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A high school athlete with a single coronary artery passing between the great vessels

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The preparticipation physical examination in the athlete is commonly encountered and performed in Family Practice. The primary care physician needs to take care during this examination to be alert to the various risks and warning signs that may signal a risk for sudden cardiac death. This case involves a 17-year-old athlete and a single coronary artery passing between the great vessels.
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Case

A 17-year-old high school basketball player and track athlete presented for a routine preparticipation examination, i.e., sports physical. On his intake questionnaire, he denied any past medical history or any previous instances of chest pain, shortness of breath, or fainting regardless of physical activity. He also wrote on the intake form that he had no family history of sudden death.

During the examination, all of the same questions were asked of the student. His answers differed. He admitted that he sometimes had "sharp pains through my heart," which he said occurred at rest but not during physical activity. The pain went from sharp to dull, lasting 4 to 5 minutes. It was a shooting pain and had last occurred two weeks before this examination. He denied any other related symptoms, signs, or radiation of the pain.

The remainder of the examination was as follows:

17-year-old African-American male; height: 5'10 ½"; weight 150 lbs; blood pressure 118/72; pulse 60 bpm; temperature 96.3 °F.
General: No acute distress; awake, alert, and oriented to person, place, and time
Heart: Irregular; 60 bpm with a III/VI systolic murmur loudest at the third left ntercostal space, which increased with Valsalva maneuver and inspiration; questionable ectopy
Lungs: Clear to auscultation bilaterally without rales, rhonchi, or wheezes
Chest: Contour normal
Tanner: Stage 5
Back: Full range of motion and without scoliosis
Extremities: No cyanosis, clubbing, or edema
Neuro: Cranial nerves II through XII were intact; no focal neural deficits appreciated

The remainder of his physical examination, including endocrine and lymphatic, was grossly unremarkable.

Based on the history and physical examination, an electrocardiogram was performed. It showed: sinus bradycardia at 45 bpm, QRS axis 80 degrees, QT interval normal. The rate was regular. There were no Q waves or ST changes.

Because of the nature of the murmur, the patient was sent for a cardiac evaluation and echocardiography. The patient's father accompanied him to his cardiac evaluation. The patient denied any past medical problems; however, his father added that the patient had Kawasaki's disease as a toddler. This information was neither provided at the initial exam by the patient nor written on the preparticipation form.

The two-dimensional echocardiogram revealed a questionable coronary artery fistula, possible anomalous right

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coronary origin from the left sinus of Valsalva, possible recanalized coronary aneurysm. The right coronary artery was described as irregular and ectatic. A low-velocity flow appeared to go from right to left and coursing between the aortic root and pulmonary artery. He was sent for further imaging and workup. The history of Kawasaki's disease suggested a possibility for a coronary artery aneurysm that healed by recanalization, possibly appearing like ectasia.

The further workup revealed that the left coronary artery originated from the single, right anomalous coronary artery and that the left passed between the great vessels. This carries a risk for sudden cardiac death. The patient then visited with a pediatric cardiothoracic surgeon.

Computed tomographic angiography confirmed the single right coronary artery arising from the left sinus, and the left coronary artery arising from the right artery and traversing between the great vessels. After the angiography, maximal exercise stress test, stress echo, and stress perfusion scans all were normal. The unique anatomy of his coronary artery origin—his left coronary artery passing between the great vessels and the coronary vessels not running intramurally—make surgical options limited and risky.

Discussion

High school and collegiate athletes routinely have preparticipation examinations to ensure that they are healthy for competitive sports. One of the goals is "recognition of 'silent' cardiovascular abnormalities that can progress or cause sudden cardiac death."[1] Often, the family physician is called on to perform this examination. A thorough history is an important precursor to the examination. Many times key information may not be learned until a third history is performed. During the examination, the doctor may appreciate a murmur, an irregular heartbeat, obtain information that the athlete has experienced chest pain, shortness of breath, or syncope—sometimes during exertion. All of these symptoms must be taken seriously, especially because chest pain and syncope can be signs of coronary artery anomalies and other cardiac problems that could result in sudden cardiac death.[2]

Sudden cardiac death is an unexpected and abrupt death with a cardiac cause and occurs without any other potential fatal conditions.[3] Death usually occurs within one hour from the onset of symptoms.[3] When this happens in a young athlete, it is often catastrophic because of the highly publicized nature of the event given that competitive athletes are often viewed as some of the healthiest members of their community.[4] Congenital coronary arteries are one of the common causes of exercise-induced/related sudden cardiac death in young athletes as a result of myocardial ischemia.[4] Symptoms preceding death in these athletes may be syncope during exertion or rest, typical and atypical angina, palpitations with and without exertion, and dizziness during activity.[4] Interestingly, in the study by Basso et al., none of the athletes who had symptoms before death had a family history of cardiovascular disease or sudden cardiac death.[4]

The incidence of congenital coronary artery anomalies is approximately 0.5 to 1.5% based on adults having coronary artery angiography.[5] However, specific congenital conditions may lead to a higher incidence of anomalies, such as tetralogy of Fallot.[6] Although most anomalies are benign, anomalies in which the coronary artery passes between the great vessels is a cause for sudden death, particularly in young athletes.[5] Approximately one-third of coronary artery anomalies result from the coronary artery having an anomalous origin from the aorta, including arising from the wrong aortic sinus of Valsalva or being located too high or low in the correct sinus, but distorting the shape of the arterial ostium.[7] An anomalous left coronary emerging from the right and passing between the great vessels is associated with sudden cardiac death, usually during strenuous activity, becuase of dilation of the vessels, which in turn causes torsion or kinking of the left coronary artery and leads to ischemia.[7,8] Despite the incidence being greater when the left coronary artery emerges from the right sinus, both anomalies have a high association with sudden death.[8]

The presence of a single coronary artery is even rarer, with an incidence of approximately 0.04 to 0.066%.[2,7] These patients often have other congenital cardiac abnormalities that may include one coronary supplying the entire heart muscle while the other artery is absent (type 1) or the single artery dividing into branches and forming the two arteries (type 2).[7]

Task Force 2 regarding congenital heart disease as published in the *Journal of the American College of Cardiology,* and distributed as part of the 2005 Bethesda Conference Guidelines, reviews various congenital defects.[9] The guidelines regarding congenital coronary artery anomalies make the following recommendations:

1. Detection of coronary anomalies of wrong sinus origin in which a coronary artery passes between great arteries should result in exclusion from all participation in competitive sports.
2. Participation in all sports three months after successful operation would be permitted for an athlete without ischemia, ventricular or tachyarrhythmia, or dysfunction during maximal exercise testing.
3. Athletes with previous myocardial infarction (MI) should follow the appropriate recommendations in Task Force 6: Coronary Artery Disease.[9]

The clear position is that surgery is required. Although there are surgeries available for various coronary artery anomalies, the specific anomaly dictates the surgery as well as the specific risks.[5]

Conclusion

The preparticipation physical is important before allowing an athlete to engage in competitive sports. The physician should not only be aware of the many risks and warning signs that may be learned during a proper history and

physical, but they must be prepared to discuss with the patient and family the consequences of allowing an athlete to compete when the athlete is at risk for a cardiac event, especially sudden cardiac death. The athlete described in this paper is currently exploring surgical remedies, but, unfortunately, his anomaly is twofold. His right coronary artery comes off the left sinus; his left coronary artery is a branch-off of the right and passes between the great vessels.

Epilogue

This athlete in this case study completed his freshman year of college without having undergone the corrective surgery. He still had not decided if the benefit to him outweighed the risks of the procedure.

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