Sudden Death in Young Athletes

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SUMMARY  The causes of sudden and unexpected death in 29 highly conditioned, competitive athletes, ages 13–30 years, are summarized. Sudden death occurred during or just after severe exertion on the athletic field in 22 of the 29 athletes. Structural cardiovascular abnormalities were identified at necropsy in 28 of the 29 athletes (97%), and in 22 (76%) were almost certainly the cause of death. The most common cause of death in this series was hypertrophic cardiomyopathy, which was present in 14 athletes. Other cardiovascular abnormalities that occurred in more than one athlete were anomalous origin of the left coronary artery from the right (anterior) sinus of Valsalva, idiopathic concentric left ventricular hypertrophy, coronary heart disease and ruptured aorta. Cardiac disease was suspected during life in only seven of the 29 patients, and in only two of the seven was the correct diagnosis made clinically. Hence, in this series of young athletes, sudden death was usually due to structural cardiovascular disease, and hypertrophic cardiomyopathy was a frequent cause of sudden death; atherosclerotic coronary heart disease was relatively uncommon.

ALTHOUGH YOUNG, competitive athletes epitomize the conditioned, healthy segment of society, sudden and unexpected death does occur in such individuals.1,4 However, no large series of sudden deaths in athletes has been systematically compiled, and therefore the principal causes of such deaths have not been adequately defined. Because such information might help to identify athletes who are “at risk” of sudden death, the present study was undertaken. In this investigation we have attempted to determine the causes of sudden, unexplained death in a group of highly competitive athletes.

Materials and Methods

Selection of Patients

The 29 athletes with sudden and unexpected death who constitute the study group were initially identified from the following sources: (1) prospectively from news media reports of sudden death in competitive athletes from 1976–1979 (17 athletes: two died in the Washington D.C. metropolitan area and the other 15 died elsewhere); (2) retrospectively from review of the autopsy registry of the Cardiovascular Division of the Armed Forces Institute of Pathology, 1950–1978 (seven athletes who were military dependents and whose hearts were referred for consultation because of the occurrence of sudden death and suspicion of heart disease); and (3) retrospectively from review of the autopsy registry of the Pathology Branch, National Heart, Lung, and Blood Institute (NHLBI) (five athletes). The hearts of four of these patients had been initially referred to the NHLBI for consultation because of the occurrence of sudden death and suspicion of heart disease; subsequently these patients were identified as having been competitive athletes during life. Six of the 29 study patients have been included in published studies.5,6

For inclusion in the study each patient must have been an active and highly conditioned member of an organized athletic team for at least 2 years at the junior high school, high school, collegiate or professional level. Four of the 29 athletes had withdrawn from organized team sports 1–5 years before death, but had continued to maintain a high level of conditioning. It also was necessary that a complete necropsy examination be performed in each patient to exclude trauma-related causes of death and that, in particular, the heart be in suitable condition to permit examination.

Toxicologic tests for levels of alcohol, barbiturates and amphetamines were performed postmortem on blood from 15 patients, and the levels of each were normal. None of the 29 athletes had a history of drug usage.

Necropsy Studies

In addition to gross visual examination, measurements were made of the maximum thickness of the ventricular septum and posterior left ventricular free wall. Trabeculae, papillary muscles and crista supraventricularis were not included in the measurement of wall thickness. The extramural coronary arteries were studied in the intact specimen by making multiple cross sections of the vessels (3–5 mm apart) and viewing the arterial lumens grossly.

Blocks of myocardium were taken from the ventricular septum for histologic study of cardiac muscle cell arrangement. When pertinent, the extent of cellular disorganization was assessed in tissue sections by a previously described quantitative method.7

In four athletes (three with idiopathic concentric
left ventricular hypertrophy and one with hypertrophic cardiomyopathy), the heart was in suitable condition to permit removal of the sinoatrial and atrioventricular node areas for histopathologic study of the conduction system.\textsuperscript{8, 9} Serial sections 6 \( \mu \) thick were cut from each conduction system block; every tenth section was stained with hematoxylin and eosin or elastic van Gieson.

