Sudden cardiac death in apparently healthy young persons: Is it preventable?

Harendra Kumar, MD, PhD
Senior Consultant Cardiologist and Director, Indira Gandhi Institute of Cardiology, Patna Medical College Campus, Patna, India

Abstract

Many people have sudden cardiac death (SCD) without known structural heart disease. Although coronary artery disease (CAD) may be the commonest cause of SCD, genetic cardiac diseases are increasingly being recognized as culprit for SCD in the young. Death may be the first symptom in inherited cardiac disease as well as in CAD. With increasing investigative facilities and awareness more and more cases can be diagnosed and preventive strategies for SCD can be undertaken. The mystery of SCD with structurally normal heart at autopsy is gradually disappearing. The number of idiopathic ventricular fibrillation (VF) is shrinking as more cases are being diagnosed. Some of the triggering factors of SCD in patients with CAD or inherited cardiac diseases which are often overlooked are mental stress, acute physical stress including competitive sports, electrolyte imbalance, etc. History of syncopes, cardiac arrest, family history of premature SCD, competitive sports, electrolyte imbalance, etc. History of syncopes, cardiac arrest, family history of premature SCD, and mental stress are important risk factors. Further advancement in technology for wider clinical use and its highly effective to diagnose genetic diseases, but needs further advancement in technology for wider clinical use and it should be cost-effective too.

Key Words
- Sudden cardiac death
- Inherited cardiac disease
- Mental stress
- Physical stress
- Death in athletes
- Coronary artery disease
- Lethal arrhythmias

Introduction

Sudden cardiac death (SCD) is responsible for 80% of deaths annually in western countries, which is much more when compared to deaths from stroke, lung and breast cancer, and AIDS put together. There is no data in India, but based on US data of 1996 which showed 0.16% incidence of SCD in total population, the projected annual incidence of SCD in India will be around 17.6 lacs at present, considering population of 1.10 crore and same incidence of 0.16% is taken for calculation.

SCD is defined as natural death due to cardiac causes, heralded by abrupt loss of consciousness within one hour of onset of symptoms. Preexisting cardiac disease may have been known to be present, but the time and mode of death are unexpected. In cardiac arrest, there is abrupt cessation of cardiac mechanical function which may be reversible by a prompt intervention, which will lead to death in its absence. Although SCD can occur in patients with known structural heart disease which may be symptomatic or asymptomatic, in this article, SCD in persons without known heart disease will be mainly discussed.

Causes of SCD

Some of the important causes of SCD are shown below (for full list of causes, reader is advised to consult textbooks):
- CAD: Atherosclerotic CAD like MI, ischemia; Congenital anomalies of coronary arteries, coronary artery embolization like from valvular endocarditis, coronary artery spasm, etc.
- Cardiomyopathy: Dilated cardiomyopathy (non-inherited) myocarditis, peripartum, cardiomyopathy (CMP), left ventricular hypertrophy (LVH), etc.
- Valvular heart disease and mechanical defects of myocardium: AS, AR, MVP rupture of structure of MV apparatus, prosthetic valve dysfunction, etc.
- Congenital heart disease: Established diseases, e.g., TOF, TGA, etc.
- Sudden infant death syndrome (SIDS): Long QT syndrome, myocarditis.
- Inherited cardiac diseases: Hypertrophic cardiomyopathy (HCM), DCM, ARVC, LQTS, Brugada syndrome, catecholaminergic polymorphic VT, short QT, early repolarization syndrome, etc.
- Primary electrical abnormality: Heart blocks, sinus node disease, idiopathic VT/VF
- Drugs and toxins
- Acute physical and mental stress
- Miscellaneous: Cardiac tamponade, rupture of aortic aneurysm massive, pulmonary thromboembolism (PTE), etc.

