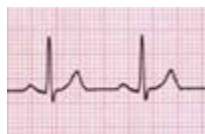


Patient Resources: Basic Electrophysiology Fact Sheet

What is electrophysiology?

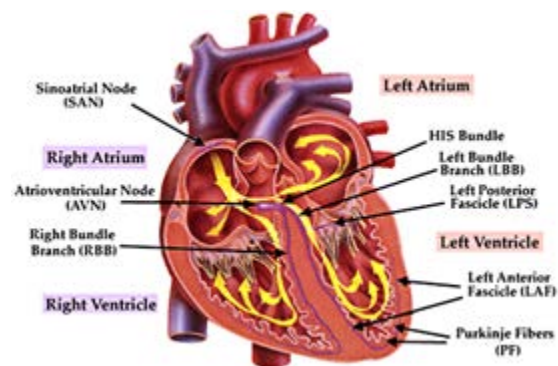


Electrophysiology (EP) is the study of the electrical system of the heart. This electrical system is what gives the heart its rhythm. Cardiologists with specialized training in how the heart's electrical system works are called electrophysiologists. If you think of the heart like a house, an electrophysiologist is the electrician.

What is the heart's "electrical system" and what does it do?

The heart is a muscular organ that is divided into four chambers: the left atrium and right atrium on top and the left ventricle and right ventricle on bottom. The heart's job is to pump blood through the body and in order to do this efficiently, four chambers need to squeeze and relax in an organized fashion. The heart coordinates this automatically through an electrical system that generates and transmits signals that tell the chambers when to squeeze. This keeps them from all squeezing or relaxing at the same time.

How does this electrical system make the heart beat?



The electrical system has groups of cells called nodes that can generate or receive signals and special tracts of cells that transmit, or conduct, signals. These tracts are also called conduction pathways. The signal for a heartbeat is first generated by a group of cells in the right atrium called the sinus node, also known as the sinoatrial node, SA node, or the heart's natural "pacemaker." The heart actually has multiple groups of cells that can function as a pacemaker, but the SA node is usually the fastest – and the best. An impulse that begins in the sinus node travels down a pathway to the atrioventricular node (AV node) located near the center of the heart, between the atria and the ventricles. The AV node delays the signal temporarily so that the atrial contraction can fill the ventricles with blood. The signal then continues down through the Bundle of His (pronounced "hiss"), which splits into left and right branches. These branches split further into smaller pathways called fascicles, to conduct the signal across both ventricles. This tells the ventricles to contract and pump blood out to the body. The next heartbeat begins with a brand new signal generated by the SA node.

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What is an arrhythmia?

An arrhythmia is an abnormal rhythm in the heart. In broadest terms, to say someone has an arrhythmia means that their heart is beating too fast (tachycardia), too slow (bradycardia), or in a way that does not follow the heart's regular pattern of electrical activity. Arrhythmias are often given names based on where they originate in the heart. For example, "supraventricular tachycardia" (SVT) is a fast ("tachy-") heart rate that originates above ("supra-") the ventricles, while "ventricular tachycardia" (VT) is a fast heart rate that originates in one of the ventricles itself. Also, there are even more precise terms that describe the specific mechanisms that cause SVT or VT in children.

What is a channelopathy?

The heart's electrical activity is generated by the flow of charged particles, called ions, into and out of the heart's cells through special channels. Sometimes these channels do not function like they are supposed to – they may allow too much (or too little) of a particular ion through. When this happens, a child is said to have a channelopathy. Channelopathies increase a child's risk for developing life-threatening arrhythmias. Examples of channelopathies include Long QT Syndrome (LQTS) and Brugada Syndrome.

What other kinds of patients are seen by electrophysiologists?

Electrophysiologists see more than patients who are already diagnosed with arrhythmias and channelopathies. They may also see patients with symptoms suggesting arrhythmias, such as palpitations, dizziness, or syncope (fainting) in order to diagnose or rule out certain conditions. They also see patients at risk for developing arrhythmias or channelopathies. This may include children with abnormal electrocardiograms (ECGs), children with a sibling, parent, or close relative diagnosed with a condition treated by an electrophysiologist, or children who have a strong family history of serious symptoms suggesting arrhythmias or channelopathies. Children with congenital heart disease are also at risk for developing arrhythmias for multiple reasons. First, since the physical structure of their heart is abnormal, this may introduce changes in the structures and pathways responsible for the heart's regular electrical activity. Second, surgery for congenital heart disease creates scar tissue in the heart, which may change existing pathways of electrical activity, or create new ones capable of causing arrhythmias. Electrophysiologists also see patients with heart block, a condition where the signals generated by the SA node do not reach the bottom chambers of the heart. Heart block may be congenital (a child is born with it) or acquired (a child suffers an injury to the heart's electrical system from surgery, infection, or some other cause).

