CHAPTER 213

Sudden Cardiac Death in Young People: Can it be Prevented

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INTRODUCTION

Sudden Cardiac Death (SCD) is defined as unexpected natural death from a cardiac cause within short period, generally less than one hour from the onset of symptoms in a person without any prior condition that would be fatal[1]. But death in young and previously healthy individuals with no identifiable cause at autopsy, termed sudden arryhthmic death syndrome (SIDS), contributes upto 5% of SCD in general population aged 16-64 years and approximately 25-35% in less than 40 years of age group[2]. SCD occurs in those who have a substrate in presence of triggering factors.

This chapter deals with mainly those people who are not known patients of cardiac diseases.

AETIOLOGY

1. In those over 50 years of age

- CAD: responsible for 75-80% of SCD
- Cardiomyopathies, Myocarditis
- Aortic Stenosis
- Drug and Toxins : which is same for those below 50 years of age.

2. In persons below 50 years of age

- Premature CAD
- Anomalous coronaries
- Hypertrophic Cardiomyopathy (HCM)
- Left ventricular Hypertrophy (LVH) due to any cause
- Long QT Interval
- Short QT Interval
- Catacholaminergic Polymorphic Ventricular Tachycardia (CPVT)
- Myocarditis
- Dilated CMP
- Valvular Heart Disease
- Drugs and Toxins

INCIDENCE

In USA, the incidence of SCD was 53/100000 population.[3] In China, the incidence of SCD was 41.8/100000 in a study on 678718 persons.[4] In India, a small study on over 22,000 population in Andhra Pradesh, there was autopsy incidence of SCD in 10-17% population.[5] Based on US data of 1996 which showed 0.16% incidence of SCD in total US population[6] , if we apply same incidence of 0.16% in 110 crore population of India, it was projected to be 17.6 lacs in India.[7] Since, 60% of world's heart disease is expected to occur in India by the end of decade, the incidence of SCD is expected to rise proportionately.[8]

FACTORS INFLUENCING SCD

Age : Increasing age enhances chances of SCD. In men of 50 years of age, the incidence of SCD was 100 per one lac people, which increased to 800 per lac in 75 years old i.e. 8 times higher.[9]

Sex : Women have less incidence of SCD.[10]

Smoking: Smokers have 2-3 times higher risk of SCD.

Obesity: Also enhances risk of SCD.

ECG: ECG change of myocardial infarction (MI) are well known.

Prolonged and short QT interval enhance chances of SCD.

LBBB an LVH: Enhance 1.5 times risk of SCD. LVH is an independent predictor of SCD.

Hypertension: Hypertension can cause SCD due to LVH and Resultant arrythmia.

Others : Low Socio-economic status[11]

Social Isolation

Psychiatric illnesses etc.

SCD IN CAD

Many young people develop premature CAD and MI and so they have chances of SCD. The bad thing about CAD is that in about 30% cases, SCD may be first symptom of MI. The risk of SCD in those patients who have developed in MI, is enhanced during first month and it gradually decreases with passage of time. However, with considerable remodeling and Scar formation, patient may be vulnerable to ventricular arrhythmia, the so called scar VT.[12,13] The risk of SCD also increases if patient develops heart failure and ischaemic events recur. The patients of CAD who are of older age, male, hypertensive, diabetic, smokers, obese having increased cholesterol have comparatively more chances of SCD. The other concomitant features such as heart failure, L.V. dysfunction, LVH, Tachycardia, other abnormalities in ECG, poor functional status enhance chances of SCD.[1,14,15]

INHERITED CARDIAC DISEASES

Many people dying suddenly and autopsy being normal, raised curiosity to find out the cause of death. Many such people were thought to have natural death unfortunately. In these victims of SCD with normal autopsy, post-mortem genetic testing showed that about 20-35% deaths were due to inherited arrhythmic disorders.[16-18]

HYPERTROPHIC CARDIOMYOPATHY (HCM)

It is the commonest cause of SCD in this group. In those with history of pain chest, syncope, cardiac arrest, VT, family history of SCD, then one must be alerted. Echocardiography shows LV wall thickness (>30mm in high risk case) and LV outflow obstruction may be present.[19]

Prevention of SCD

Beta-blockers and amiodarone are being used for a long time, but there is no improvement in survival. ICD is a better option. These patients should avoid intense physical activity, competitive sports and alcohol use.