There is a long list of causes of SCD with known structural heart diseases, but the unfortunate fact is that neither most of the patients nor physicians are aware of it. Another important point is that death may be the first symptom in many such diseases, especially inherited cardiac diseases. It was found that etiology of victims of such SCD was inconclusive in many cases and many young people were reported to die a natural death! In one autopsy series on victims of SCD without structural heart disease, up to 50% of SCD was first manifestation of disease. The etiology of sudden infant death syndrome which causes death in children below one year of age remained unexplained in good number of cases even after thorough investigation. But later 10% of such deaths, SIDS, were found to be caused by inherited cardiac disease specially related to mutation in ion channels. The likely cause of SCD may be different in different age groups, though in presence of a triggering factor SCD can occur at any age.

HCM is the most common cause of SCD below 35 years of age and CAD in over 35 years of age. Several etiological factors may contribute to SCD. Increasing age, male sex, obesity, alcohol use >5 drinks/day and family history of SCD are well known risk factors. But more attention has been drawn recently on stress and inherited cardiac diseases.

Role of mental stress

Mental stress is a well-known precipitatory factor of myocardial infarction (MI). If we include all age groups CAD is responsible for about 80% of SCD. Disasters like earthquake which cause displacement from home and miseries have increased cardiac events. In an earthquake in 1994 in the USA, the number of cardiac deaths in victims was 2 to 5 times higher than in those who had no physical trauma or increased physical exertion. In the month after terrorist attack on the world trade center in New York in 2001, the rate of shocks of ICD increased. Stressful life events like loss of job, retirement, death of spouse, death, or major illness of a close family member, loss of crop, failure in business, violence, major intra family conflict, major personal injury or illness or other major stress has been found to be a risk factor of acute MI in the INTERHEART study. People residing in areas where hostile missile attacks are common, have increased chances of acute myocardial infarction (AMI), which can cause SCD. Similar disasters like tsunami might have caused similar impact on affected population, but data are not available. Firemen who are fighting fire have 10 to 100 fold enhanced risk of developing MI than those firemen who are sitting in office or doing non-emergency work. Excess emotional stress like extreme anger, intense excitement like watching an important match can act as a triggering factor of MI. Sexual activity in persons with sedentary lifestyle or doing extravaganital sexual activity which has added emotional stress factor may also act as triggering factor.

Role of physical stress

It is known that uncustomed vigorous physical activity; especially those in habit of sedentary lifestyle have increased chances of MI/SCD. SCD during competitive sports/pace for recruitment or promotion purpose in police or marathon run or physical training in military people or physical training in schools report SCD in young people and children. Review of emergency medical record has shown that 11–17% of victims of cardiac arrest collapsed during or immediately after exertion. SCD has been found to be leading medical cause of death in university student athletes in the USA.

Intense physical activity may also act as trigger for arrhythmia in various inherited cardiac diseases and
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Key Words

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• Inherited cardiac disease
• Mental stress
• Physical stress
• Death in athletes
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Sudden cardiac death (SCD) is responsible for 8 lacs deaths annually in western countries, which is much more when compared to deaths from stroke, lung and breast cancer, and AIDS put together.1 There is no data in India, but based on US data of 1996 which showed 0.16% incidence of SCD in total population, the projected annual incidence of SCD in India will be around 17.6 lacs at present, considering population of 110 crore and same incidence of 0.16% is taken for calculation.

SCD is the most common cause of SCD below 35 years of age and CAD in over 35 years of age. Several etiological factors may contribute to SCD. Increasing age, male sex, obesity, alcohol use > 5 drinks/day and family history of SCD are well known risk factors. But more attention has been drawn recently on stress and inherited cardiac diseases.