**Echocardiographic Studies**

M-mode echocardiograms were obtained in a total of 75 first-degree relatives (ages 4–58 years, mean 28 years) of 15 athletes in whom the necropsy diagnosis was hypertrophic cardiomyopathy or idiopathic concentric (symmetric) left ventricular hypertrophy. Thicknesses of the ventricular septum and posterior left ventricular free wall were measured as previously described.\textsuperscript{10}

**Results**

**Demographic Data and Circumstances of Death**

The 29 athletes ranged in age from 13–30 years (mean 19 years). Twenty-six athletes were male and three were female; 20 were white and nine were black. Body surface areas ranged from 1.7–2.4 \( \text{m}^2 \) (average 2.0 \( \text{m}^2 \)).

At the time of death the athletes had most commonly reached the high school or junior high school level of competition (17 athletes), although eight were participating in intercollegiate sports programs and four were professional athletes. The athletes had participated in a wide variety of sports, but basketball and football were most common (table 1). The duration of participation at competitive levels ranged from 2–18 years.

In 28 of the 29 athletes, death occurred suddenly without warning and was virtually instantaneous; the other athlete survived 12 hours in a terminal state after her collapse. In 22 of the 29 athletes, death occurred during or just after severe exertion. Each of these 22 deaths occurred during an athletic contest or during training. Two of the remaining seven patients died during or just after relatively mild exertion unrelated to participation in sports, and five athletes died during sedentary activities.

**Necropsy Findings: Causes of Sudden Death**

Structural cardiovascular alterations were identified at necropsy in 28 of the 29 athletes (97%), but no significant anatomic abnormality was found in an organ other than the heart (fig. 1). The remaining patient, a 14-year-old high school football player, had a normal heart by both gross and histologic examination. ECGs in five first-degree relatives of this athlete showed no abnormalities (including no evidence of prolonged QT interval syndrome), but a systematic examination of the QT interval syndrome, but a systematic examination of the QT interval syndrome, but a systematic examination of the QT interval syndrome, but a systematic examination of the QT interval syndrome, but a systematic examination of the QT interval syndrome, but a systematic examination of the QT interval syndrome.\textsuperscript{11} For the diagnosis of hypertrophic cardiomyopathy to be made, each of these patients also had to have at least one of the following characteristic features of the disease: (1) asymmetric septal hypertrophy (ventricular septal-to-left ventricular free wall thickness ratio \( \geq 1.3 \); range 1.3–2.2);\textsuperscript{10, 12, 13} (2) marked disorganization of cardiac muscle cells in the ventricular septum,\textsuperscript{7, 14} involving at least 5% of the relevant areas of the tissue section;\textsuperscript{7} and (3) clinical or echocardiographic evidence of hypertrophic cardiomyopathy in one or more closely related family members.\textsuperscript{15, 16} Seven of the 14 athletes with hypertrophic cardiomyopathy had both asymmetric septal hypertrophy and marked ventricular septal disorganization and three had asymmetric septal hypertrophy without disorganization. The other four athletes had concentric (symmetric) left ventricular hypertrophy (septal–free wall ratio \( < 1.3 \)), including two with marked septal disorganization and two without disorganization; each had echocardiographic evidence of hypertrophic cardiomyopathy in relatives.

**Patients with Unequivocal Evidence of Cardiovascular Disease**

Of the 28 athletes with structural cardiovascular alterations, 22 were considered to have unequivocal cardiovascular disease that caused sudden death (fig. 1); the diseases were congenital in 19 of the 22 patients and acquired in the other three.

**Hypertrophic Cardiomyopathy**

The cardiovascular disease most commonly identified was hypertrophic cardiomyopathy, which was present in 14 athletes (figs. 1 and 2; table 2). Each of these patients had a hypertrophied, nondilated left ventricle in the absence of another cardiac or systemic disease that could produce left ventricular hypertrophy (such as valvular, hypertensive or coronary heart disease or infiltrative cardiomyopathy).\textsuperscript{11} For the diagnosis of hypertrophic cardiomyopathy to be made, each of these patients also had to have at least one of the following characteristic features of the disease: (1) asymmetric septal hypertrophy (ventricular septal-to-left ventricular free wall thickness ratio \( \geq 1.3 \); range 1.3–2.2);\textsuperscript{10, 12, 13} (2) marked disorganization of cardiac muscle cells in the ventricular septum,\textsuperscript{7, 14} involving at least 5% of the relevant areas of the tissue section;\textsuperscript{7} and (3) clinical or echocardiographic evidence of hypertrophic cardiomyopathy in one or more closely related family members.\textsuperscript{15, 16} Seven of the 14 athletes with hypertrophic cardiomyopathy had both asymmetric septal hypertrophy and marked ventricular septal disorganization and three had asymmetric septal hypertrophy without disorganization. The other four athletes had concentric (symmetric) left ventricular hypertrophy (septal–free wall ratio \( < 1.3 \)), including two with marked septal disorganization and two without disorganization; each had echocardiographic evidence of hypertrophic cardiomyopathy in relatives.

