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Congenital Heart Defect AndSudden Cardiac Death In Children And Adolescents

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Abstract: A congenital heart defect is a problem with the structure of the heart that a child is born with.Some congenital heart defects in children are simple and don't need treatment. Others are more complex. The child may need several surgeries done over a period of several years.

Index Terms -.Children, hypertrophic cardiomyopathy, long QT syndrome, sudden cardiac death.

I. INTRODUCTION

Serious congenital heart defects usually are found soon after birth or during the first few months of life. Symptoms could include:

1) Pale gray or blue lips, tongue, or fingernails. Depending on the skin color, these changes may be harder or easier to see.

2) Rapid breathing.

3) Swelling in the legs, belly or areas around the eyes.

4) Shortness of breath during feedings, leading to poor weight gain.

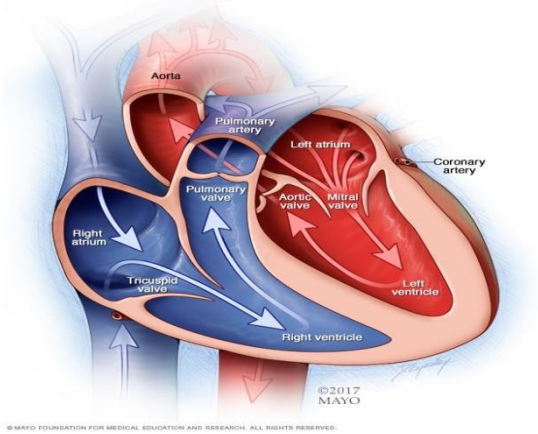
5) Less-serious congenital heart defects may not be found until later in childhood. Symptoms of congenital heart defects in older children may include:

6) Easily getting short of breath during exercise or activity.

7) Getting tired very easily during exercise or activity.

8) Fainting during exercise or activity.

9) Swelling in the hands, ankles or feet.



Sudden cardiac death (SCD) is defined as death that is abrupt, unexpected, and due to a cardiovascular cause.

ETIOLOGIES OF SCD IN THE YOUNG:-There are several known causes of sudden death in young people These etiologies can be divided into two categories:

1.arrhythmic

2.non-arrhythmic.

The majority of SCD in young people occurs due to arrhythmic causes. These usually result in an abrupt loss of consciousness, with or without a sensation of palpitations. The arrhythmia may be proven or presumed. Non-arrhythmic etiologies result in circulatory collapse in the setting of an organized rhythm. Examples of the latter include congestive heart failure, embolic phenomena, or aneurysm ruptures. A variety of identifiable conditions have been associated with SCD in the young.

Causes	Relative incidence (%)
Hypertrophic cardiomyopathy	36
Increased cardiac mass	10
Coronary artery anomalies	24
Marfan’s syndrome	6
Congenital heart disease	5
Myocarditis	3
Dilated cardiomyopathy	3
Arrhythmogenic right ventricular dysplasia	3
Ischemic heart disease	2
Commotiocordis	<1

HIGH RISK POPULATIONS

1.Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy (HCM) is the most common cause of SCD. The disease prevalence has been estimated as high as 1 per 500 in young adults. It is typically non-obstructive and presents in mid to late adolescence. HCM is often clinically silent, but the ECG typically may show left ventricular hypertrophy or T-wave abnormalities. The diagnosis traditionally has been best confirmed by echocardiography. However, carriers of an HCM genetic mutation may have little or no hypertrophy, especially earlier in life. Associated

sudden death is often exertional and is usually secondary to malignant ventricular arrhythmias. HCM is an autosomal dominant congenital disorder typically characterized by asymmetric septal hypertrophy and marked disarray of ventricular muscle fibers, which contribute to the risk of arrhythmias even in patients with minimal hypertrophy and no evident left ventricular outflow tract obstruction. HCM has been found to be caused by genetic abnormalities involving primarily sarcomeric contractile proteins (such as b-myosin and troponin T).

2.Dilated cardiomyopathy

Although less common than HCM, dilated cardiomyopathy (DCM) is also a known cardiac risk factor for SCD. DCM is characterized by cardiac dilation and decreased systolic function. It can be acquired from ischemic injury, myocarditis or toxins, or it can be inherited, usually as an autosomal dominant trait, with variable penetrance. There are currently more than 20 different gene mutations identified that cause DCM.

Coronary artery anomalies

In the United States, coronary artery anomalies are the second most common cause of SCD in the young. The most common abnormality associated with SCD is origin of the left main coronary ostia from the right sinus of Valsalva when the coronary artery traverses between the aorta and the pulmonary artery. Ischemia has been proposed to occur during exertion when the great vessels increase in size and compress the left main coronary artery. Although anomalous origin of the right coronary artery from the left sinus of Valsalva traversing between the aorta and pulmonary artery is far more commonit seems to have a much lower association with SCD than the left coronary artery originating from the right sinus. Coronary artery anomalies are unlikely to be identified without imaging studies unless complaints of early fatigue, angina, or exercise-induced syncope lead to a directed evaluation. Further, coronary anomalies may be difficult to screen for with a routine echocardiogram.