Role of mental stress

Mental stress is a well-known precipitatory factor of myocardial infarction (MI). If we include all age groups CAD is responsible for about 80% of SCD.2 Disasters like earthquake which cause displacement from home and miseries have increased cardiac events. In an earthquake in 1994 in the USA, the number of cardiac deaths in victims was 2 to 5 times higher than in those who had no physical trauma or increased physical exertion.3 In the month after terrorist attack on the world trade center in New York in 2001, the rate of shocks of ICD increased.4 Stressful life events like loss of job, retirement, death of spouse, death, or major illness of a close family member, loss of crop, failure in business, violence, major intra family conflict, major personal injury or illness or other major stress has been found to be a risk factor of acute MI in the INTERHEART study.5 People residing in areas where hostile missile attacks are common, have increased chances of acute myocardial infarction (AMI), which can cause SCD. Similar disasters like tsunami might have caused similar impact on affected population, but data are not available. Firemen who are fighting fire have 10 to 100 fold enhanced risk of developing MI than those firemen who are sitting in office or doing non-emergency work.6 Excess emotional stress like extreme argument, anger, intense excitement like watching an important match can act as a triggering factor of MI.7 Sexual activity in persons with sedentary lifestyle or doing extramarital sexual activity which has added emotional stress factor may also act as triggering factor.8

Role of physical stress

It is known that unaccustomed vigorous physical activity; especially those in habit of sedentary lifestyle, have increased chances of MI/SCD. SCD during competitive sports/race for recruitment or promotion purpose in police or marathon run or physical training in military people or physical training in schools report SCD in young people and children. Review of emergency medical record has shown that 11–17% of victims of cardiac arrest collapsed during or immediately after exertion.9 SCD has been found to be leading medical cause of death in university student athletes in the USA.10 Intense physical activity may also act as trigger for arrhythmia in various inherited cardiac diseases and
High risk SCD

- Females
- Longer QT interval
- Unexplained syncope
- Past history of SCD/cardiac arrest
- Documented torsades/VT
- Inducible ventricular arrhythmia during EPS
- Prolonged mental stress

Acute mental stress can cause prolongation of QT interval. Mental stress can also trigger arrhythmias and SCD. In one study,7 mental stress has been found to be triggering factor for cardiac events in patients with LQTS who had first cardiac arrest before age of 20 years. Prolonged mental stress due to stressful life events may be a trigger for arrhythmic events in LQTS.

The underlying mechanism may be adrenergic stimulation, change in autonomic function, prolongation of QT interval, and electrical instability of heart. So, there are triggering factors which increase chances of arrhythmic events in LQTS and this explains why all patients with LQTS do not behave in the same way.

Treatment for prevention of SCD in LQTS

- Drugs that prolong QT should be stopped
- Beta-blockers are most preferred drugs for prevention of syncope and arrhythmia, but in LQTS 3 these may worsen the problem
- ICD (with beta-blockers) in those with history of SCD/cardiac arrest
- Left cardiac sympathetic neural denervation who are symptomatic despite beta-blockers

Triggering factors to be avoided are:

- Acute physical stress
- Competitive sports
- Acute mental stress
- Electrolyte imbalance

Acquired long QT syndrome

The main causes of acquired LQTS are:

- Electrolyte abnormalities like:
  - Hypokalemia
  - Hypomagnesemia
  - Hypocalcemia
  - Hypothyroidism
  - CNS injury: Most common – subarachnoid hemorrhage
  - Drugs
  - Antiarrhythmics:
    - Amiodarone
    - Sotalol
    - Quinidine
    - Procainamide
    - Disopyramide, etc.
  - Other drugs:
    - Tefedrine
    - Astemizole
    - Cisapride
    - Lithium

Odds with less hazard potential:

- Chlorbromycin
- Haloperidol
- Doperidin
- Thiordanizine
- Chlorpromazine, etc.

Management is according to etiology.

Short QT syndrome (SQTS)

SQTS was recognized in the year 1999. It is an inherited cardiac disease encoding three potassium channels and there is propensity to syncope, life threatening cardiac arrhythmias and SCD. QT interval of 300 ms was thought as cut off point but now it is believed to be 350 ms.

In a series of 53 patients with SQTS, about 90% of patients had a family or personal history of SCD. SCD/cardiac arrest was the presentation in 32% persons.

