

# Sudden Cardiac Death in Athletes

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## Clinical Points

- SCD in athletes is rare but significant as it can result in the unexpected death of a young, apparently healthy person
- The trigger for SCD is exercise in the presence of certain cardiovascular diseases, which can lead to fatal arrhythmias of the heart during physical exertion
- The most common causes for SCD in a young person:
  - a. Hypertrophic cardiomyopathy
  - b. Commotio cordis - unusual cause, but still second most common (especially in children due to their fragile thoracic skeletons)
  - c. Congenital abnormalities of the coronary arteries
- Atherosclerotic coronary artery disease is the most common cause of SCD in athletes over 35. The incidence is 10-20 times more than in a younger person
- A selective screening strategy for SCD is the most favoured approach. Identifying those at risk via history and physical exam before undergoing further investigations is crucial

## ABSTRACT

With the death of several well known athletes in the last thirty years, sudden cardiac death (SCD) has become of interest among many physicians. A series of studies conducted by American cardiologist and researcher, Dr. Barry J. Maron, have identified the major aetiologies in young athletes. In Ireland, Dr. Fionnuala Quigley, GP and lecturer in sports medicine, conducted a study that identified the main causes of SCD occurring in sport from 1987 to 1997. This data emphasized the causes of SCD in athletes over 35. This paper gives an overview of the main conditions that can lead to SCD in athletes, and discusses the methods and challenges of implementing a screening program.

## INTRODUCTION

In 490 B.C., the Persian army attacked Greece in the Battle of Marathon. Despite being outnumbered three to one, the Greek army managed to encircle the Persians and emerge victorious. According to legend, a long-distance messenger, known as Phidippides ran 26 miles from Marathon to Athens, delivering the news of the Greek triumph. Upon delivering his message he dropped to the ground and died<sup>1</sup>. This story not only gave rise to an Olympic running event known today as the marathon but it was also the first recorded incidence of SCD in an athlete<sup>2</sup>. There have been very few documented cases of SCD occurring in sports since.

At the end of the twentieth century, the unexpected death of several high profile athletes led to an increased awareness of SCD. In 1986, 32 year old U.S. Olympic volleyball player Flo Hyman died on the sidelines after being substituted out during a game in Japan. In 1990, 23 year old U.S. college basketball player, Hank Gathers, collapsed on the court and died<sup>3</sup>. In 1994, 28 year old Russian Olympic figure skater Sergei Grinkov had a massive heart attack and died while practicing for an upcoming performance<sup>4</sup>. In 1999, 25 year old Gaelic footballer, Gerald Gallagher died of a heart attack during a

match<sup>5</sup>. More recently, in 2004, 18 year old John McCall from Northern Ireland suddenly collapsed and died during a rugby match against New Zealand.

### Aetiology in Younger Athletes

A 2003 study by Maron<sup>6</sup> compiled several causes for SCD in 387 American athletes aged 12-35 (Table 1). Male athletes were found to be three times more affected than female athletes, with the frequency of SCD increasing with age. The trigger for cardiac arrest was noted to be participation in sport in the presence of cardiovascular diseases, which can lead to fatal ventricular arrhythmias during physical exertion. Thus, most athletes who died suddenly were found to have cardiomyopathies, congenital coronary abnormalities, or atherosclerotic coronary artery disease (Table 1).

Hypertrophic cardiomyopathy is the most common cause of SCD in young athletes<sup>6</sup>. Half of those affected will describe symptoms of chest pain, dizziness, fainting, and abnormal breathlessness, particularly with exercise. Others may be asymptomatic, with SCD being the first sign of disease. Twenty percent of cases of hypertrophic cardiomyopathy are inherited, with the disease being passed on in an autosomal dominant fashion<sup>2</sup>.

