Following the Trail of Broken Hearts

A congenital cardiovascular abnormality has become a leading killer of young athletes in the U.S. So why isn’t more being done to save those who have it?

BY DAVID EPSHTEIN | Photograph by Prestige Portraits

THE KID’S playing with me, William Batts figured. One second 17-year-old DeCarlo Polk had gone up confidently for a jumper, his chiseled 5’ 9”, 237-pound frame rising just beyond the free throw line in their game of one-on-one. The next, he had tumbled to the Nashville blacktop like a piece of laundry at the end of a dryer’s cycle. Now, on the afternoon of June 27, DeCarlo lay flat on his back in the simmering heat. Real funny, the 37-year-old Batts thought. But then he took a step closer and saw that DeCarlo’s pupils had rolled back into his skull, and he knew it was no joke.

Somewhere mid-jump the electrical signals that cued DeCarlo’s heart to pump had misfired terribly. Rather than flexing and contracting rhythmically, his heart trembled like jelly in a shaken jar. His left ventricle, the chamber that takes oxygenated blood from the lungs and sends it through the body, stopped working, causing a circulatory traffic jam. As blood piled up in the lungs’ capillaries—vessels so small that red blood cells have to pass through them single file—water in the bloodstream breached the capillary walls and settled into the air sacs of his lungs, spaces normally reserved for oxygen. DeCarlo had stopped breathing and started drowning. Batts called his boss—DeCarlo’s uncle, Jimmy Boulton—as his pulse slowed to three then down, and Boulton came running with Rick Jones, a coworker. Jones became DeCarlo’s lungs, blowing oxygen-rich air into his mouth.

MIXED SIGNALS An enlarged heart often indicates a superbly conditioned athlete, but for 17-year-old DeCarlo it was a sign of his hypertrophic cardiomyopathy (HCM).
Anatomy of a Failing Heart

In the HCM heart the septum, the muscle wall that separates the ventricles, grows abnormally thick and invades the left ventricular cavity, where oxygen-filled blood collects before being pumped through the body. Normally thinner than 1.2 centimeters, a septum thicker than 1.6 cm is nearly always indicative of HCM. When the septum is between 1.3 and 1.5 cm, however, doctors find it difficult to determine whether the heart has been enlarged from exercise or from HCM.

Then he became his heart, pushing down on his chest to force the oxygenated blood through the lungs and into the rest of his body. But CPR could only buy time.

Don't let this happen, Boulton prayed, not now. On June 29, DeCarlo had graduated from Hillside High in Durham, N.C., where he had been an honor student, homecoming king and star defensive lineman. Football scholarship to Division II St. Augustine's College in nearby Raleigh in hand, he was baptized two days later, then jumped in Boulton's track for his annual summer visit to Nashville, his hair still parted from the baptismal dip. Within a week of his arrival DeCarlo became the proud cousin of Jimmy's third child, James Kerrigan Boulton Jr.

It took at least 10 minutes for the rescue workers to arrive; already DeCarlo's oxygen-deprived brain cells were dying in droves. The paramedics applied defibrillators to DeCarlo's chest, trying to fix the deadly rhythm that had begun in his left ventricle. Throughout his heart electrical signals were ricocheting like pinballs, spurring spastic muscle twitching. With each jolt from the paddles DeCarlo's powerful body would lift and then flop limply to the ground. His 17-year-old heart was broken.

DeCARLO was one of an estimated 500,000 Americans with hypertrophic cardiomyopathy (HCM), a genetic disease that causes the walls of the left ventricle to enlarge, usually during adolescence; the thickened ventricle does not relax fully, inhibiting the flow of blood into the heart. While many will never exhibit a symptom, some 6,000 will die from HCM each year, more than from asthma and Hodgkin's disease combined. In those HCM victims, the left ventricle's muscle cells, rather than stacking up flesh like bricks in a chimney, are arranged at odd angles, all askew, as if the bricks had been tossed into a Dungaree. With the cells in such disarray, an electrical signal crossing them is liable to travel in an erratic path and fatally misfire.

