The Pediatric & Congenital Electrophysiology Society

Patient Resources: Arrhythmias and Congenital Heart Disease

Overview

Arrhythmias (abnormal heart rhythms) can develop in patients with congenital heart disease (CHD) due to thickening/weakening of their heart muscle, abnormal heart anatomy, abnormal circulation or scarring from prior surgeries. These arrhythmias include SVT, atrial flutter/atrial fibrillation, sinus node dysfunction, junctional rhythm, ventricular tachycardia, and various degrees of heart block (AV node block). Each will be described below and details regarding the normal conduction system of the heart can be found in the Basic Electrophysiology Fact Sheet.

Diagnosis of Arrhythmias

When there is a suspicion that a patient with CHD may be having arrhythmias your Cardiology/Electrophysiology team may order testing. Some of the tests that may be ordered are (see Basic Electrophysiology Fact Sheet for more details):

Electrocardiograms (ECGs)

An ECG, sometimes called an EKG, is a noninvasive test that measures the heart's electrical activity. For this test, small, sticky patches called electrodes are placed at specific locations on the skin. These electrodes measure the heart's electrical activity and are connected by wires to an ECG machine. The ECG machine then generates an image, called a tracing, to record the electrical activity measured by the electrodes.

Resting ECGs are done in clinics and hospitals and require a child to remove his or her shirt so that electrodes may be placed on their chest, arms, and legs. The ECG machine records the electrical activity for approximately one minute and produces a one-page tracing, usually showing only a few beats from each electrode to represent the overall activity measured during the test. Typically, a child lies down during a resting ECG.

Echocardiogram (or echo): an ultrasound of your child's heart to assess how well the heart is functioning (squeezing) and whether there are any structural abnormalties

Exercise ECGs, also called Exercise Stress Tests, are performed in exercise laboratories and require electrodes to be placed on the skin just like resting ECGs. However, instead of lying down, your child engages in physical activity, like walking on a treadmill or pedaling a stationary bike, with the ECG recording the entire time. This test is used to look for changes in the ECG that may occur as the result of exercise and the increased work being done by the heart. Your child may be asked to exercise until they are too tired to keep going or the test may be stopped if certain changes are seen on the ECG. Your child's ECG will also usually be monitored for a little while after the test while they are recovering from exercise.

Holter monitors are ECGs recorded over a period of 24 hours or more. Electrode stickers are

placed on the child's chest and connected with wires to small ECG recorder the child wears on his or her belt or carries in their backpack. They then go home and can engage in their regular activities (other than bathing, swimming, or activities that cause excessive sweating or may cause the leads to become loose or to fall off). With a continuous Holter, the device is worn for a specific duration, typically 24 or 48 hours, and then returned to clinic so the information can be analyzed. The Holter records every beat that a child has while wearing it, providing a lot of information for your healthcare team to review. It can be used in children who have suspected arrhythmias or in children who cannot feel or communicate their symptoms.

Event monitors are a special type of Holter monitor worn for up to 60 days. Some are programmed to automatically record arrhythmias that they detect on their own, and the recorder also has a button you or your child can press to record the rhythm when they have symptoms. As they are collected, these recordings are typically transmitted to a monitoring service on a regular schedule, which then reports them to your healthcare provider. Event monitors record every beat while they are worn, but only save "events" that are automatically triggered or patient activated. The limitation of event recorders is that the child must be able to feel his or her symptoms and be able to press the activator button or communicate their symptoms to an adult to activate the recorder.

Implantable loop recorders are small event monitors implanted under the skin that are battery-powered and can stay in place for 2-3 years. They may be placed with local anesthesia, conscious sedation, or general anesthesia depending on the age of the child. Like other event monitors, implantable loop recorders can be programmed to automatically detect arrhythmias and have a patient-activated recorder for symptomatic episodes. Implantable recorders communicate wirelessly with the recording-activating button (which needs to be with your child at all times) and with the special home monitoring system that transmits information about your child's rhythm back to your healthcare team. These transmissions can be scheduled automatically or can be initiated by families when a symptomatic event has been recorded. The information on the device can also be read in your electrophysiologist's office. Implantable loop recorders can be helpful for patients who have very infrequent symptoms that may indicate an arrhythmia.

Common Atrial arrhythmias in CHD

Atrial Flutter: The atria (upper chambers) of the heart beat faster than the ventricles (lower chambers). It is usually a regular rhythm caused by abnormal circuits in the electrical system.

Atrial flutter can sometimes change to atrial fibrillation.

Atrial Fibrillation: Disorganized, rapid electrical discharges in the atria, creating an irregular heartbeat in the top chambers (atria) that often does not coordinate with the heartbeat in the bottom chambers (ventricles).