The heart weights in these 14 patients were all increased over normal\textsuperscript{17} and ranged from 360–630 g.
(mean 475 g). None of the 12 patients in whom blood pressure recordings were available had systemic hypertension. Mild-to-moderate right ventricular dilatation was also present in five patients.

Abnormal intramural coronary arteries (two or more per tissue section) were identified in the ventricular septal myocardium of four athletes. The abnormal arteries had thickened walls (due to intimal proliferation or medial hypertrophy or both) and narrowed lumens.18, 19 One other athlete with hypertrophic cardiomyopathy showed marked narrowing of the arteries to the sinoatrial and atrioventricular nodes due to fibromuscular hyperplasia (figs. 3A–C); the conduction system was otherwise normal, as were the other intramural coronary arteries.

**Anomalous Origin of the Left Coronary Artery**

Four athletes had anomalous origin of the left coronary artery from the right (anterior) sinus of Valsalva,24, 26–28 including one of the patients with hypertrophic cardiomyopathy. Three were 17 years old and the fourth was 22 years old. In these patients the left coronary artery made an acute leftward and posterior angulation from its origin in the right sinus of Valsalva and followed a course through the narrowed area between the aorta and pulmonary artery (fig. 4); the left main coronary artery appeared to be narrowed and obstructed by virtue of its oblique course. In each of the four patients with anomalous origin of the left coronary artery, the distribution of both right and left coronary arteries was normal, and the entire coronary tree was free of significant atherosclerosis.

**Coronary Heart Disease**

Three patients had acquired abnormalities of the coronary arteries. These patients were 24, 26 and 28 years of age. Two of the three patients showed severe atherosclerosis in each of the three major extramural coronary arteries (> 75% cross-sectional area luminal narrowing by atherosclerotic plaque). One of these

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**Figure 1.** Diagram summarizing the cause of death in 29 competitive athletes. CV = cardiovascular; LVH = left ventricular hypertension.

*Includes one patient who also had anomalous origin of the left coronary artery from the anterior sinus of Valsalva. The hearts of four other patients in this subgroup were initially referred for evaluation because of suspected cardiac disease; subsequently, these patients were identified as having been competitive athletes during life.

†Includes one patient who had gross and histologic findings at necropsy consistent with mitral valve prolapse, as well as a family history of that condition. This patient (as well as one other with idiopathic concentric left ventricular hypertrophy) also had marked narrowing of the artery to the atrioventricular node.

**Figure 2.** Heart of a 13-year-old male football and baseball player showing disproportionate thickening of the ventricular septum (VS) with respect to the posterior left ventricular free wall (LV). RV = right ventricular wall.
TABLE 2. Necropsy Findings in 19 Athletes with Hypertrophic Cardiomyopathy or Idiopathic Concentric Left Ventricular Hypertrophy

<table>
<thead>
<tr>
<th>Pt</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Heart weight (g)</th>
<th>Ventricular wall thicknesses</th>
<th>Thickened AML</th>
<th>% VS disorganized*</th>
<th>ASH in relatives*</th>
</tr>
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<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>VS (mm)</td>
<td>LVFW¶ (mm)</td>
<td>VS/LVFW</td>
<td>VS plaque†</td>
<td></td>
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<tr>
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<td>M</td>
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<td>21</td>
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<td>+</td>
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<td>14</td>
<td>M</td>
<td>360</td>
<td>18</td>
<td>12</td>
<td>1.5</td>
<td>0</td>
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<tr>
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<td>17</td>
<td>F</td>
<td>440</td>
<td>15</td>
<td>8</td>
<td>1.9</td>
<td>0</td>
</tr>
<tr>
<td>PB</td>
<td>17</td>
<td>M</td>
<td>530</td>
<td>21</td>
<td>12</td>
<td>1.8</td>
<td>+</td>
</tr>
<tr>
<td>JG</td>
<td>17</td>
<td>M</td>
<td>370</td>
<td>16</td>
<td>10</td>
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<td>M</td>
<td>460</td>
<td>21</td>
<td>14</td>
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<tr>
<td>SW ‡</td>
<td>17</td>
<td>M</td>
<td>490</td>
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<td>630</td>
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<td>+</td>
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<tr>
<td>WD</td>
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<td>+</td>
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<tr>
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<td>KK</td>
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<td>530</td>
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Idiopathic concentric LVH