"Hypertrophic cardiomyopathy is the most common cause of sudden death in young athletes," says Dr. Barry Maron, director of the Hypertrophic Cardiomyopathy Center at the Minneapolis Heart Institute Foundation and one of the top experts on HCM. "And it's the most common cause of [natural] sudden death in young people in the general public. But it's still a little-known disease.

For those who suffer from some of the nation's most common health problems, such as diabetes, high cholesterol and coronary artery disease, exercise is a potent remedy. But for those with HCM it is the most active individuals—the young (usually teenage) para-threads of fitness—who are at higher risk of sudden death because of their athletic pursuits. At least every two weeks an athlete with HCM will die during or immediately after exertion, when his or her abnormally thick heart, triggered by strenuous exercise, will start beating in a lethal cadence.

Some of the victims are famous, such as San Francisco 49ers offensive lineman Thomas Herrick, 23, who collapsed in the locker room minutes after a preseason game in August 2005, and Jason Collier, 28, the Atlanta Hawks center who died in the middle of the night two months later. But the majority are less well-known, their lives just beginning, such as 16-year-old Adam Litten, who went lifelessly sliding across a hockey rink after he collapsed during an October practice in St. Peters, Mo., and DeCarlo Polk.

And Kevin Richards. During my senior year at Evanston (Ill.) Township High, Kevin, a precocious freshman, became my friend and training partner in track. After I went to Columbia in August 1998, Kevin helped Evanston win its first 4 × 800-meter state title the following spring. By his junior year Kevin, the son of Jamaican immigrants, was on his way to becoming the first member of his family to attend college, most likely on a track scholarship. Indiana was high on his list.

On the afternoon of Feb. 12, 2000, Kevin was locked in a tight race in the indoor mile. The bell hollowly signaled the final lap as Kevin kicked to the shoulder of Dan Gla, a top Illinois distance runner from Amos Alonso Stagg High in Palos Hills. During that lap Kevin's heart struggled to function. Still he kept coming, finishing second by a meter. After crossing the finish line Kevin...
DeCarlo's heart—perhaps enlarged from working out, and certainly enlarged with HCM—weighed 600 grams. (Ryan Shag, 28, who died 55 miles into the Olympic marathon trials in New York City last month, and Toronto Blue Jays pitcher Joe Kennedy, 28, who died at his in-laws' home three weeks later, also reportedly had enlarged hearts. Their autopsies are pending.)

If, during high school, DeCarlo had been given an electrocardiogram, or ECG, a $50 test that takes a few minutes and

**A CRUEL TOLL** Herron (left) dropped dead after a preseason game, while Collier died at home in the middle of the night.

AT LEAST EVERY TWO WEEKS IN THE U.S., A HIGH SCHOOL, COLLEGE OR PRO ATHLETE WITH HCM WILL DIE.

gorous ballplayer. "Right now we really don't know most of the time who is at risk of sudden death," says Dr. Paul D. Thompson, a cardiologist at Hartford Hospital. The first diagnosis is often made by a medical examiner, if it is made even then. With few live patients to examine, clinicians have had trouble getting a fix on the disease since British pathologist Donald Tsarey first compared HCM to a tumor of the heart in 1959. The disease would acquire more than 30 different names, from apical hypertrophy to subvalvular aortic stenosis, until becoming widely known as hypertrophic cardiomyopathy within the last decade.

While an enlarged heart might serve as a conspicuous sign of trouble, it is also characteristic of a highly conditioned athlete. It is no surprise to anyone who saw DeCarlo race sideline to sideline for a tack that he had, literally, a lot of heart. A normal, fully mature human heart weighs about 300 grams, or two thirds of a pound.

records the electrical signals in the heart, a cardiologist might have noticed a suspicious pattern indicative of an enlarged ventricle. The doctor might have followed up with an echocardiogram—or echo, for short—an ultrasound that gives a real-time picture of the heart and costs $1,000 to $2,000. To a cardiologist who had seen HCM before, an echo could determine whether the heart was merely enlarged by exercise (in which case both the muscle of the left ventricle and the chamber it surrounds would expand) or whether it was afflicted by HCM (in which case the muscle walls would grow but the chamber would not). The doctor might have recommended that DeCarlo trade his cleats for golf clubs, tools of a less vigorous sport that he loved. He could have lived a longer life with a surgically implantable cardiac defibrillator (ICD) standing sentinel inside his chest. The titanium-encased, computerized device is about the

size of a matchbox and is programmed to give the bearer's heart a 750-volt shock when it detects an abnormal rhythm.