Arrhythmic channelopathies and primary arrhythmias

A variety of relatively rare conditions can cause primary arrhythmias in young people .Although there are cases where SCD is the first symptom with these conditions, recurrent syncope often precedes more malignant symptoms. Fortunately, the surface 12-lead ECG is abnormal in most of these conditions. So a minimal evaluation of an ECG and a careful history are indicated for recurrent syncope, and have been advocated as screening tests for athletes by many investigators and several organizations.

Primary arrhythmias and SCD

Arrhythmogenic right ventricular dysplasia

Long QT syndrome

Andersen–Tawil syndrome

Short QT syndrome

Brugada syndrome

Catecholaminergic polymorphic ventricular tachycardia

Wolff–Parkinson–White syndrome

Congenital complete heart block

Long QT Syndrome

The congenital form of the long QT syndrome (LQTS) is a familial genetic disorder that occurs in about 1 in 2500–3500 individuals. It manifests primarily as ventricular repolarization abnormalities caused by cardiac ion channel mutations. For symptomatic patients, the presenting symptom is usually syncope, which is due to the form of ventricular tachycardia known as "torsades de pointes." The syncope may occur with specific triggers, such as stress, swimming, and loud auditory stimuli, or it may occur when the child is relatively bradycardic, as during rest or sleep. In most cases, the corrected QT interval is prolonged, but there is considerable overlap with the normal distribution of QT intervals in the healthy population. In fact, 15–25% of patients with LQTS may have a QTc that falls within the "normal range." This has made clinical diagnosis difficult. In 1993, Peter Schwartz developed a set of diagnostic criteria that combined ECG and clinical criteria into a point system to aid in the diagnosis of LQTS.Family history of LQTS or sudden unexplained cardiac death are important factors in these criteria. However, points are also given for syncope, which occurs in up to 30% of the healthy population, and for QTc>450, which can be found in 2–5% of the healthy population. Further clinical evaluation of the QT interval during exercise or with epinephrine infusion can aid in the diagnosis, as QTc prolongation may become more exaggerated at increased heart rates as the QT interval fails to shorten in some patients. There are a variety of genetic abnormalities of ion channels which have now been identified, and it is estimated that up to 80% of patients with a high index of suspicion for LQTS have a mutation in one of the known LQTS genes.The specific phenotype may be predicted from the genetic mutation found and may aid in the assessment of risk for sudden death or response to therapy.Currently, the mainstay of therapy remains beta-blockade, which prevents severe symptoms and sudden death in most cases, but may be less effective for LQTS3. If symptoms recur on beta-blocker therapy, implantation of an ICD is generally considered indicated.

Catecholaminergic polymorphic ventricular tachycardia

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is characterized by ventricular ectopy induced by exercise or emotional stress. It is a genetic channelopathy, most commonly caused by mutations of the ryanodine receptor gene (*RYR2*) which encodes a sarcoplasmic calcium ion channel. The onset of CPVT symptoms typically occurs in childhood and adolescence. Exercise stress testing is important for diagnosis, since CPVT cannot be diagnosed on surface ECG. During exercise stress testing, ectopy is enhanced at greater levels of activity and often a "bidirectional" VT with a beat-to-beat 180° rotation of the QRS complex is observed. If left untreated, CPVT is lethal in 30–50% of patients. Although beta-blockers are the recommended therapy, many patients will continue to have arrhythmic symptoms and may need to have an ICD placed.

Brugada's syndrome:- Brugada's syndrome is an inherited arrhythmogenic syndrome characterized by specific ECG abnormalities and life-threatening ventricular arrhythmias.It commonly presents in men in the fourth decade as syncope and ventricular arrhythmias. The characteristic ECG findings are coved or saddle-back ST-segment elevation in leads V1–V3 with complete or incomplete right bundle branch block and T-wave inversion. The ECG abnormalities may not be evident until unmasked by an infusion of a sodium channel blocker (flecainide, ajmaline or procainamide). A handful of genetic mutations have been discovered to cause Brugada's syndrome; however, *SCN5A* is the most commonly occurring defect. Treatment is limited to ICD implantation.

Arrhythmogenic right ventricular dysplasia

Arrhythmogenic right ventricular dysplasia or cardiomyopathy (ARVD/ARVC) is a rare cause of SCD in the United States, but has been reported as the most common cause of SCD in the young athletes in Italy.Clinical presentation typically occurs in early adolescence to young adulthood. It is a heritable, progressive cardiomyopathy characterized by fatty and fibrous replacement of the myocardium, classically causing thinning of the right ventricular (RV) free wall. However, biventricular and left-dominant patterns are now being recognized.It is also characterized by electrical instability, and patients may present with a variety of ventricular arrhythmias, which are often provoked by exercise. The diagnosis is often difficult to make clinically. The RV changes are very hard to detect by echocardiography, but if suspicion is high, it can be detected by RV angiography and magnetic resonance imaging (MRI). The resting ECG may be mildly abnormal with ventricular premature beats with a left bundle morphology, T-wave inversion in leads V2 and V3, a so-called epsilon wave following the QRS in lead V1, and a wide QRS in leads V1–V3. Genetic testing may aid in the diagnosis, particularly if there is a strong family history. Although both drug therapy

and catheter ablation are occasionally successful, implantation of an ICD is usually recommended for patients with significant symptoms.

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