ORIGINAL RESEARCH PAPER
Volume-9 | Issue-4 | April-2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

SUDDEN CARDIAC DEATH IN YOUNG PEOPLE - A REVIEW

Medicine

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ABSTRACT

Although sudden cardiac death(SCD) is rare in young people,but when it occurs, it's a devastating event. A lot of research has been done in this area. This review focuses on epidemiology of SCD and to stratiges for prevention, resuscitation, and identification of those at high risk of SCD.This review will also cover the pathophysiology of SCD and the predisposing disease processes.

KEYWORDS

Cardiomyopathy, Myocarditis, Sudden Cardiac Death, Ventricular Tachycardia.

INTRODUCTION

Sudden cardiac death is defined as sudden unexpected death attributed to cardiac arrest, which if witnessed occurs within one hour of symptom onset and In unwitnessed cases, the definition is often expanded to include unexpected deaths where the subject was documented to be well within the preceding 24 h. 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[1].SCD occurs in those who have a substrate in presence of triggering factors.

INCIDENCE

In USA, the incidence of SCD was 53/100000 population. In India, a small study on over 22,000 population in Andhra Pradesh and there was autopsy incidence of SCD in 10-17% population.[2] 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.

Cardiovascular Conditions Associated with Sudden Death in the young people are-

1.**Structural Heart Diseases**

a. hypertrophic cardiomyopathy
b. Coronary artery anomalies
c. Left ventricular hypertrophy of indeterminate cause
d. Myocarditis
e. ruptured aortic aneurysm (Marfan syndrome)
f. arrhythmogenic right ventricular cardiomyopathy
g. Myocardial bridging
h. aortic valve stenosis
i. atherosclerotic coronary artery disease
j. Dilated cardiomyopathy
k. Myxomatous mitral valve degeneration
l. asthma (or other pulmonary condition)
m. heat stroke

2.**Channelopathies**

a. Long Qt syndrome
b. Short Qt Syndrome
c. Catecholaminergic polymorphic ventricular tachycardia
d. Brugada syndrome
e. Idiopathic ventricular fibrillation

3.**ELECTROLYTE IMBALANCE**

a. Hypokalemia,
b. Hypomagnesemia,
c. Hypocalcemia.

In people younger than 35 years of age, inherited diseases such as HCM, arrhythmogenic right ventricular cardiomyopathy/dysplasia, and congenital coronary artery abnormalities of wrong sinus origin are the most common causes of sudden death . In people older than 35 years, coronary artery disease is the most common cause of sudden death.

Hypertrophic Cardiomyopathy

HCM is the most cause of sudden cardiac arrest in young people.HCM is a genetically transmitted disease characterized by asymmetric LV hypertrophy with disproportionate septal thickening and reduction in LV chamber size. Decrease in LV compliance can contribute to increased wall stress and inadequate intramural coronary blood filling with exercise. Dynamic LV outflow tract obstruction at rest or with exercise is demonstrable in some patients. 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.[3]The characteristic histopathologic marker of HCM is myocardial disarray, with disorganized patterns of myocytes in association with increased interstitial fibrosis and often replacement scarring. Prevention of SCD-Those persons who are at high risk of SCD,should get ICD implantation done. Beta-blockers and amiodarone are being used for a long time, but there is no improvement in survival. These persons should be advised to avoid intense competitive sports, physical activity and alcohol use.

Arrhythmogenic right ventricular cardiomyopathy /dysplasia (ARVC)

ARVC is an inherited heart muscle disorder which is characterized by fibrofatty replacement of right ventricular myocardium. It is a genetically determined heart muscle disease[4,5]. It can be diagnosed by Echo which demonstrate right ventricular global or regional morphologic and functional abnormalities. ECG shows depolarization and repolarization abnormalities in right precordial leads. Commonly, premature ventricular contractions or sustained monomorphic ventricular tachycardia with left bundle morphology originate from the right ventricle and are associated with exercise and can cause syncope and in some case SCD. Symptomatic patients may need ICD.