Treatment

Quinidine and hydroquinidine are useful drugs. But ICD is most preferred form of treatment in those with history of cardiac arrest to prevent SCD. Till more data are available, competitive sports and vigorous physical activity should not be allowed in such patients.

Brugada syndrome

Brugada syndrome kills in 3rd and 4th decade of life. It is due to a mutation in SCN5A gene which causes a reduction of sodium current leading to abnormal conduction and depolarization of ventricular myocardium. The hallmark of Brugada syndrome is a right bundle branch block (RBBB) with ST segment elevation in at least two leads of the right precordial area (V1-V2) and this pattern is observed in about 15% of the general population. The diagnosis is confirmed by the presence of a J-point elevation of at least 0.2 mV and a positive terminal force of at least 0.1 mV in at least two of the three right precordial leads. The prevalence of Brugada syndrome is estimated to be about 1 in 1000 individuals and it is more common in men than in women (with a male-to-female ratio of 5:1). The disease is autosomal dominant and the gene responsible for Brugada syndrome is the SCN5A gene located on chromosome 3q21.1-q21.3. Genetic testing is important for diagnosis and genetic counseling.

Treatment options include:

1. Beta-blockers: They are the first-line treatment for Brugada syndrome. Beta-blockers work by slowing the heart rate and reducing the risk of ventricular arrhythmias.
2. Antiarrhythmic drugs: These drugs are used to prevent or control arrhythmias. Examples include amiodarone and sotalol.
3. ICD implantation: An ICD is an automatic internal defibrillator that can detect and treat fatal arrhythmias by delivering an electric shock to the heart.
4. Catheter ablation: This procedure involves using a catheter to deliver energy to the heart to prevent arrhythmias.
5. Lifestyle modifications: Avoiding alcohol and caffeine, quitting smoking, and managing stress can help reduce the risk of arrhythmias.
6. Diet: A healthy diet rich in fruits, vegetables, and whole grains can help reduce the risk of arrhythmias.

It is important to discuss treatment options with a healthcare provider to determine the best course of action.

High risk LQTS

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- Documented torsades/VT
- Inducible ventricular arrhythmia during EPS
- Prolonged mental stress

Acute mental stress can cause prolongation of QT interval. Mental stress can also trigger arrhythmias and SCD. In one study, mental stress has been found to be triggering factor for cardiac events in patients with LQTS who had first cardiac arrest before age of 20 years. Prolonged mental stress due to stressful life events may be a trigger for arrhythmic events in LQTS.

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- Electrolyte abnormalities like:
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  - CNS injury: Most common – subarachnoid hemorrhage
  - Drugs
    - Antiarrhythmics:
      - Amiodarone
      - Sotalol
      - Quinidine
      - Procainamide
      - Disopyramide, etc.
    - Other drugs:
      - Tefedrine
      - Astemizole
      - Cisapride
      - Lithium
  - Drugs with less hazard potential:
    - Chlorbromycin
    - Haloperidol
    - Doperidin
    - Thiordanizine
    - Chlorpromazine, etc.

Management is according to etiology.
structural heart diseases like hypertrophic cardiomyopathy (HCM), long QT syndrome (LQTS), short QT syndrome (SQTS), Brugada syndrome, arrhythmogenic right ventricular cardiomyopathy (ARVC), catecholaminergic polymorphic VT (CPVT), abnormal/congenital defects in coronary arteries, severe aortic stenosis, etc. During physical stress, there is activation of sympathetic nervous system and catecholamine surge which increase heart rate, vascular tone, besides many other changes. These can trigger arrhythmia/MI, especially in presence of substrate. In a 10 years review study on 114 persons 18 years of age or older who were taking organized physical training, 31.6% SCD were attributed to exertional death during running. In a study on marathon runners in which cardiac magnetic resonance imaging was done immediately after running 26.2 miles, acute dilatation of right atrium and right ventricle was seen in one third of participants along with diminished RV ejection fraction in some.  

