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SUDDEN LOSS OF CONSCIOUSNESS (SYNCOPE) AND SUDDEN DEATH IN CHILDREN:

The Emerging Importance of the Long QT Syndrome

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INTRODUCTION

Sudden and unexpected cardiac death in young persons is being recognized with increasing frequency. The sudden death of a young athlete during a sports competition is the most obvious and reported example, but death during non-competitive physical activity, during emotional upsets, or even during sleep are being recognized with progressively greater frequency. These events are usually due to a disturbance of the heart's electrical system, causing an abnormality of the heart beating or rhythm, called an arrhythmia. Fortunately, many of these deaths are preventable if the underlying conditions correctly recognized and properly treated. Unfortunately, many times the condition is unsuspected and, therefore, not treated, with a tragic sudden death the outcome. Particularly, a condition known as the **long QT syndrome** is being recognized as a common cause of the loss of consciousness and sudden death episodes, it is often not recognized prior to a sudden death.

WHAT CAUSES SUDDEN LOSS OF CONSCIOUSNESS?

The **arrhythmias** cause loss of consciousness by interfering with the blood pumping function of the heart with the consequence that the blood flow to the vital organs, particularly the brain, is diminished or absent. When the brain is deprived of blood flow for more than a few seconds, loss of consciousness occurs. This form of loss of consciousness is referred to as **syncope**. Syncope can also occur when the blood pressure falls dramatically, again resulting in lack of blood flow to the brain. The common faint, known medically as a **vasovagal** event, is primarily due to a fall in blood pressure usually accompanied by a slowing of the heart rate. Another common cause of loss of consciousness is a **seizure**. In seizure disorders, the blood flow to the brain and other organs is normal, but loss of consciousness occurs because of abnormal electrical discharges in the brain.

WHAT ARE THE COMMON CAUSES OF ARRHYTHMIAS AND CARDIAC SYNCOPE IN CHILDREN?

Almost any heart disease can cause arrhythmias and/or syncope. Anatomical (structural) heart disease such as congenital (present at birth) disorders, like a narrowed or leaking heart valve, a hole in the heart (septal defects), or more severe malformations, or rheumatic heart disease which affects the heart valves and muscle, are well known examples. Most of the time these conditions are not manifest by sudden loss of consciousness, but by shortness of breath or fatigue, or by physical exam findings detected by the physician.

Of more concern are inherited diseases, which principally affect the heart's electrical system, such as the long QT syndrome, and the heart muscle, such as hypertrophic cardiomyopathy. The long QT syndrome is particularly of concern because there are no physical abnormalities to detect, only a subtle electrical disturbance which is easily, and therefore, often overlooked.

WHEN SHOULD A LIFE THREATENING CARDIAC PROBLEM BE SUSPECTED?

Vasovagal episodes and seizures must be separated from cardiac arrhythmias, for while they are important they do not commonly lead to sudden cardiac death. A vasovagal event is most often precipitated by obvious factors such as pain, nausea, seeing blood, or other unpleasant stimuli. The individual becomes dizzy, warm and sweaty, has blurred vision, and usually has time to sit or lie down, which might lessen the symptoms or prevent the loss of consciousness. If a loss of consciousness does occur, it is very brief, and the person regains consciousness within a few seconds, being alert and oriented immediately. Seizures, on the other hand, usually have no obvious precipitating factors, have no preceding symptoms other than occasionally a strange feeling of various types and are associated with movement of the extremities such as stiffening of the muscles and/or body and then

shaking, and often with tongue biting and loss of urine or bowel control. The event may last around a few seconds to a minute or more. Following the event, the individual is often confused, disoriented and often lethargic for some time.

In contrast, the syncope of life threatening cardiac arrhythmia disorders is sudden, usually without warning, often occurs during or immediately following exercise, and is accompanied by gasping or absence of breathing, absence of a pulse, and in some cases by cyanosis or turning blue. Usually the loss of consciousness lasts from one of several minutes and in some cases requires resuscitation. These differences in presentation usually allow the conditions to be correctly separated, although some cases are confusing. Confusion often exists when a superficial rather than a comprehensive history of the event is obtained.

FAMILY HISTORY

Because these cardiac conditions are often inherited, a family history of syncope or unexplained sudden cardiac death in young persons may be present, and should be inquired about. The absence of a family history of these events, however, does not exclude these diseases.

DIAGNOSIS OF LONG QT SYNDROME

When a cardiac condition is suspected, medical evaluation is of utmost importance to prevent sudden cardiac death. A careful history and physical examination will eliminate many of other causes of syncope. The principle test required is the electrocardiogram (ECG, EKG), and specific observations for a prolonged QT interval and certain T-wave abnormalities. Please note, 10-40% of long QT syndrome patients have a normal QT interval on initial evaluation, so a normal QT interval *does not* exclude the condition. Diagnosis can be further clarified by an exercised EKG (stress test) and often by examining electrocardiograms on parents and siblings. In most cases, one parent is a gene carrier, therefore 50% of siblings would be at risk of inheriting the condition. Once an individual is diagnosed with long QT syndrome, it is extremely important that all members of the extended family be screened by EKG and medical histories gathered to identify others who are at risk of sudden cardiac death.

TREATMENT

Most of the cardiac conditions associated with unexpected sudden cardiac death are treatable, and events can be prevented or diminished. This is particularly true for long QT syndrome, as treatment is very effective in most instances by administration of daily medications.

HOW CAN YOU HELP?

If you observe children who have loss of consciousness, particularly during physical activity or emotional stress, discuss the situation with their parents or other responsible parties such as a school nurse who could then contact parents or other individuals responsible for the child. *A medical examination and careful evaluation may save the life of the child at risk!*

A child should be seen by a doctor if she/he has:

- Family history of unexpected, unexplained sudden death in a young person
- Fainting (syncope) or seizure during exercise, excitement or startle
- Consistent or unusual chest pain and/or shortness of breath during exercise