, approximately nine per year in Texas.
- This paper explains the three most likely reasons for "sudden cardiac death" in high school athletes during sports.
- Effective mass screening to prevent these deaths is nearly impossible and prohibitively expensive.
- A physician can diagnose and cure two of these three causes, but warning signs must first be present.

Problem Statement

- Approximately 100 athletes under age 20 die suddenly and unexpectedly each year in the U.S. during competition. Among them, 9 percent—or nine deaths—happen in Texas. For comparison, 4,054 teenagers die each year in auto accidents in the U.S.
- These seemingly inexplicable deaths are called Sudden Cardiac Death (SCD).
- Reliable demographic data is sparse, and autopsy results are very limited. Most conclusions are based on anecdote, inference, and guesswork.
- The three most likely reasons—each described below—for SCD are:
  I. Hypertrophic cardiomyopathy (HCM)
  II. Coronary artery [coursing] between the great arteries (CBGA)
  III. Rapid heart beat, or rhythm disturbance
- This discussion excludes traumatic causes of death such as bleeding in the brain from boxing or football, projectile injury during shooting competitions, auto or bicycle accidents during races, etc.

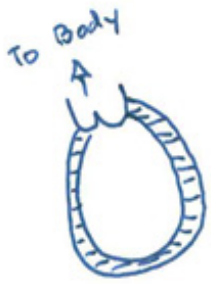
I. Hypertrophic cardiomyopathy

- Also called: idiopathic hypertrophic subaortic stenosis (IHSS), and asymmetrical septal hypertrophy (ASH).
- Diagnosis made by:
  - *Electrocardiogram* (ECG; cost ≈$75): limited usefulness because of too many false positive and false negative results.
  - *Echocardiogram* (cost ≈$2,000): only reliable method, 100 percent accurate, and gives both anatomy as well as severity.
- Treatment: There are both drug treatments and surgical techniques that may reduce the risk, but no treatment eliminates the risk of sudden death. Children have died despite having had appropriate treatment. Even with treatment, a guarantee of safety in children with HCM is not currently possible.
- Drawings that follow explain how HCM can cause sudden cardiac death.

continued

Hypertrophic Cardiomyopathy can cause sudden cardiac death, especially during exercise

Normal heart, filling

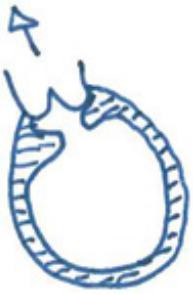


When the heart is filling with blood, the wall is relaxed to allow the blood to flow in. The aortic valve, leading out of the body, is closed.

Normal heart, ejecting blood

To force the blood out into the body, the muscles in the heart wall all contract. This "squeezes" the blood out of the pumping chamber and opens the valve. There is nothing getting in the way of the blood going to the body.

Heart with HCM, filling



With HCM, the part of the heart wall muscle just under the aortic valve is abnormally thick. During filling, this has no life-threatening consequence, but the abnormal thickness does make it harder for blood to flow in.

Heart with HCM, ejecting

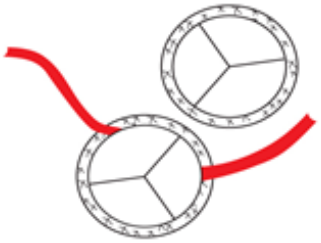
When the heart starts to force blood toward the body, all the muscle fibers contract, including the ones under the valve, the same ones that are too thick. This obstructs the flow. To compensate, the heart wall gets thicker both in general and specifically where it is already too thick. This makes things worse and can lead to a vicious cycle causing sudden cardiac death.

II. Coronary Between Great Arteries (CBGA)

- Diagnosis:
  - *ECG* is virtually useless.
  - *Echocardiogram* by highly experienced sonographer can suggest CBGA.
  - *MRI* (magnetic resonance imaging; cost ≈$2,500) can make diagnosis but can also miss the abnormal coronary course.
  - *Angiography* (cost ≈ $4,000) is the best, surest way to make diagnosis. It requires insertion of cardiac catheters and injection of contrast directly into the coronary arteries to define their precise position.
- Treatment: It is possible with surgery to move the coronary so it is no longer in danger of being compressed. Surgery is curative and carries very low risk.
- Drawings that follow explain how CBGA can cause sudden death.