Anomalous Origin Of Left Coronary Artery

It can precipitate sudden and unexpected cardiac arrest with exercise probably related to acute ischemia. Most commonly, the anatomic finding at the time of autopsy is the left main coronary artery arising from the right coronary sinus. The acute angle relative to the aorta taken by the anomalous coronary artery leaves a narrowed and compromised lumen, frequently characterized as “slit like,” which limits coronary blood flow and myocardial perfusion with exercise. Evidence of myocardial ischemia caused by anomalous origin of the coronary artery is not generally manifested on resting ECG and is rarely elicited with stress testing. Therefore, patient may have false-negative exercise stress tests. The diagnosis of anomalous origin of the coronary artery requires a high index of suspicion in young people presenting with exertional chest pain and/or syncope.[6] Once diagnosed, anomalous origins of the coronary arteries are generally treated with CABG.

Atherosclerotic Coronary Artery Disease

Because sudden cardiac death (SCD) in those people older than 35 years of age most commonly occurs because of atherosclerotic coronary artery disease, it is recommended that a careful history for coronary risk factors or symptoms be taken. If an person is identified as being at risk for coronary artery disease or if symptoms are suggestive of myocardial ischemia, an exercise stress test should be performed. Stress testing is also recommended in males older than 40 years of age or females older than 50 years of age in whom coronary artery disease is suspected based on the presence of at least two risk factors other than

age and sex or one marked abnormal finding.[7] If exercise stress test is positive, then detailed cardiovascular evaluation should be done and these patient should be put on anti-ischemic drugs and advised to avoid intense exertion and not to take part in competitive sports.

Myocarditis

Myocarditis either acute or healed, is also associated with sudden death in the presumptively by resulting in an inflammatory or fibrotic pathologic substrate predisposing to ventricular arrhythmias. Life-threatening ventricular arrhythmias can be associated with focal myocarditis that is clinically silent and not reliably detected by endomyocardial biopsy.

LONG QT SYNDROME

Approximately 10% of young athletes who die suddenly with exercise have no evidence of structural heart diseases. In many such patients, the cause of sudden death is likely a primary electrical heart disease. Prolonged QT means QTC is >480 ms. QTC prolongation more than 500 ms markedly increase the risk of SCD due to ventricular arrhythmia like VT/VF, torsades de pointes.

Types of long QT syndrome

Long QT syndrome type 1:
Reduced repolarizing current IKs due to mutation in KCNQ1 gene.
Long QT syndrome type 2:
Reduced repolarizing current IKr due to mutation in KCNH2 gene.
Long QT syndrome type 3:
Delayed inactivation of the INa due to mutations in SCN5A gene .

Others: Several other types of Long QT syndromes have been described; long QT types 1, 2, and 3 account for 80–90% of cases . 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.[8] Triggering factors to be avoided are : Acute physical stress, hypothermia, drugs which are known to prolong QT interval such as Terfenadine, Cisapride, Lithium, Qunidine, Sotalol, Amiodarone, Domperidon, Clarithromycin 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 insufficient.

Catacholaminergic Polymorphic Ventricular Tachycardia (CPVT)-CVPT accounts for 14-21%. Of SCD observed in young adults. [9]This rare familial syndrome is due to mutations in the cardiac ryanodine receptor and less commonly, the sarcoplasmic calcium binding protein, calsequestin 2. These mutations result in abnormal sarcoplasmic calcium handling and polymorphic ventricular arrhythmias. The VT is polymorphic or has a characteristic alternating QRS morphology termed bidirectional VT. Patients usually present during childhood with exercise or emotion induced palpitations, syncope, or cardiac arrest. Beta-adrenergic blockers (e.g., nadolol and propranolol) and an implantable defibrillator are usually recommended. Verapamil or flecainide or surgical left cardiac sympathetic denervation reduces or prevents recurrent VT in some patients.