<table>
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<tr>
<th>Pt</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Heart weight (g)</th>
<th>VS (mm)</th>
<th>LVFW¶ (mm)</th>
<th>VS/LVFW</th>
<th>VS plaque†</th>
<th>Thickened AML</th>
<th>% VS disorganized*</th>
<th>ASH in relatives*</th>
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<td>M</td>
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<td>0</td>
<td>0</td>
<td>0</td>
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<td>KR§</td>
<td>17</td>
<td>M</td>
<td>530</td>
<td>20</td>
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<td>1.0</td>
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<td>0</td>
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<td>M</td>
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<td>1.1</td>
<td>0</td>
<td>0</td>
<td>0</td>
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</table>

*Echocardiographic identification of a septal–free wall thickness ratio of 1.3 in at least one first-degree relative of the athlete.
†Thickening involving the mural endocardium in left ventricular outflow tract, presumably resulting from contact of the free edge of the anterior mitral leaflet with the ventricular septum.
‡Also had an origin of the left coronary artery from the anterior sinus of Valsalva.
§Also had gross and histologic findings consistent with mitral valve prolapse, as well as a family history of that disease.
¶Measured behind the midpoint of the posterior mitral leaflet at the level of the inferior margins of the mitral leaflets.

Abbreviations: AML = anterior mitral leaflet; ASH = asymmetric septal hypertension; LVH = left ventricular hypertrophy; LVFW = left ventricular free wall; VS = ventricular septum; VS/LVFW = ventricular septal-to-left ventricular free wall thickness ratio; + = present; 0 = absent; — = data not available.

patients had total occlusion of the right coronary artery by an antemortem thrombus and hemorrhage into an atherosclerotic plaque. This athlete, a 28-year-old professional football player, had a large myocardial infarct that was about 2 months old in the posterior left ventricular wall at necropsy. The other athlete with severe coronary atherosclerosis also showed extensive atherosclerotic plaque in the ascending aorta. There was no evidence of necrosis or scarring in this heart. The remaining patient had about 50% luminal narrowing of the left anterior descending coronary artery by atherosclerotic plaque and no, or only minimal, atherosclerosis in each of the other two major extramural coronary arteries. There was no myocardial necrosis or scarring. However, the left anterior descending coronary artery was completely occluded by an antemortem thrombus 2 cm from the bifurcation of the left coronary artery. This thrombus was 1 cm in length and adherent to the underlying atherosclerotic plaque.

Aortic Rupture

Two athletes (ages 18 and 21 years) died of aortic rupture, with massive mediastinal hemorrhage. In each, the ascending aorta was markedly dilated (about 10 cm in diameter) (fig. 5A) and had a linear tear in its wall. In one patient the aortic rupture was associated with a dissecting aneurysm of the ascending aorta. Histologic examination of the aortic wall in both athletes showed markedly diminished numbers of elastic fibers in the media, compatible with "cystic medial necrosis" (fig. 5B). One of these athletes had the typical physical stigmata of the Marfan syndrome; the other athlete was tall (81 inches) and relatively thin (210 pounds), but did not have the other physical features of that syndrome.

Patients with Probable Cardiovascular Disease

Six of the 29 athletes had cardiovascular alterations that were considered probable (but not definitive)
FIGURE 3. Small arteries in the conduction system of two athletes who died suddenly. (A–C) The artery to the sinoatrial node of a 24-year-old basketball player with hypertrophic cardiomyopathy (hematoxylin and eosin stain). (D and E) Arteries to the atrioventricular node of a 17-year-old basketball player with idiopathic concentric left ventricular hypertrophy (elastic van Gieson stain). (A) Artery to sinoatrial node has markedly narrowed lumen due to extreme thickening of the arterial wall (magnification × 95). (B) Same artery as in panel A, but at a different level and at lower magnification (×55), showing calcium deposits in adjacent myocardial fibers (outlined by broken line). (C) Higher magnification view (×400) of the calcium-containing fiber shown in panel B. (D) Low-magnification view (×40) of the portion of atrioventricular nodal tissue that penetrates the central fibrous body. Three distinctly abnormal small arteries are evident (arrows). (E) High-magnification view (×130) of one of the vessels shown in panel D, but at a different level. The lumen is markedly narrowed due to thickening of the arterial wall.
evidence of cardiovascular disease. Five had a hypertrophied, nondilated left ventricle unassociated with a cardiac lesion that could produce left ventricular hypertrophy; heart weights were increased over normal\textsuperscript{15} and ranged from 420–530 g. However, each of these five hearts differed from the other hearts in this study considered to represent hypertrophic car-

