# Case Report

# Sudden Cardiac Death in Young Adults

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## Abstract

Sudden cardiac death is defined as death from unexpected circulatory arrest resulting from cardiac arrhythmia which occurs within one hour of the onset of symptoms. Hypertrophic cardiomyopathy (HCM) is the leading cause of sudden cardiac death in young athletes in the United States and the most common genetic cardiovascular disorder. HCM is characterized by a heavy muscular hyper contracting heart and is a diastolic disorder of heart with asymmetric hypertrophy of ventricular septum. HCM is mostly asymptomatic until sudden cardiac death occurs. Sudden death due to cardiac failure is the most common cause of death and particularly likely in young males with familial HCM or with a family history of sudden death. Major risk factors for sudden death in individuals with Hypertrophic cardiomyopathy include prior history of cardiac arrest, ventricular fibrillation, spontaneous sustained ventricular tachycardia, family history of premature sudden death, unexplained syncope, left ventricular thickness more than or equal to 30mm, abnormal exercise blood pressure and non-sustained ventricular tachycardia. We are hereby reporting few rare cases of sudden cardiac deaths, the bodies of which were brought to the mortuary of S.C.B Medical College, Cuttack for autopsy.

**Key Words:** Hypertrophy, Cardiomyopathy, Sudden Death, Young Male

## Introduction:

Many a times Forensic experts encounter cases of deaths which are sudden, unexpected, clinically unexplained and suspicious in which there is usually no unnatural element. According to WHO, sudden death is defined as those deaths which occur within 24 hours from the onset of symptoms. [1]

Diseases of cardiovascular system account for about 45-50% of all sudden deaths. [2] Hypertrophic Cardiomyopathy (HCM) is one of the most common causes of sudden and unexplained death in young athletes. [3]

Hypertrophic cardiomyopathy is defined by the presence of unexplained myocardial hypertrophy typically involving the interventricular septum more than the posterior or free wall of the left ventricle. [4] The classic pattern of HCM is disproportionate thickening of the ventricular septum as compared with the free wall of the left ventricle for which it is also known as asymmetric septal hypertrophy. [3]

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Marked hypertrophy can involve the entire septum but it is usually most prominent in the sub-aortic region. [3]

In this disorder the ventricular cavity is compressed in to a banana like configuration as elicited on cross section. [3] Variable amount of scarring may be seen in the septum suggesting previous ischaemic episodes. [5] The exact cause of HCM is not yet known. [6]

However, in approximately half of cases the disease is familial and the pattern of transmission is autosomal dominant. [3] Rest of the cases is sporadic in nature. [3] About 60% of the HCM is caused by mutation of beta-myosin heavy chain, cardiac myosin binding protein C, cardiac troponin-T and I. [7] Majority of the patients with HCM have at least one other affected first degree relative. [6]

Most of the patients of hypertrophic cardiomyopathy have few or no symptoms but in hospital based populations dyspnea, chest pain, unexplained syncope, palpitation and fatigue are commonly seen. [7] The most important complication of HCM is sudden death. [7]

Sudden death most commonly occurs during mild exertion or sedentary activities. [4]

## Case One:

A 22 year young man while climbing the stairs of his office suddenly fell down on the platform, sustained few non-fatal injuries on different body parts and became unconscious. He was then immediately shifted to hospital where the doctor declared him brought dead.

The dead body was sent to the central morgue of S.C.B Medical College, Cuttack for post-mortem examination with an allegation of death due to accidental fall in the company premises. During autopsy, surgical bandage was found on the head. Nail beds were bluish in color and rigor mortis present all over the body.

There were presence of several fresh abrasions of small size on face, left ear and dorsum of left elbow with presence of two small superficial lacerations on right side eyebrow and left ear. On Internal examination, scalp was found contused (2cmx2 cm) on right side frontal area. Skull, duramater and brain were found intact without any injury.

Heart was enlarged, weighing about 400gms with gross hypertrophy of left ventricular wall and inter ventricular septum. There was severe narrowing of left ventricular lumen.

Multiple small fibrotic patches were found in the interventricular septum. Stomach was found to contain semi digested food without emitting any characteristic smell. All other internal organs were found intact.

## Case Two:

A 37 year old post-graduate student of Orthopedics suddenly became unconscious in the hospital and died immediately. During autopsy, face was congested, nail beds were bluish in color and there was no external or internal injury present on the body.

Hearts was enlarged in size, weighing about 450gms with thickening of left ventricular wall and inter ventricular septum and there was narrowing of left ventricular cavity. Other internal organs were found intact.

## Case Three:

A young 27 years man suddenly became unconscious during a police training camp. He was then immediately shifted to hospital where he was declared brought dead.

During autopsy, few superficial abrasions were found on the body. Hearts was found enlarged in size, weighing about 380s gm with thickening of left ventricular free wall and inter ventricular septum with narrowing of left ventricular cavity. Stomach was found to contain water and other internal organs were intact.

## Case Four:

A man of 32 years without any prior history of heart disease felt sudden chest pain in one morning and fell down and became unconscious. Immediately he was shifted to nearest hospital where he was declared brought dead by the doctor. During autopsy, no injury either external or internal detected on the body.