DeCarlo would have undergone at least an ECG had he grown up in Italy, where all competitive athletes, from grade-schoolers to pros, must, by law, have their hearts undergo government-subsidized screening. When an American child's heart fails, crestfallen parents often, and understandably, speak out in favor of a nationwide screening program that includes ECGs. But Italy has the richest and most sporting of nations?

For one, the U.S. has 37% fewer doctors per capita than Italy, a nation that opened the doors wide to its medical schools after World War II. Some Italian doctors have spent almost their entire careers screening athletes. With an American citizenry that is far more geographically and genetically diverse, and that has more than twice as many high school athletes (7.3 million) as Italy has high-school-age people, marshaling enough experts to do quality, uniform screening is impractical, with the costs prohibitive. "The U.S. health care system does not have the mechanisms to pay for an ECG for every athlete," says Lisa Salberg, who has HCM and founded the Hypertrophic Cardiomyopathy Association. "Nor do we have enough trained professionals to evaluate the results. There would be a lot of athletes placed on alert for no reason, and a lot of missed diagnoses."

And yet, immediate steps can be taken given the hereditary nature of HCM. "Asking whether there is anyone in the family who has died before the age of 50 of cardiovascular causes is incredibly valuable," says Dr. David Glover, a Warrensburg, Mo., physician and expert in prosports participation screening, "and that doesn't cost a nickel."

Although Glover has seen improvement in preparticipation screening questions, he has also documented a dangerous trend. In 1997 there were 11 states that allowed chiropractors or other nonphysi-
cians to perform the exams that deem a high school athlete fit to play. By 2000 that
total increased to 18 states that sanctioned.
Glover says, "practitioners with little or no
cardiovascular training" to conduct athletic
screening. "You need somebody that listens
to hearts every day," Glover says. "A trained
examiner knows if a heart murmur is very
loud, or if it occurs during the relaxa-
tion phase of the heart, or if it gets louder
when you stand up, it raises suspicion that
a person could have HCM."

THERE IS a sleek, steel-and-glass
building about a mile away from
Boston's Fenway Park where the
men's and women's rest rooms are labeled
with diagrams of chromosomes. On its
second floor is Harvard's Seidman Lab,
rung by the husband-and-wife team of Jon-

SHOULD ALL PATIENTS KNOW
EVERYTHING ABOUT THEIR GENOME?
EXPERTS AREN'T SURE.

than and Christine Seidman. "Looking at
a patient's genome can tell us that somebody
has HCM or is going to develop HCM as
they get older," says Christine. The lab
is putting mice genetically bred to have
HCM through the first HCM drug trial,
with the hope that, Christine says, "in a
decade or two, the [manifestation of the]
disease may be preventable."
Perhaps those future patients will have
had HCM diagnosed with absolute certainty
through their genes at the nearby Harvard-
Partners Center for Genetics and Genom-
ics, where each week researchers identify
a new HCM mutation, of which more than
800 are currently known. For $3,000, the
center can pop the blood of a living person
(or a deceased one, if a medical examiner is
sufficiently eager to extract a sample) in
what looks like an over-sized gray microwave
and find out if that individual got HCM from any
of the currently known mutations.
And once a mutation is identified in one
member of a family, other members can be
tested for the same mutation—at $250 apiece.
That's what Jimmy Kogut's three younger
siblings are doing. A 21-year-old junior at
Pittsburgh, Jimmy spent his leisure time

DONT TEST, DONT TELL Curve refused to
undergo screening that might have revealed
HCM, so the Bulls traded him to New York.