These atrial arrhythmias are more common in congenital heart diseases that cause the top chambers of the heart to enlarge, such as atrial septal defects after surgical repair, Tetralogy of Fallot, classic and lateral tunnel Fontan circuits, and transposition of the great arteries after Senning/Mustard repair. Leaking of the valves that separate the atria from the ventricles can cause the atria to become large over time, and increase the risk of atrial arrhythmias.

Symptoms

The symptoms of atrial arrhythmias vary, ranging from no symptoms at all to shortness of breath, palpitations, dizziness and fatigue. If the atrial arrhythmia is not under control, weakening of the heart muscle and/or blood clot formation in the heart can occur, increasing the risk of stroke.

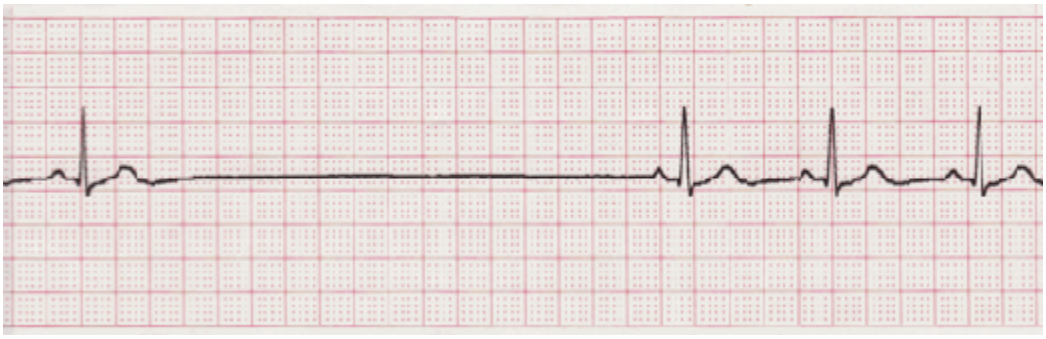
Treatment

Management of atrial flutter and/or fibrillation is directed at controlling fast heart rates that can lead to symptoms and overall weakening of the heart muscle, and avoiding blood clots that can lead to stroke. A variety of cardiac medications and blood thinners may also be prescribed by your cardiologist. On occasion, a cardioversion (a controlled electrical shock while under sedation) may be necessary to restore a patient's normal rhythm. In select patients either an electrophysiology study (EPS) and catheter ablation or surgical ablation (see Basic EP Fact

Sheet) can be used to treat the problem and destroy the sites of abnormal electrical discharges. Your cardiologist will discuss the treatment options with you.

Sinus Node Dysfunction (SND)

The sinus node (SA node) consists of a group of specialized cells within the right atrium and is the heart's natural pacemaker. Normally, the heart's electrical impulse starts in the SA node. This important structure allows the heart rate to increase or decrease in response to the body's activity level. Sinus node dysfunction can occur in CHD patients who have had cardiac surgery which results in scarring within the right atrium.



An ECG tracing showing a pause in the heart rhythm due to inactivity of the SA node.

Symptoms

Symptoms associated with SND vary from no symptoms to fatigue, bradycardia (slow heart rate), syncope (fainting), shortness of breath at rest or with exertion, and palpitations.

Treatment

Patients with significant symptoms often require and benefit from pacemaker implantation (see ICD/pacemaker overview) to increase their heart rate and have appropriate heart response to activity. If you might benefit from a pacemaker, your cardiologist will discuss the treatment options with you.

Junctional Rhythm

Junctional rhythm occurs when the electrical impulse in the heart starts in the atrio-ventricular node (AV node) instead of the SA node. Patients who have had heart surgery for CHD can develop junctional rhythm from damage to the AV node which causes it to fire automatically, "taking over" the SA node. Junctional rhythm can also occur if the SA node slows or is damaged from surgery. Junctional rhythms can often be normal, but sometimes they are too fast or too slow.

A junctional rhythm does not have a "P wave" that is associated with the impulse from the SA node.

Symptoms

Symptoms such as shortness of breath, fatigue, dizziness, palpitations, and syncope (fainting) may indicate that the function of the heart has decreased because the heart is not beating in a coordinated fashion or at an appropriate rate.

Treatment

CHD patients with junctional rhythms may require medical therapy or a pacemaker, or sometimes both. Your cardiologist will discuss the treatment options with you.

Ventricular Tachycardia (VT)

Ventricular tachycardia in CHD occurs because of poor heart function and/or scarring in the ventricles. See Ventricular Tachycardia page for more information.

AV Node Block

Various degrees of heart block in CHD occur because of damage to the AV node during surgical repairs. See AV Node Block page for more information.