Heart was found enlarged in size, weighing about 420gms with gross asymmetrical hypertrophy of left ventricular wall and interventricular septum and narrowing of left ventricular lumen. Multiple small fibrotic patches were found in the left ventricular wall and inter ventricular septum. Stomach was empty. All other internal organs were found intact.

## Case Five:

A 38 year old man felt sudden chest pain in the morning and then immediately shifted to S.C.B Medical College, Cuttack for treatment where he was received dead by the casualty medical officer. During post-mortem examination, face was found congested without presence of any injury on the body.

Heart was enlarged weighing about 400gms with asymmetric hypertrophy of left ventricular wall and inter ventricular septum with presence of small fibrotic patches throughout them. Internal structures of neck were found intact and stomach was empty. Other internal organs were intact and congested.

In all these cases heart was sent for histopathology and subsequently histopathology report showed presence of extensive myocyte hypertrophy, myofibre disarray and extensive interstitial fibrosis suggesting hypertrophic cardiomyopathy of heart. [Fig. 1 & 2] After going through all the post-mortem reports, autopsy conducting doctors opined cardiac arrest resulting from hypertrophic cardiomyopathy as the cause of death in these cases.

## Discussion:

Hypertrophic cardiomyopathy was established as a diagnostic entity in 1950. [7] Males are more commonly affected than females. [4] It is the most common inherited cardio vascular disorder affecting up to 1 in 500 of the general population. [7]

The pathologic hallmark of this disease is myocardial hypertrophy, myocyte disarray (usually in association with myocardial fibrosis) and small vessel disease. [7] The most important microscopic feature of HCM is myocyte disarray which consists of:

(1) Loss of normal parallel arrangement of the myocytes
(2) Abnormal intercellular connections
(3) Variation in the diameter and length of individual myocyte. [7]

In HCM myofibre disarray affects about 5 to 40% of the total myocardium. [7] Many patients have abnormal small intramural arteries with apparent narrowing of their lumen. [7] This leads to obvious mismatch between myocardial

mass and coronary artery blood flow resulting in myocardial fibrosis and scarring. [7]

The clinical presentation of the disease are heterogeneous ranging from asymptomatic gene carriers who have very minor electrocardiographic and echocardiographic abnormalities to patients who have severe hypertrophy, diastolic dysfunction, arrhythmias and disabling symptoms. [7]

Arrhythmias are commonly seen in HCM. Paroxysmal episodes of atrial fibrillation occur in approximately 20-25% of patients. Ventricular arrhythmia is found to be the major cause of sudden death in HCM. [7]

Sudden death occurs throughout life with a peak in adolescence and young adult hood and may be the initial disease presentation occurring without warning sign and symptoms. [4] The presence of multiple risk factors in a patient subsequently increases the risk of sudden death. [4]

The incidence of sudden cardiac death has declined from 4% per annum to 1% nowadays as a result of evolving diagnostic criteria, family screening and modern treatment procedures. [4] The mortality for premature death from HCM is approximately 2-3% per annum and the great majority of such deaths are sudden and unexpected. [8]

ECG, echocardiography and magnetic resonance imaging are investigation of choice in hypertrophic cardiomyopathy. [7] However, echocardiography is the mainstay investigation for the diagnosis of HCM. [7] Patients suffering from this disease are benefited by symptomatic treatment, beta blockers, septal alcohol ablation and surgery. [4]

Patients affected with Hypertrophic cardiomyopathy with multiple risk factors for sudden death should be considered for Implantable cardioverter-defibrillator therapy. [4]

## Conclusion:

Hypertrophic cardiomyopathy is a relatively common cardiac disorder in which sudden unexpected death is the most unwelcome component, occurring throughout life, often in asymptomatic patients.

Hence, priority should be given to identify HCM patients who are at high risk and appropriate measures should be taken to prevent sudden death. Awareness should be created among young people regarding the course and outcome of this condition.

Patients complaining of unexplained episodes of syncope, collapse and chest pain require appropriate clinical, electrocardiographic and echocardiographic assessment. The family members of patients dying suddenly of HCM should undergo a noninvasive risk stratification assessment, including clinical history, echocardiography, maximal exercise tolerance testing and genetic screening to further prevent its hazardous outcome.

Steps should be initiated at the National level to decrease the mortality rate in our country. Forensic experts should keep in mind the condition of HCM while dealing with cases of sudden death in young people.

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**Fig. 1: Myocardium Showing Extensive Myocyte Hypertrophy (400x, H&E stain)**

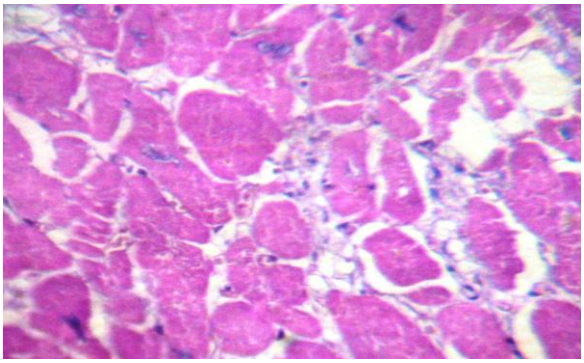

**Fig. 2: of Myocardium Showing Interstitial Fibrosis (400x, H&E stain)**