- **Coronary artery disease (CAD)**

  CAD is responsible for about 80% of SCD. Nearly 50% of all CAD deaths are sudden and approximately 15% of these deaths are the first clinical manifestation of the disease. It can cause SCD later also due to scar induced ventricular arrhythmia. Silent ischemia may be an important cause, as the person is caught unaware. Prevalence of premature cardiac troponin in healthy persons. It is likely to be a benign process.

- **Hypertrophic cardiomyopathy (HCM)**

  HCM is the most common genetically determined cardiovascular disease. It is the most common cause of SCD in young and athletes, responsible for one third deaths. **High risk patients**
  
  - History of (H/O) cardiac arrest
  - Undiagnosed syncope
  - Family H/O premature SCD
  - H/O VT
  - Onset of symptoms in childhood
  - LV wall thickness > 30 mm
  - LV outflow obstruction
  - Myocardial ischemia

  Unexplained syncope in HCM patients was associated with 5 times high risk of SCD in one study. In patients with HCM who had giddiness or lightheadedness only, study with ECG loop recorder revealed prolonged episodes of non-sustained VT as the cause of above symptoms.

  **Treatment for prevention of SCD in HCM**

  Beta blockers are being used for symptomatic relief for a long time. Amiodarone has been also used for treatment, but no improved survival with amiodarone or beta blockers has been seen. ICD implantation is the best option to prevent SCD.

  Patients with HCM should avoid competitive sports including weight lifting, intense exercise, alcohol intake (as it enhances LVOT gradient), hypokalemia. So diuretics should be used with caution in patients with HCM with a watchful eye on serum potassium level.

- **Long QT syndrome (LQTS)**

  It may be genetic or acquired and is an important cause of SCD. A QTc of > 480 ms is considered prolongation but QTc > 500 ms, the risk of SCD below 40 years of age is 50%. In the International LQTS registry, risk of SCD was associated with QTc > 530 ms and if there was history of syncope in the past 10 years. Life threatening ventricular arrhythmias like VT, torsade-de-pointes and VF are common and cause SCD. Twelve types of LQTC have been identified, but the most common are as follows:

  - **LQTS 1:** The involved gene controls slow inactivating potassium channel (Kv1.4).
  - **LQTS 2:** The involved gene controls rapidly inactivating potassium channel (IKr) cardiac dependent
  - **LQTS 3:** The involved gene controls sodium channel. Arrhythmias occur only when heart rate is slow like during sleep.

  One ominous fact is that up to 30% of patients with LQTS have a normal or borderline QT interval, necessitating other methods for confirmation including genetic testing, phenotype detection, or both. Fortunately some patients with LQTS do not have arrhythmias in their life, whereas many others face life-threatening arrhythmias.

  **High risk LQTS**

  - Females
  - Longer QT interval
  - Unexplained syncope
  - Past history of SCD/cardiac arrest
  - Documented torsades/VF
  - Inducible ventricular arrhythmia during EPS
  - Prolonged mental stress

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  **Treatment for prevention of SCD in LQTS**

  - **Drugs**
    - Beta-blockers are most preferred drugs for prevention of syncope and arrhythmia, but in LQTS 3 these may worsen the problem
    - ICD (with beta-blockers) in those with history of cardiac arrest or syncope/VT despite beta-blockers
    - Left cardiac sympathetic neural denervation who are symptomatic despite beta-blockers

    Triggering factors to be avoided are:
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    - Competitive sports
    - Acute mental stress
    - Electrolyte imbalance

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  The main causes of acquired LQTS are:

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  In a series of 53 patients with SQTS, about 90% of patients had a family or personal history of SCD. SCD/cardiac arrest was the presentation in 32% persons.