**Figure 4.** Diagrammatic representation of anomalous origin of the left coronary artery from the right (anterior) sinus of Valsalva. The left coronary artery may have a common (or separate) ostia with the right coronary artery that also arises from the right sinus of Valsalva. Note the acute leftward and posterior course of the left coronary artery between the aorta and pulmonary artery. Normal origins of the left coronary artery from the left sinus of Valsalva and of the right coronary artery from the right sinus of Valsalva are shown below for comparison. \(A\) = anterior coronary cusp; \(L\) = left coronary cusp; \(P\) = posterior (noncoronary) cusp; \(L.A.D.\) = left anterior descending coronary artery; \(L.CIRC.\) = left circumflex coronary artery; \(PUL.\ A.\) = pulmonary artery; \(R.C.A.\) = right coronary artery.

**Figure 5.** (A) Posteroanterior chest radiograph obtained from an 18-year-old male collegiate swimmer with the Marfan syndrome who died 2 months later of a ruptured aorta. Note prominent dilatation of the ascending aorta. (B) Histologic section of ascending aorta from the second patient in this study who died of a ruptured aorta. Note the markedly diminished numbers of elastic fibers (which stain darkly) in the media. Intima is at the top and adventitia is at the bottom. Elastic van Gieson stain; magnification \(\times 80.\)
diomyopathy by virtue of the absence of: (1) asymmetric septal hypertrophy (septal–free wall ratios ranged from 1.0–1.2) (fig. 6); (2) disorganization of cardiac muscle cells in the ventricular septum; and (3) clinical or echocardiographic evidence of hypertrophic cardiomyopathy in closely related family members.

These five patients with idiopathic concentric left ventricular hypertrophy ranged in age from 16–28 years. Two were normotensive, but blood pressure recordings had not been obtained in the other three. In three of these five patients the degree of left ventricular wall thickening was particularly marked (average of ventricular septal and posterior left ventricular free wall thickness ≥ 20 mm); in the other two patients, left ventricular wall thickening was only moderate (thicknesses of 16 mm and 18 mm) (table 2). Mild-to-moderate right ventricular dilatation was present in three of these patients.

Two of the five athletes with idiopathic concentric left ventricular hypertrophy had associated cardiac malformations. In one, a 17-year-old football player with a 530-g heart, the anterior and posterior mitral leaflets had an appearance consistent with relatively mild mitral valve prolapse — thickened, gelatinous and mildly scalloped leaflets with excessive mucoid material, histologically. This patient’s mother and two siblings had clinical or echocardiographic evidence of mitral prolapse. In addition, this athlete had an anatomic alteration of the artery to the atrioventricular node, involving severe fibromuscular hyperplasia of the vessel wall and marked narrowing of the lumen (figs. 3D and 3E). One other athlete with concentric ventricular wall thickening had a similar alteration in the artery to the atrioventricular node. The conducting tissue was otherwise normal in these latter two athletes. None of the athletes who had idiopathic concentric left ventricular hypertrophy, with or without alterations in the artery to the atrioventricular node, had abnormal intramural coronary arteries in other areas of ventricular myocardium.

The remaining athlete with probable evidence of cardiovascular disease was a 17-year-old distance runner whose heart was of normal weight (280 g), without luminal narrowing of the extramural coronary arteries. The left main and left anterior descending coronary arteries were normal. However, only two small posterior descending branches to the posterior left ventricular wall arose from the left circumflex coronary artery. The right coronary artery was short and small in caliber and it gave off only two small branches to the posterior wall of right ventricle and none to the posterior wall of left ventricle (fig. 7). Hence, the coronary arterial distribution to the posterior wall of both ventricles was diminished.