**Table 1** - Adapted from B.J. Maron. Sudden death in young athletes. *N Engl J Med.* 2003 (6)

Cardiovascular Abnormality	%of Cases
Hypertrophic cardiomyopathy	26.4
Commotio cordis	19.9
Coronary artery anomalies	13.7
Left ventricular hypertrophy of indeterminate causation	7.5
Myocarditis	5.2
Ruptured aortic aneurysm (Marfan syndrome)	3.1
Arrhythmogenic right ventricular cardiomyopathy	2.8
Tunneled (bridged) coronary artery	2.8
Aortic valve stenosis	2.6
Atherosclerotic coronary artery disease	2.6
Dilated cardiomyopathy	2.3
Myxomatous mitral valve degeneration	2.3
Asthma (or other pulmonary condition)	2.1
Heat stroke	1.6
Drugabuse	1.0
Other cardiovascular cause	1.0
Long QT syndrome	0.8
Cardiac sarcoidosis	0.8
Trauma causing structural cardiac injury	0.8
Ruptured cerebral artery	0.8

Presentation is usually during the second decade of life and is characterized by dramatic ventricular thickening which particularly involves the ventricular septum. This excessive thickening of the heart muscle can lead to ventricular fibrillation, and death.

Commotio cordis, literally meaning “concussion of the heart,” involves a blunt, non-penetrating impact to the precordial region of the chest<sup>7</sup>. The kinetic energy of the impact is transmitted to the heart muscle and causes a fatal ventricular arrhythmia. For the impact to be fatal, it is thought that it must occur during the vulnerable phase of the cardiac cycle when the heart is relaxed. Commotio cordis is an unusual cause of SCD, but still ranks as the second most common aetiology<sup>6</sup>. Sports such as rugby, football, baseball, or martial arts, which may involve such trauma to the chest, are associated with an increased risk of commotio cordis<sup>8</sup>. Children are especially vulnerable due to their fragile thoracic skeletons. Pre-existing structural or electrical abnormalities of the heart have been speculated to render the individual more susceptible, yet there is no hard evidence to support this theory.

Congenital coronary artery abnormality is another common cause of sudden death in young athletes<sup>6</sup>. Such anomalies include the abnormal origin of the coronary

arteries from the aorta, incomplete development of the arteries, coronary arteries buried deep within the heart muscle, and an abnormal origin of the left coronary artery from the pulmonary artery instead of the aorta<sup>9</sup>. The specific mechanism underlying death in these conditions is assumed to be insufficient blood flow to the heart during increased physical activity.

### Aetiology in Older Athletes

A retrospective study was conducted by Quigley in 2000, which covered a 10 year period from 1987 to 1997, to observe the incidence of SCD associated with sports in the Republic of Ireland (Table 2)<sup>10</sup>. Fifty-one cases of SCD were reported by coroners. Ages of the individuals ranged from 15 to 80 years with a median age of 48 years. Males represented 50 of the cases. The cause of SCD in 42 cases was atherosclerotic coronary artery disease. Thirty-nine of these deaths occurred in men over the age of 35. Of interest, SCD in young athletes was rare, with only one case of hypertrophic cardiomyopathy; one case of a coronary artery abnormality; and no cases of commotio cordis.

**Table 2** - Adapted from Quigley F. A survey of the causes of sudden death in sport in the Republic of Ireland. *The Brit J of Sports Medicine.* 2000 (10)

Type of sport associated with SCD	# of Sudden Deaths
Golf	16
Gaelic football	11
Jogging	5
Tennis	5
Marathon	2
Soccer	2
Swimming	2
Badminton	1
Table tennis	1
Squash	1
Rugby	1
Bowls	1
Horseriding	1
10 km Run	1
Pitch and putt	1