BUT WILL people feel safe to seek
that knowledge? In September
2006, six months after Chicago
Bulls center Eddy Curry felt his heart
skipping beats, the team added a genetic
testing clause to the one-year, $5 million
contract offer it had on the table. If the
tests showed that Curry had HCM, the
team would not let him play, but it still
promised to pay him $400,000 a year for
the next 50 years. Fearing that the public
disclosure of the results might jeopardize
his career, Curry refused, and the
Bulls traded him to the New York Knicks.
"As far as DNA testing, we're just at
the beginning of that universe," Alan
Milestone, Curry's attorney, told the Associ-
ated Press. "Pretty soon, though, we'll
know whether someone is predisposed
to cancer, alcoholism, obesity, baldness
and who knows what else — Hand that
information to an employer and imagine
the implications."
The Genetic Information-Nondiscrimi-

Introuducing the new Everio GZ-HD3 camcorder, the perfect holiday gift for the
highly defined sports enthusiasts on your list. The GZ-HD3 records in stunning
1440x1080 HD with no tapes or disks to load, and features 3 CCD technology
for bright, vibrant colors. With a 60GB hard drive, you get up to 7 hours of recording
time with an HD lens developed by the camera specialist, KONICA MINOLTA. It's
HD priced affordably. Making it the camcorder choice you don't want to pass on. 
against a person based on the content of their genetic code. In effect, it would give people like Curry the freedom to submit to a potentially lifesaving test without fear of reprisal. Last April the House of Representatives voted 420-3 to pass the bill, and President George W. Bush has already said that he will sign it should it clear the Senate. But Senator Tim Coburn (R., Okla.), a physician who voted for a similar version of the bill that passed the Senate 98-0 in 2005 (but never made it to a vote in the House), has placed a “hold” on the latest GINA, forestalling a vote. Coburn initially argued for specific language ensuring the rights of the fetus. The language of the bill was altered to that end, but his hold persists.

Coburn says that he supports a genetic nondiscrimination law, but that the current version of GINA does not provide enough protection for employers. “What if an employee files a form to take family leave from work and they write on the form that they have to take care of their mother who has breast cancer?” he says. “Because breast cancer can be hereditary, the employer may have gathered genetic information accidentally, and they are vulnerable in a lawsuit. We need protection for [employers and insurance companies] who don’t mean to discriminate, but have accidentally collected information.”

As far as Christine Seidman is concerned, the passage of GINA, which was first introduced in 2002, is overdue. “Americans have to support this,” she says. “I have Alzheimer’s and breast cancer in my family. You may have schizophrenia. We all have something. I believe that families have a right to know everything they can.”

But should all patients know everything about their genome? Even experts aren’t certain. “As some kids, and they don’t have a family history of death and they don’t have symptoms or a very thick heart, and I don’t think a lot of them are at great risk,” says Thompson, a former marathoner who competed in the 1972 Olympic trials. “I usually say to them, ‘I don’t think you’re at great risk. I wish I have to sleep at night, and I can’t take a chance with you, so I’m prohibiting you.’”

For some acne-stained 17-year-old who’s accepted at that high school because he’s a good linebacker, to tell him that’s gone is a load.”

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Sudden Deaths

HCM may be the leading killer of young athletes, but other cardiovascular conditions have also claimed prominent lives.

**FLO HYMAN**, died in 1989 at age 31 (Marfan syndrome)

Most often seen in tall, lanky individuals such as the 6'6" Hyman, a volleyball star who made the 1984 Olympics, Marfan syndrome is a congenital disease that weakens the body's connective tissues. Affecting one in 5,000 people, it is often characterized by cardiovascular abnormalities—such as Hyman's weakened aorta, which ruptured during a Japanese-league match.

**PETE MARAVICH**, died in 1980 at age 40 (coronary artery anomaly)

Most people have two arteries that bring blood to the heart, but Maravich's autopsy showed that he was born with only one. While Pistol Pete, the 1976-77 NBA scoring leader, was playing a pickup game, he suffered a heart attack.

**HANK GATHERS**, died in 1990 at age 23 (heart arrhythmia, cause unknown)

After passing out during a December '89 game, the NCAA's reigning scoring and rebounding champion was found to have occasional ventricular tachycardia, a potentially lethal heart rhythm that starts in the ventricles. Doctors prescribed a beta-blocker to control the arrhythmia, but the dosage was reduced after Gathers felt sluggish. He dropped dead during a game the following March.