Catacholaminergic Polymorphic Ventricular Tachycardia (CPVT)

It is not uncommon. The presenting feature is syncope/ventricular arrhythmia in children and young adults, triggered by physical or mental stress. CPVT may be suspected by PVCS on exertion or by their appearance during exercise testing. 24 hour ECG may also help. CVPT accounts for 14-21%. Of SCD observed in young adults.[20]

LONG QT SYNDROME

Prolonged QT means QTC is >480 ms, when QTC prolongation is over 500 ms, then the risk of SCD is markedly increased due to ventricular arrhythmia like VT/VF, torsades. Risk of SCD is increased when there is unexplained syncope/cardiac arrest, documented lethal arrhythmias like VT/VF.

Incidence of SCD in long QT syndrome may be 2.3 to as high as 5 times.[21]

Triggering factors to be avoided are :

Acute mental and physical stress, hypothermia etc. Some drugs are known to prolong QT interval: Terfenadine, Cisapride, Lithium, Qunidine, Sotalol, Amiodarone, Domperidon, Clarithromycin, Haloperidol etc.

Prevention of SCD in long QT

- Offender drug should be stopped
- Beta-Blocker drugs are most preferred drugs for prevention of arrhythmia and syncope.
- ICD may be needed when beta blockers are not sufficient.

ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY/DYSPLASIA (ARVC)

In this disease right ventricle is affected and is replaced by fibrofatty tissue, but L.V. can also be affected. It is a genetically determined heart muscle disease.[22,23] It can cause syncope, VT and in some case SCD. It can be diagnosed by Echo. Symptomatic patients may need ICD.

EARLY REPOLARIZATION SYNDROME

It is a common ECG finding in 2-5% population. It was considered a benign ECG finding for over half century. It can cause life threatening ventricular arrhythmia in some cases.[24]So, those persons who have early repolarization form of ECG and ventricular arrhythmia and unexplained syncope, 24 hour ECG, EP study should be done.

Brugada syndrome

Brugada syndrome can also cause SCD below 50 years of age, but it is rarely seen. The typical ECG pattern is ST elevation in lead V_1 –V_3 with RBBB. Death is due to polymorphic VT or VF.[25]

TRIGGERING FACTORS OF SCD

These are important because avoidance and taking care of these may prevent SCD.

MENTAL STRESS

Stress not only causes MI, but also triggers life threatening arrhythmias. It is already known that about 75-80% SCD is due to CAD. The INTER HEART study[26] has shown that diverse stressful life events like death or major illness of spouse or a close family member, major self-illness, conflict among family members, loss of job etc. can cause SCD. Disasters like earthquake is known to cause miseries in the form of displacement from house, increase in mental stress immensely. In such a study of earthquake victims, the cardiac death was enhanced by 2-5 times even they had no physical trauma by earthquake.[27]Mental stress is also known to prolong QT interval.[28]

PHYSICAL STRESS

There is data that physical exertion was responsible for collapse and cardiac arrest of 11-17% persons.[29] In an athlete meet in USA, SCD was found to be a leading cause of death.[30] In fact many SCDs occur during competitive sports, run for job for recruitment in police or military, vigorous physical activity by a person who is spending sedentary life. In most of these situations, there must be some substrate in the form of CAD or genetic cardiac disorders or structural heath disease.

ELECTROLYTE IMBALANCE

Hypokalemia, Hypomagnesemia, Hypocalcemia can also trigger SCD.

WHAT ARE WARNING SIGNS

- Undiagnosed syncope
- History of cardiac arrest
- Family history of SCD
- History of VT/VF
- Warning sign of ECG
- Echo evidence of disease
- History of Angina on effort

These persons should be meticulously investigated. Simple tests like ECG, Echo, exercise test, 24 hour ECG, ELR should be done first and EPS, coronary angiography,

CT Angio etc. should be done whenever required.

With increasing awareness about genetic cardiac diseases, more and more causes of SCD/Cardiac arrest are being diagnosed. Earlier many cases were dumped as idiopathic VF. Many young victims of SCD were thought to die a natural death. The CASPER trial[31] has shown the path, as cause of cardiac arrest was detected in 70% people.

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