What causes arrhythmias and channelopathies?

Arrhythmias occur when there are changes in the structure or timing of the heart's electrical system. One reason might be the SA node firing more quickly than it should or not firing fast enough to beat another one of the heart's other pacemakers. Also, the electrical system usually only conducts signals in one direction, but sometimes extra pathways exist that allow



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conduction to go other directions, which may cause tachyarrhythmias. Channelopathies are caused by a change in one or more of the proteins that make up the channels that let ions flow into and out of heart cells. An individual child's risk for an arrhythmia or channelopathy depends on a number of factors. While healthcare providers cannot perfectly predict when a child will develop one of these conditions, once a diagnosis has been made, that information is vital in determining the appropriate treatments to be offered.

How are patients with known or suspected electrophysiology problems evaluated by their healthcare providers?

There are many different tests and techniques that can be used by electrophysiologists evaluate their patients. Not every child needs every test, and the same test might be repeated multiple times with different results in the same child. Many of the tests are noninvasive, but some may require placement of an IV or other invasive techniques. Some of the tests your child may undergo are:

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 a tracing to record the electrical activity measured by the electrodes. There are many types of ECGs that may be ordered by your healthcare provider.



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. A resting ECG can be a good tool for

evaluating arrhythmias and channelopathies with known characteristic ECG changes, such as Wolff-Parkinson-White (WPW) syndrome or LQTS.

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 stress on 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 after the test while they are recovering

from exercise. This test is valuable in evaluating children for Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) and certain other arrhythmias.



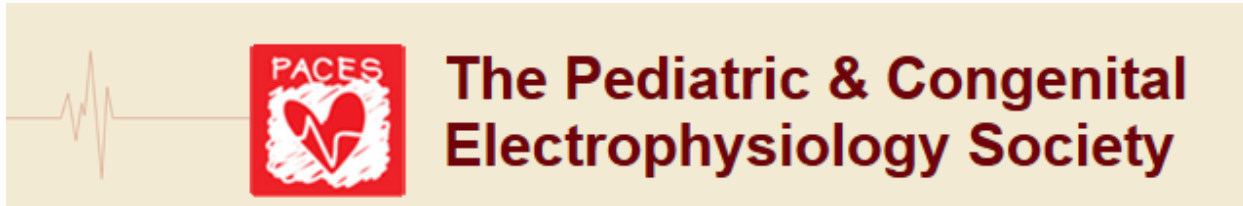
Holter monitors are ECGs recorded over a period of 24 hours or more. Three electrodes are placed on the child's chest and connected to small ECG recorder the child wears on his or her belt or carries in their backpack. They then go home and can engage in their regular activities (other than bathing, 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. They are programmed to automatically record arrhythmias that they detect on their own, but 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 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.

Tilt Table Test

A tilt table test is used to evaluate patients with frequent episodes of syncope (fainting). During this test, your child lies down on a table and is secured in place. The table then rotates your child from lying to standing while his or her ECG and blood pressure is monitored for



changes. The speed with which the table rotates and the time spent standing or lying down can vary depending on the testing protocol used by your healthcare team. Sometimes, medication or fluids may be administered through an IV to prevent or to treat syncope during the test.

Electrophysiology (EP) Study

An EP study is an invasive test that examines the electrical activity of your child's heart in detail. An EP study is almost always performed with the patient under general anesthesia. Long, soft, flexible wires called catheters are inserted into the large blood vessels in your child's groin and/or neck and advanced through these vessels into your child's heart. Sometimes, a special probe may be placed in your child's esophagus, which runs right next to the heart, to obtain additional information about their rhythm. These tools can sense the activity of the heart's electrical system to help determine where an arrhythmia is coming from in the heart. Once that information has been determined, your healthcare team will be able to better guide treatment for your child's arrhythmia going forward.

Genetic Testing

Genetic testing is an important tool in many aspects of healthcare today, but making the decision to have (or not to have) your child undergo genetic testing is a complicated process. Many arrhythmias and channelopathies have known genetic causes that can be tested for, but not every condition has an identifiable cause and not every patient with the same condition will have the same genetic testing results. While genetic testing is covered by many insurance plans, coverage is not universal and initial testing can be expensive based on the numbers of genes that must be examined. Waiting for results to come back, which may take months, can be stressful for families. When a genetic diagnosis is made, it can provide vital information for you, your child, their siblings, their other family members, and even their future children. A genetic diagnosis also allows for more focused testing in other family members, which may be less expensive than the initial tests that are required. Ultimately, the decision to pursue genetic testing is very individualized and requires open, direct communication among a patient, their family, and their healthcare providers.