  **Treatment**

  Quinidine and hydroquinidine are useful drugs. But ICD is most preferred form of treatment in those with history of cardiac arrest to prevent SCD. Till more data are available, competitive sports and vigorous physical activity should not be allowed in such patients.
Sports and severe physical exertion should be forbidden. As a preventive measure for SCD, participation in treatment is mainly with beta-blockers which has catecholamine infusion can also unmask occult CPVT. Exercise testing. Holter study may also be useful. Suspected by ventricular ectopics on exercise or during exercise. It is a disorder of intracardiac calcium handling which can cause SCD. The usual presentation is syncope, VT/VF, in children and adolescents, usually triggered by physical or exercise testing. Holter study may also be useful. It is an inherited heart muscle disease, but also ion channel disorder. It mostly affects RV with fibrofatty tissue replacement causing RV dysfunction; but involvement of LV with enlargement occurs in 50–75% cases. The usual presentation is palpitation, arrhythmia, syncope and in some cases SCD. The commonest arrhythmia is VT arising from RV. Majority of patients with ARVC have some ECG changes which may be non-specific. They include T inversion in leads V1-V3 and epsilon wave. Medical treatment is amiodarone or sotalol when ICD is not feasible. ICD is advised in those with VT/VF despite optimal medical treatment. Early repolarization syndrome

Early repolarization syndrome is a common ECG finding in young healthy persons. It was considered benign for 50 years. In the year 2000, one study revealed that early repolarization can lead to VT/VF in some cases. Earlier these SCWs were possibly included in idiopathic VT/VF group, but higher incidence of SCD in those with early repolarization pattern linked SCD to this disorder. Involvement of inferior and lateral leads were more associated with SCD.

Catecholaminergic polymorphic ventricular tachycardia (CPVT)

It is a disorder of intracardiac calcium handling which can cause SCD. The usual presentation is syncope, VT/VF in children and adolescents, usually triggered by physical or emotional stress. The usual mortality is 30–50% by age 50 years. It is often familial. ECG is usually normal or there may be bradycardia with “U” waves. CPVT may be suspected by ventricular ectopics on exercise or during exercise testing. Holter study may also be useful. Catecholamine infusion can also mask occult CPVT. Treatment is mainly with beta-blockers which has significantly reduced events of cardiac arrest/SCD in these cases. As a preventive measure for SCD, participation in sports and severe physical exertion should be forbidden including those on beta-blockers.

Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC)

It is an inherited heart muscle disease, but also ion channel disorder. It mostly affects RV with fibrofatty tissue replacement causing RV dysfunction; but involvement of LV with enlargement occurs in 50–75% cases. The usual presentation is palpitation, arrhythmia, syncope and in some cases SCD. The commonest arrhythmia is VT arising from RV. Majority of patients with ARVC have some ECG changes which may be non-specific. They include T inversion in leads V1-V3 and epsilon wave. Medical treatment is amiodarone or sotalol when ICD is not feasible. ICD is advised in those with VT/VF despite optimal medical treatment.

Left ventricular hypertrophy (LVH)

LVH is a strong independent risk factor for SCD irrespective of etiology, and it enhances chances of arrhythmia and heart failure. The two possible mechanisms are scarring in a hypertrophied mass or ischemia or both.

Idiopathic ventricular fibrillation

As more and more inherited heart diseases, especially without structural cardiac defects are being recognized as cause of SCD, the incidence of SCD attributable to idiopathic VF is shrinking.

Electrolyte disorders

Lower blood levels of potassium, magnesium, calcium, and acidosis may trigger arrhythmias particularly VT. Hyperkalemia may cause 1st degree to 3rd degree heart block, asystole, etc. Hypomagnesia is known to cause torsade-de-points and SCD. Electrolytes should be checked if patients are on diuretics or if there is any condition where its lower or higher levels are expected.

So, the triggering factors of SCD may be acute mental stress, undue physical exertion, especially in those with sedentary lifestyle, electrolyte disturbance, drugs or toxins, changes in hemodynamic status, etc.

How to evaluate a suspected case

• History: Past H/O syncope, heart disease, drug history, cardiac arrest family H/O cardiac arrest/syncpe/SCD

Blood tests

• ECG: Ischemia, conduction disease, QRS duration, QT interval, Brugada pattern, LVH, PVCs, NSVT/VT early repolarization, etc.