Clinical Findings: Suspicion of Cardiovascular Disease During Life

Of the 29 athletes, 21 had been entirely asymptomatic, even upon retrospective questioning of closely related family members. The remaining eight athletes had transient symptoms that may have been due to cardiac disease, including syncope in three, presyncope in one (a single episode 2 months before
TABLE 3. Findings in Seven Competitive Athletes in Whom Cardiovascular Disease Was Suspected or Identified During Life

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Reason disease first suspected</th>
<th>ECG</th>
<th>Cardiac cath</th>
<th>Clinical diagnosis</th>
<th>Necropsy diagnosis</th>
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</thead>
<tbody>
<tr>
<td>DM</td>
<td>14</td>
<td>M</td>
<td>Syncope</td>
<td>WPW pattern; LAD; T-wave abn. I, aV_{1}, ( V_{r} - V_{4} ); ( \pm ) LVH</td>
<td>Mild bilateral PA branch stenosis</td>
<td>WPW syndrome</td>
<td>Hypertrophic cardiomyopathy</td>
</tr>
<tr>
<td>MM</td>
<td>14</td>
<td>M</td>
<td>Family history</td>
<td>LVH with &quot;strain&quot;; Q waves II, III, aV_{F}, aV_{6}</td>
<td>Rest LVOT grad. = 15 mm Hg; LVEDP = 24 mm Hg</td>
<td>Hypertrophic cardiomyopathy</td>
<td>Hypertrophic cardiomyopathy</td>
</tr>
<tr>
<td>JG</td>
<td>17</td>
<td>M</td>
<td>Heart murmur</td>
<td>Q waves III, aV_{F}, ( V_{r} - V_{4} ); ( \pm ) LVH</td>
<td>None</td>
<td>VSD</td>
<td>Hypertrophic cardiomyopathy</td>
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<tr>
<td>SS</td>
<td>18</td>
<td>M</td>
<td>Exam for Marfan syndrome</td>
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<td>None</td>
<td>Marfan syndrome; AR</td>
<td>Ruptured ascending aorta</td>
</tr>
<tr>
<td>EW</td>
<td>20</td>
<td>M</td>
<td>Heart murmur</td>
<td>Deep, symmetric T-wave inversion II, III, aV_{F}, ( V_{r} - V_{4} ); LVH</td>
<td>None</td>
<td>Normal athlete's heart</td>
<td>Hypertrophic cardiomyopathy</td>
</tr>
<tr>
<td>OB</td>
<td>23</td>
<td>M</td>
<td>VT on routine exercise test</td>
<td>Deep, symmetric T-wave inversion I, II, III, aV_{F}, ( V_{r} - V_{4} ); LVH*</td>
<td>Rest LVOT grad. = 0; LVEDP = 5 mm Hg; LV cavity obliteration (EF = 95%)</td>
<td>Normal athlete's heart</td>
<td>Hypertrophic cardiomyopathy</td>
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<tr>
<td>KK</td>
<td>30</td>
<td>F</td>
<td>Mild fatigue</td>
<td>LVH with &quot;strain&quot;; diffuse T-wave abn.</td>
<td>Rest LVOT grad. = 0; LVEDP = 10 mm Hg</td>
<td>Normal athlete's heart</td>
<td>Hypertrophic cardiomyopathy</td>
</tr>
</tbody>
</table>

*Initially this patient underwent a treadmill exercise ECG as part of the preparticipation examination for his university basketball team. During exercise, ST-segment depression of 2 mm with negative slope and frequent premature ventricular complexes were present. During the second minute of recovery, a 4-second period of ventricular tachycardia occurred (fig. 8). A subsequent 24-hour ambulatory ECG showed only occasional premature ventricular complexes and episodes of junctional rhythm; a second 24-hour ECG recorded during a basketball scrimmage showed no arrhythmia.

Abbreviations: AR = aortic regurgitation; abn. = abnormality; cath = catheterization; EF = ejection fraction; grad. = gradient; LAD = left-axis deviation; LV = left ventricular; LVEDP = left ventricular end-diastolic pressure; LVH = left ventricular hypertrophy; LVOT = left ventricular outflow tract; PA = pulmonary artery; WPW = Wolff-Parkinson-White syndrome; VSD = ventricular septal defect; VT = ventricular tachycardia; \( \pm \) = probable.

death), chest pain in two, periodic mild fatigue in one, and mild fatigue, presyncope and palpitations in one.

In two athletes with transient symptoms, as well as in five other asymptomatic athletes, cardiovascular disease had been suspected by an examining physician during life (table 3). In the latter five athletes, the suspicion of cardiovascular disease initially arose because of a heart murmur in two, a family history of heart disease in one,\(^{8}\) identification of ventricular tachycardia on an exercise ECG performed as a routine part of a preparticipation athletic examination in one (fig. 8B), and during evaluation for the Marfan syndrome in one.