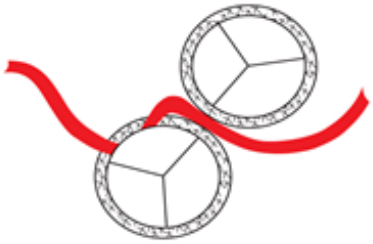
Normal coronary arteries

Coronary arteries come out of the aorta, sending blood to the right and left pumping chambers. No coronary runs between the great arteries.



CBGA, at rest

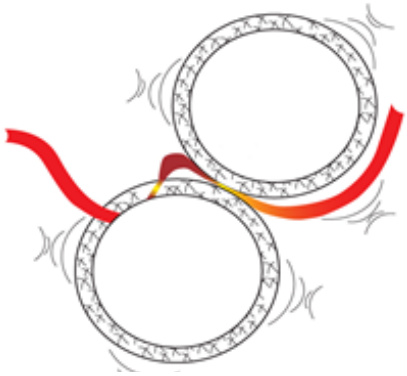
The left coronary artery arises from right side of the aorta and must pass between the great arteries to give blood to left side of heart.



CBGA, during exercise

When there is CBGA and the heart is pumping hard and fast, as during athletics, both great arteries pulsate and compress the artery passing between them.

At the very time that more blood is needed—during strenuous exercise—the obstruction makes less blood available. This can cause sudden cardiac death.



III. Rhythm Disturbance

- In general, there are three kinds of cardiac rhythm disturbances: too slow, too fast, and irregular. The last does not cause sudden death. The first almost never occurs in children. "Too fast" is the athletes' problem.
- There are many different forms of "too fast." They can come on suddenly without any warning.
  - The most common form of "too fast," called Wolff-Parkinson-White is almost never fatal on the first or second episode.
  - By contrast, the genetic disorder called Long QT Syndrome can be fatal the first time it is manifest.
- Diagnosis:
  - *ECG*: If an ECG happens to capture an abnormal rhythm, then the ECG can make the diagnosis. However, a normal ECG does not rule out episodes of "too fast."
  - *Holter monitor* (cost ≈ $500): 24 or 48 hours of continuous ECG recording is more likely to pick up a "too fast" event, even if it is brief and self-limited. But if events only happen occasionally, Holter may miss it.
  - *Event monitor* (cost ≈ $750): a device the patient carries for 2-8 weeks going about all normal activities. Average cost for six weeks is $750. When patients feel something that is not right, they push a button on the device and recording starts. This digital record is then transmitted electronically to the doctor's monitoring station.
  - *Electrophysiologic cardiac catheterization* (cost ≈ $5,000): the best way, sometimes the sole way, to diagnose the exact problem. Moreover, it is the only way to truly cure the patient by permanently altering how electrical impulses flow inside the heart.
- Treatment: Most forms of "too fast" can be suppressed with medications. There are some rhythm problems that go away on their own. Some require a cardiac catheter procedure to get rid of an extra or an abnormal electrical pathway inside the heart.

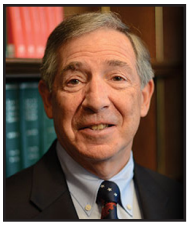
References

American Heart Association. 2016. "Hypertrophic Cardiomyopathy." Last updated March 2016.

Bezold, Louis I. et al. "Coronary Artery Anomalies." Medscape, January 5, 2015.

Ellis, Christopher R. et al. "Wolff-Parkinson-White Syndrome." Medscape, January 8, 2017. Note: there are many other forms of "too rapid heart beat." Wolff-Parkinson-White is probably the most common and certainly the best known.

Sovari, A. A. 2015. "Long QT Syndrome." *E-medicine*, December 31.



About the Author

Dr. Deane Waldman, MD, MBA, is the director of the Center for Health Care Policy at the Texas Public Policy Foundation. He is a retired pediatric cardiologist and system theorist analyst for American health care. He brings 37 years of clinical and administrative experience as chief of Pediatric Cardiology at University of Chicago.

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