• Echocardiography: Large LV, LVH, ARVC and many more

• Other imaging modalities: CT, MRI for CMP, myocarditis, myocardial scar, etc.

• Exercise test:
  o Ischemia
  o To detect occurrence of PVCs, NSVT, VT, etc.

• Provocative test

• Holter study, loop recorders: May detect arrhythmias

• EPS: For knowing mechanism and treatment of arrhythmia

• Coronary angiography

Importance of meticulous investigations in cardiac arrest survivors in CASPER trial is an eye-opener, as it resulted in detecting cause in apparently unexplained cardiac arrest in 70% of patients. These persons had normal ECG, echocardiogram and without CAD. The reason of cardiac arrest was detected in 35 out of 50 patients, which are as follows:

<table>
<thead>
<tr>
<th>Disease</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>LQTS</td>
<td>8</td>
</tr>
<tr>
<td>CPVT</td>
<td>8</td>
</tr>
<tr>
<td>ARVC</td>
<td>6</td>
</tr>
<tr>
<td>Early repolarization syndrome</td>
<td>5</td>
</tr>
<tr>
<td>Coronary artery spasm</td>
<td>4</td>
</tr>
<tr>
<td>Brugada syndrome</td>
<td>3</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>1</td>
</tr>
</tbody>
</table>

Additionally, genetic testing identified disease carrying mutations in 47% of patients. Succeeding of family members of these patients identified 24% affected persons in the same study.

Prevention at community level

• Increasing awareness about SCD

• Screening programs with basic ECG and echocardiography whenever possible

• Placement of defibrillators with trained paramedics at strategic points like Railway stations, airports and crowded places which are comparatively distant from hospitals.

In future, as the awareness will increase, people will have genetic horizon rather than astrological one. We still have a long battle to fight.

References

In suspected cases, challenge with a sodium channel blocker like flecainide may reproduce classical ECG pattern. In suspected cases, challenge with a sodium channel blocker like flecainide may reproduce classical ECG pattern.

- **Life-threatening arrhythmias**
- **Persistent ECG pattern**
- **Family H/O SCD**
- **H/O syncopes**

- **Electrolyte disorders**

  - Lower blood levels of potassium, magnesium, calcium, and acidosis may trigger arrhythmias particularly VT. Hyperkalemia may cause 1st degree to 3rd degree heart block, asystole, etc. Hypomagnesia is known to cause torsade-de-pointes and SCD. Electrolytes should be checked if patients are on diuretics or if there is any change in hemodynamic status, etc.

  - So, the triggering factors of SCD may be acute mental stress, undue physical exertion, especially in those with sedentary lifestyle, electrolyte disturbance, drugs or toxins, changes in hemodynamic status, etc.

- **How to evaluate a suspected case**

  - History: Past H/O syncopes, heart disease, drug history, cardiac arrest family H/O cardiac arrest/syncope/SCD

- **Blood tests**

- **ECG: Ischemia, conduction disease, QRS duration, QT interval, Brugada pattern, LVH, PVCs, NSVT/VT early repolarization, etc.**

- **Echocardiography: Large LV, LVH, ARVC and many more**

- **Exercise test:**
  - Ischemia
  - To detect occurrence of PVCs, NSVT, VT, etc.

- **Provocative test**

- **Holter study, loop recorders:** May detect arrhythmias

- **EPS:** For knowing mechanism and treatment of arrhythmia

- **Coronary angiography**

  - Importance of meticulous investigations in cardiac arrest survivors in CASPER trial is an eye-opener, as it resulted in detecting cause in apparently unexplained cardiac arrest in 70% of patients. These persons had normal ECG, echocardiogram and without CAD. The reason of cardiac arrest was detected in 35 out of 50 patients, which are as follows:

    - **Disease**
    - **Number**

    - **LQTS**
    - 8

    - **CPVT**
    - 8

    - **ARVC**
    - 6

    - **Early repolarization syndrome**
    - 5

    - **Coronary artery spasm**
    - 4

    - **Brugada syndrome**
    - 3

    - **Mycarditis**
    - 1

  - Additionally, genetic testing identified disease carrying mutations is 47% of patients. Sequencing of family members of these patients identified 24% affected persons in the same study.