Six of the seven athletes in whom cardiovascular disease was suspected during life had hypertrophic cardiomyopathy confirmed at necropsy. However, in only one of those six athletes with hypertrophic cardiomyopathy was the correct diagnosis made clinically. In the other five athletes, the diagnosis made during life was "normal athlete heart" in three, ventricular septal defect in one and Wolff-Parkinson-White syndrome in one. Each of the latter five athletes with hypertrophic cardiomyopathy had a distinctly abnormal ECG (figs. 8A and 9). Two athletes with abnormal ECGs also underwent cardiac catheterization; neither had a left ventricular outflow gradient under basal conditions. (Provocative interventions were not performed.) These findings led to the erroneous conclusion that hypertrophic cardiomyopathy was absent, even though one of these athletes also had a markedly hyperdynamic left ventricle (fig. 8C). The seventh athlete had been identified as having the Marfan syndrome with aortic root dilatation (fig. 4A) and aortic regurgitation during life; he had been advised not to compete in intercollegiate sports, but died of a ruptured aorta 2 months later during swimming practice.

In the remaining 22 athletes there had been no suspicion of cardiovascular disease during life. However, clinical data in two suggested the presence of coronary heart disease, even though it was not identified until necropsy. One of these two athletes was a 28-year-old professional football player who had an abnormally elevated serum cholesterol (350 mg%; normal < 280 mg%), as well as a family history of premature coronary heart disease and death, identified at a routine preparticipation examination for professional athletes. Subsequently, type II hyperlipoproteinemia was documented in two surviving first-degree relatives. Upon retrospective analysis of the athlete's medical history, it was ap-
parent that he had experienced symptoms compatible with myocardial infarction about 45 days before death (which was documented at necropsy), and frequent episodes of chest pain with exertion thereafter. During life, the acute myocardial infarction and chest pains had not been considered evidence of heart disease, but rather manifestations of a traumatic injury. An ECG recorded before the acute myocardial infarction (the only one obtained from this patient) was normal. A second athlete who died of severe coronary atherosclerosis had an abnormal ECG and elevated serum cholesterol (380 mg%) identified at a preparticipation athletic examination, but no further diagnostic evaluation was undertaken.

Discussion

This study shows that most sudden and unexpected deaths in young, competitive athletes are due to cardiovascular disease. In 28 of the 29 athletes, a structural cardiovascular abnormality was identified at necropsy, and in 22 athletes it was almost certainly responsible for their sudden death. Although the etiology of sudden death in one patient without structural cardiac disease is unknown, it is possible that a primary rhythm disturbance was responsible.

While a wide variety of cardiovascular malformations (primarily congenital) was identified in our 29 athletes, the most frequently encountered disease was hypertrophic cardiomyopathy, a condition known to be responsible for sudden death in young, asymptomatic subjects. Although this disease accounted for almost one-half of the deaths in the present series, our data cannot be used to define precisely the prevalence with which different diseases cause sudden death in young athletes. First, our series is relatively small, and cannot be assumed to be an en-
tirely representative sampling of sudden death in athletes. Second, unavoidable biases, intrinsic to our study design, may have influenced the selection of subjects for this investigation. For example, athletes with unequivocal cardiac disease recognized at necropsy may have been preferentially referred to the NHLBI and the Armed Forces Institute of Pathology. However, 17 of 29 subjects were obtained by follow-up of news media reports describing sudden death in athletes. Hence, most patients in this series were identified initially as competitive athletes who had died suddenly, and not as patients with heart disease who were subsequently proved to have been athletes. Even when this latter segment of our patient population was considered separately, our conclusions were similar to those from the entire study population. For example, eight of the 17 athletes initially identified through the news media had hypertrophic cardiomyopathy, compared with 14 of the 29 athletes in the overall study group.

The fact that we chose to limit our investigation to competitive athletes, who were necessarily relatively...
young, also importantly influenced the relative prevalence of cardiovascular diseases found. In such young athletes, atherosclerotic coronary disease would be expected to be an infrequent cause of death. Conversely, had we studied recreational athletes over the age of 30 years at death, the frequency of coronary heart disease would surely have been much higher.