Atherosclerotic coronary artery disease is rare in young athletes, as the genetic factors that predispose one to premature disease are uncommon<sup>11</sup>. However, there is an increase occurrence of atherosclerosis in males over 35. Various risk factors can lead to lipid deposition and hardening of the arteries characteristic of the disease (Table 3). In general, moderate exercise is recommended to prevent early death from atherosclerosis<sup>12</sup>. As the study by Quigley reveals, there are a few instances in

which physical exertion in the presence of atherosclerotic disease can lead to sudden death. The mechanism involves fatal ventricular fibrillation provoked by decreased blood flow to the heart due to atherosclerotic coronary arteries and exacerbated by exercise<sup>13</sup>. SCD due to atherosclerosis is anywhere from 10 to 20 times more common than SCD due to hypertrophic cardiomyopathy, coronary abnormalities, or commotio cordis<sup>6</sup>. Yet, the incidence of SCD in athletes with atherosclerosis is extremely low when compared to death from coronary artery disease in the general population. In Ireland, there were 42 SCDs in athletes from atherosclerosis over a 10 year period from 1987 to 1997 compared to 8,000 annual deaths from coronary artery disease during the same time period<sup>14</sup>.

**Table 3** - Adapted from Burke AP et al. Coronary risk factors and plaque morphology in men with coronary disease who died suddenly. *N Engl J Med.* 1997 (13)

Major Risk Factors	Minor Risk Factors
Hyperlipidaemia	Male gender
Hypertension	Obesity
Smoking	Sedentary lifestyle
Diabetes	Type A personality (stress) <ul style="list-style-type: none"> <li>• Impatient</li> <li>• Excessively time-conscious</li> <li>• Insecure</li> <li>• Hostile/Aggressive</li> <li>• Highly competitive</li> </ul>
	Elevated homocysteine
	Oral contraceptive use
	Increasing age
	Genetic factors

In the past, there was much debate about implementing athletic screening programs for SCD. It was felt that with the low prevalence of cardiac anomalies, a large number of athletes would have to be screened to identify the single individual who would die suddenly<sup>15</sup>. The likelihood of 'false positives', where tests might incorrectly indicate that someone was at risk, and 'false negatives', where a defect might not be picked up, made screening programs expensive, inefficient and potentially counter-productive<sup>16</sup>. A previous study by Maron, in 2002, estimated the cost of echocardiogram screening of all American athletes for hypertrophic cardiomyopathy to be \$2000 per individual<sup>7</sup>. In addition to the high cost, false positive test results could have an emotional toll on athletes and may cause them to unnecessarily give up sports altogether.

Today most cardiologists are in favour of a selective screening strategy. The selective process involves taking the patient history in an attempt to single out those who

may be at risk for SCD<sup>18</sup>. Specific questions focus on the family history with regards to SCD, premature death from heart disease, cardiomyopathies, or congenital heart abnormalities. Any past cardiac symptoms such as shortness of breath, palpitations, chest pain, or fatigue would also be explored. A physical exam would then look for abnormalities in blood pressure or heart murmurs. Individuals who are thought to be at risk would then undergo further testing with an echocardiogram and electrocardiogram (ECG).

The success in screening athletes for SCD has been seen in Italy, where all athletes ages 12 to 35 have been screened on an annual basis since 1971<sup>19</sup>. The screening program includes a history and physical examination, exercise and pulmonary function testing, and an ECG. Italy's National Health Service funds the program and has seen a 90% drop in SCD from 3.6/100,000 to 0.4/100,000 in Italy's Veneto region<sup>20</sup>. These favourable results have inspired several physicians and athletic trainers in the United States and Europe to initiate their own screen programs.

## CONCLUSION

SCD in athletes is rare but has seen an increase in awareness due to the high profile deaths of several young athletes in the last three decades. Hypertrophic cardiomyopathy, commotio cordis, and coronary artery anomalies are the main causes of SCD in younger athletes, while atherosclerotic coronary artery disease is the main aetiology affecting athletes over the age of 35. The implementation of screening programs for SCD can help identify both young and old athletes who may be at risk. While mass screening for SCD is not appropriate for most countries, a selective screening approach is likely more economical and efficient. As athletic screening programs for SCD start to become more widespread it is hoped that future tragedies may one day be prevented.

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