**REGGIE LEWIS**, died in 1993 at age 27 (myocarditis)

He collapsed during an April '93 playoff game and died while shooting baskets three months later of an inflammation of the heart typically caused by a virus that damages the organ's muscle cells.

**SERGEI GRINKOV**, died in 1995 at age 28 (coronary atherosclerosis and high blood pressure)

Atherosclerosis refers to the narrowing and hardening of the arteries. The Russian Olympian's figure skating champion in 1989 and '91 had a heart attack while practicing with Ekaterina Gordeeva, his wife and skating partner, in Lake Placid, N.Y. An autopsy found that two of Grinkov's coronary arteries were almost totally blocked.

**DARRYL KILE**, died in 2002 at age 33 (coronary atherosclerosis)

The St. Louis Cardinals' pitcher was found dead in a Chicago hotel room, atherosclerosis having impeded the blood supply to the heart.

**GEORGE BOJARDI**, died in 2004 at age 22 (possible concomito cordis)

Translation from Latin as a "commotion of the heart," concomito cordis is a sudden, disordered heart rhythm caused by a blunt impact; Bojardi, a senior lacrosse player at Cornell, was struck in the chest by a ball during a game.

**CHAD SCHIEBER**, died in 2007 at age 39 (mitral valve prolapses)

A condition in which one of the heart valves does not close correctly; mitral valve prolapses affects more than 2% of adults but rarely leads to complications. The backflow of blood can cause the heart to work too hard; it’s not known if that's what killed Schieber, who died during the Oct. 7 Chicago Marathon.
Since Knapp's ordeal, the use of implanted defibrillators has become more common in the population at large, and doctors have permitted some athletes with ICDs to compete. In January 2006, Washington guard Kayla Burt, who has Long Q-T syndrome, a generic heart disease that, like HCM, is treated with an ICD, was sitting on the bench in a game against UCLA when her device fired. "She thought someone came up behind and punched her in the head," recalls team trainer Jenn Raeliff. The shock "usually gets a 10 out of 10 on the pain scale from patients," says Dr. Mark Estes, an electrophysiologist and director of the New England Cardiac Arrhythmia Center. Burt, who had nearly died on New Year's Eve, 2002, when she went into cardiac arrest, gave up playing for good that night. "The device worked, which is good, but not exposing yourself to potentially to find. There is still much that is not known about the progression of the disease—when and how the cellular disarray develops, for example—largely because a live heart can't be put under a microscope. There is still no explanation for why Eileen Kogut's two brothers died but she, with the same HCM mutation, leads a normal life. Says Suhovich, "Somewhere between screening every kid in America for a couple of billion dollars and not doing anything lies the right answer."

 ScarJo Polk's mom, Tommie (Lady) Polk, knows a lot about HCM now, but she does not have any more children to lose. DeCarlo was her baby and she clings to his memory as if trying to keep a balloon from drifting into the sky. There are moments around every corner in her yellow clapboard house in Durham: in the

**IF ONLY SHE'D KNOWN, LADY POLK WOULD HAVE PAID FOR THE TESTS TO MAKE SURE DECARLO WAS SAFE.**

When he arrived in Evanston, however, the school's doctors, concerned that he might have a fatal heart condition, declared him ineligible. Knapp's left ventricle was slightly enlarged, and cardiologists were divided on whether he had the disease. Knapp himself doubted that he had HCM and sued the university for the right to play. Keeping him off the court, Knapp argued in his lawsuit, violated the 1973 Rehabilitation Act, which protects "otherwise qualified individuals" from discrimination based on a disability.

Knapp won in federal court, but Northwestern got the ruling overturned by the U.S. Court of Appeals for the Seventh Circuit. The court acknowledged that Knapp could play college ball, but that Northwestern also had the right to heed its doctors and to prevent him from potentially risking his own death at its facilities. Though the university honored his scholarship, Knapp, who sat out for two seasons during the litigation, transferred to Northwestern Illinois, then to Division II Ashland (Ohio) University. He still pondered what might have been: "I've played against guys who went to the NBA, like Brian Cardinal and Stephan Marbury," Knapp says, "and I was at that level."