- **Plan to prevent SCD in normal looking young people**

  - To identify high risk or suspected persons

    - **H/O palpitation with restlessness**
    - **Obesity**
    - **Un-investigated irregular pulse**
    - **Dyspnea at low level of physical activity**

  - **Investigations as required**

    - **Target school**
    - To screen students at least once during school
    - To investigate them at least with ECG and further if needed

    - **Participation in competitive sports and strenuous physical activity should be forbidden in known cases with substrate.**

    - **ECG interpretation in athletes has been recently published by European society of cardiology.** Some ECG changes are training related like sinus bradycardia, LVH by QRS voltage criteria. But abnormal ECG changes not related to training may be seen which are: pathological Q wave, RV1, RBBB or LBBB long QT or short QT, inverted T waves, ST segment depression, left atrial enlargement, ST segment elevation in V1-V3 with RRBB (Brugada type).

- **Prevention at community level**

  - Increasing awareness about SCD
  - Screening programs with basic ECG and echocardiography whenever possible
  - Placement of defibrillators with trained paramedics at strategic points like Railway stations, airports and crowded places which are comparatively distant from hospitals.

In future, as the awareness will increase, people will have genetic horizon rather than astrological one. We still have a long battle to fight.

### References


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Primary prevention of sudden cardiac death

Ashok Garg, MD, DM
Diplomate American Board of Cardiovascular disease
Electrophysiologist, Arizona Heart Clinic, Phoenix, Arizona, USA
Director, Phoenix Institute of CPR and Defibrillation, New Delhi, India

Abstract

Sudden Cardiac Death (SCD) from cardiac arrest is the most common cause of death worldwide, accounting for more than 55 percent of all cardiovascular deaths worldwide. A large percentage, about 70-80% of persons who die of SCD have no past history of heart disease. Also, SCD is the mode of death in about 50% of persons with heart disease. The best way to prevent SCD is early defibrillation within seconds or 2-3 minutes of its onset. Prophylactic implantation of defibrillators which provide immediate defibrillation have been immensely successful in persons with left ventricular ejection fraction less than 35 percent. However, ventricular fibrillation leading to SCD, occurs in much larger absolute numbers in persons with no or non-significant heart disease. Publicly accessible automated external defibrillator (AED) has been shown to be a highly successful strategy. There is however, no good strategy to reduce SCD at home, where most occurrences of ventricular fibrillation happen. Presence of AED at home may be useful where community based emergency medical services are unavailable or overwhelming.

Key Words

Sudden cardiac death • Implantable defibrillator • Automated external defibrillator

Introduction

Primary prevention of Sudden Cardiac Death (SCD) is a major challenge. SCD is common occurrence, is unpredictable and is often un-expected. It occurs in persons with heart disease, and even in apparently healthy persons, with no known heart disease. Surgical implantation of defibrillator in patients, who have survived SCD, has been shown to be immensely successful in preventing SCD. There are no clear and very successful strategies in preventing SCD in the general population or those who have never had ventricular fibrillation. The article discusses the strategies for primary prevention of sudden cardiac death.

Patient case 1

A 55-year-old man suffered a myocardial infarction (MI), for which he had primary angioplasty and stent placement. Four months later, however, his left ventricular ejection fraction was found to be only 30%. You feel he is at risk of sudden cardiac death and advised him and his family implantation of defibrillator. When he finds out its cost, he feels it is very expensive, he requests more information on preventing sudden cardiac death (SCD)?

Patient case 2

A 52-year-old man with no pre-existing illness was, at the Delhi airport, standing in the line for security check when he suddenly collapsed. A bystander immediately started cardiopulmonary resuscitation (CPR), and applied an automated external defibrillator (AED). The AED