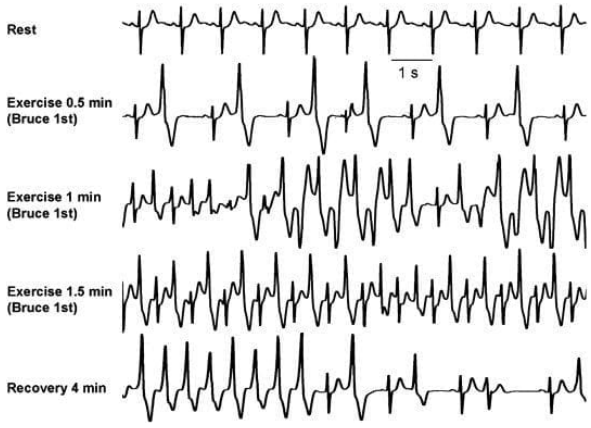

Figure1. Catacholaminergic Polymorphic Ventricular Tachycardia in a patient following exercise stress test.

BRUGADA SYNDROME

Brugada syndrome is a rare entit. Males are more commonly affected than females. Mutations involving cardiac sodium channels are identified in ~25% of cases. The characteristic ST-segment elevation can wax and wane over time and may become pronounced during acute illness and fever.

Diagnostic Criteria

Type1

- There is coved ST segment elevation >2mm in >1 of V1-V3 followed by a negative T wave.
- This is the only ECG abnormality that is potentially diagnostic and often referred to as Brugada sign..

Type2- Brugada Type2 has >2mm of saddleback shaped ST elevation.
Type3- Brugada type3: It can be the morphology of either type1 or type2, but with <2mm of ST segment elevation.

This ECG abnormality must be associated with one of the following clinical criteria to make the diagnosis of Brugada syndrome:

- Documented ventricular fibrillation (VF) or polymorphic ventricular tachycardia (VT).
- Family history of sudden cardiac death at the age of <45 years.
- Coved-type ECGs in other family members.
- Inducibility of VT with programmed electrical stimulation.
- Associated with syncope.
- Associated with nocturnal agonal respiration.

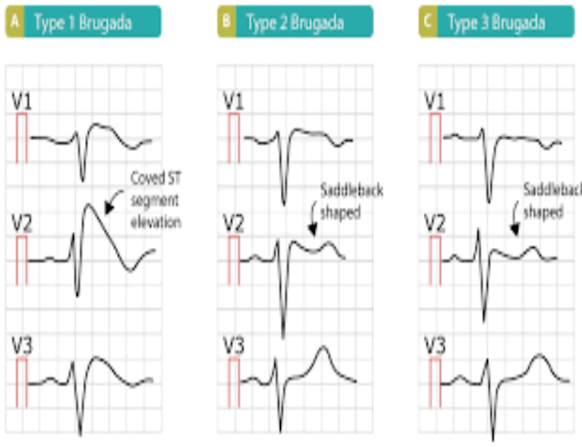

Figure2. Types of Brugada syndrome.

Administration of the sodium channel blocking drug flecainide, ajmaline, or procainamide can augment or unmask ST elevation in affected individuals. An ICD is indicated for individuals who have had unexplained syncope or been resuscitated from cardiac arrest. Quinidine and catheter ablation of abnormal regions in the epicardial right ventricular (RV) has been used successfully to suppress frequent episodes of VT.

ELECTROLYTE IMBALANCE

Hypokalemia, Hypomagnesemia, Hypocalcemia can also trigger SCD.

WARNING SIGNS OF SCD

1. Undiagnosed syncope
2. History of cardiac arrest
3. Family history of SCD
4. History of VT/VF
5. Warning sign of ECG
6. Echo evidence of disease
7. History of Angina on effort.

CONCLUSION

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. Those persons who present with warning signs of SCD 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.

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