Certain cardiovascular diseases, such as valvular heart disease, coarctation of the aorta, fixed subvalvular aortic stenosis or other acyanotic or cyanotic heart malformations, did not appear in our series of athletes. Such lesions would likely have been identified earlier in life due to the presence of a loud heart murmur or other obvious physical findings and, hence, before participation in competitive athletics had begun. Other known causes of sudden death, such as prolonged QT-interval syndrome, probably did not appear in our series due to their relatively low prevalence in the population.

Cardiovascular disease was unrecognized during life in most of our athletes, and suspected clinically in only seven of the 29. In only two of these seven athletes was the correct diagnosis established during life: one with obvious stigmata of the Marfan syndrome and the second in whom hypertrophic cardiomyopathy was identified after his brother had died suddenly of that disease. The five other athletes in whom cardiac disease was suspected clinically all died of hypertrophic cardiomyopathy. All five of these athletes had distinctly abnormal ECGs and one had an abnormal left ventricular angiocardiogram. Thus, it is likely that if high-quality echocardiograms had been obtained in these athletes, the correct diagnosis of hypertrophic cardiomyopathy would have been made during life. Each of these five athletes, after examination for possible heart disease, were permitted by their examining physician to continue participation in competitive athletics and each subsequently died on the practice field or during an athletic contest.

The five athletes who had concentric (symmetric) left ventricular hypertrophy without ancillary evidence of a primary, genetically transmitted cardiomyopathy represent a diagnostic dilemma. The significance of the ventricular hypertrophy in these patients is unclear. First, we cannot exclude the possibility that the increased cardiac mass identified at necropsy represented only the "physiologic" hypertrophy that would be expected in many highly conditioned competitive athletes.55-58 Indeed, in two of these athletes, the degree of left ventricular wall thickening was consistent with that usually present in actively competing athletes. However, in the other three athletes with idiopathic concentric hypertrophy, the magnitude of left ventricular wall thickening (≥ 20 mm) clearly exceeded that which would appear to be "physiologic" hypertrophy, even after making reasonable allowances for known discrepancies between ventricular wall thicknesses assessed by echocardiography and at necropsy.59 Second, systemic hypertension could not be definitely excluded as the cause of left ventricular hypertrophy in three patients because blood pressure had not been recorded during life; however, two of these three were less than 20 years old at death, and it is unlikely that hypertension played an etiologic role in their disease process.

In two athletes with idiopathic concentric left ventricular hypertrophy, associated abnormalities (which may have been primary or contributing factors in their sudden deaths) also complicate interpretation of the significance of the cardiac hypertrophy found in this subgroup. One of these athletes had, at necropsy, alterations of the mitral valve leaflets consistent with mitral valve prolapse and a family history of that disease. The abnormality of the mitral valve in this patient was relatively mild and therefore was probably not responsible for significant mitral regurgitation or the degree of left ventricular hypertrophy that was present. In addition, this athlete and one other with idiopathic concentric hypertrophy had severe narrowing of the artery to the atrophicventricular node. Abnormal small intramural arteries in the atrophicventricular node (as well as in the sinoatrial node), morphologically similar to those present in our patients, have been described in young, healthy subjects32,33 or in patients with hypertrophic cardiomyopathy34 who died suddenly, as well as in patients with a variety of other cardiac or systemic diseases.35 Unlike the findings in our athletes, the small-vessel abnormalities described in these reports were usually associated with significant fibrosis of the conducting tissue.

Our findings raise questions regarding the prospective identification of potentially lethal cardiovascular diseases in competitive athletes during life. Screening of athletes by personal and family history, physical examination, ECG and echocardiogram would have identified most of the athletes in our study group. However, routine comprehensive screening of this type would probably be impractical. Although screening by personal history and physical examination alone would identify many of the more obvious cardiovascular abnormalities (such as systemic hypertension or aortic valvular stenosis), it would be relatively insensitive for detecting other diseases. For example, the most frequent cause of death in this series, hypertrophic cardiomyopathy, is usually not associated with a loud heart murmur,36,37 or even always with an abnormal ECG38 when present in its nonobstructive form. Definitive identification of hypertrophic cardiomyopathy without obstruction could be made noninvasively in athletes only by echocardiography. Similarly, echocardiography could identify many of the patients with aortic root dilatation who may be at high risk of aortic rupture.

The data in this study do not provide definitive information regarding the feasibility of screening programs for detecting cardiovascular disease in competitive athletes. However, the basic knowledge and awareness of the diseases that may cause sudden death in athletes help to create the groundwork necessary for the initiation of studies that will provide such answers.

References

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