

The heart's main job is to pump blood. The upper chambers (atria) take in blood from your lungs and body, and they pass that blood on to the lower chambers (ventricles) through your heart's first set of valves. The second set of valves then open and your ventricles push your blood out to your lungs and body. All of this action is coordinated by your heart's electrical system.

How is my heart wired?

To begin pumping, your heart muscle has to contract in a uniform way. Contraction starts when an electrical message goes out to each cell in your heart muscle. Your heart's electrical system has a very intricate network of connections that use special tissue to carry this message through the heart. Although it can operate on its own, your heart responds to many factors in your body. These factors include arousal, stress, fear, fever, anxiety, thirst, exercise, blood chemistry, and your overall wellbeing. There are a number of feedback systems between your heart, your brain, and other organ systems that help maintain a normal heart rate and rhythm.



Your heartbeat starts in the top of your heart in the sinus node. The sinus node sends the impulse that makes the top of your heart contract. The impulse then travels to the middle of the heart to the atrioventricular (AV) node. The AV node then sends the message to your ventricles. The ventricles contract and pump blood to the lungs and the body. The sinus node sets the pace for your heart. It is very responsive to factors like stress or fright. The sinus node also tells your heart to slow down when you are at rest. In an adult with a normal sinus node, the heart rate will range from 50 to 100 beats per

minute (bpm) during periods of relative inactivity. The heart rate can go as high as 170 to 200 bpm during exercise. During sound sleep, it can slow to the low- to mid-40s.

My heart is beating too slow; what does this mean?

In some congenital heart defects (CHDs), the sinus node is missing or does not work properly. It can also be damaged during heart surgery. This is especially common in operations that place sutures or patches in the sinus node area. These include the Fontan, Senning, or Mustard procedures. Sometimes tubing for the bypass machine can damage the sinus node or its blood supply. If this happens, your heart may beat slower (bradycardia). Regardless of the reason, if your sinus node is not working well for any reason, your adult congenital heart disease (ACHD) cardiologist may prescribe medications or recommend a pacemaker to help increase your heart rate.

The second major part of the electrical system in the heart is the AV node. The AV node is located in the middle of the heart between the upper and lower chambers. Its main job is to help manage the electrical activity as it travels from the atrium to the ventricles. The AV node slows down the electrical message slightly. This delay allows time for your heart's valves to open and let blood travel into the ventricles. This blood is then pushed into your lungs and body when the ventricles contract. The AV node can also independently fire and contract the lower chambers of the heart without an impulse from the sinus node. This allows the AV node to function as a "backup" pacemaker when there are problems with the sinus node.

Like the sinus node, the AV node can be abnormal in certain heart defects. These include congenitally corrected transposition of the great arteries (CCTGA) and some single ventricle defects. It can also be abnormal in babies born to mothers with lupus. Because the AV node is the center of the heart, it is very susceptible to surgical damage. Such damage can occur when holes in the heart are closed or when artificial valves are placed. Your AV node can also be damaged during surgery to correct narrowing under the aortic valve.

If your AV node is not working properly, you may develop heart block. There are three types of heart block. First degree heart block is when it takes too long for your heartbeat to travel from the top to the bottom of your heart. In second degree heart block, some of the electrical signals do not

reach the ventricles. Third degree (or complete) heart block is when the electrical impulse no longer travels through the AV node at all. Patients with advanced forms of heart block frequently require pacemaker therapy.

My heart is beating too fast; what does this mean?

So far, we have discussed problems with your sinus and AV node that can cause your heart to go too slowly (bradycardia). But congenital heart patients also can experience problems with the heart rate going too fast (tachycardia). Tachycardia can happen when there are problems with your heart's anatomy. It can also be caused by problems with your heart function such as heart failure or leaky heart valves. Sometimes surgical intervention can cause this kind of problem. If these rhythm issues come from the upper chambers of the heart, they are called atrial arrhythmias (abnormal heart rhythms). Problems that begin in the lower chambers of the heart are known as ventricular arrhythmias. Sometimes the heart develops new electrical pathways from top to bottom that bypass the AV node. This allows abnormal rhythms to develop called atrioventricular reentry arrhythmias.

Heart rhythm problems should always be treated in the context of the “whole picture” of the defect and heart history.

Medications are often used to treat rhythm problems. There are also pacemakers and/or implantable cardioverter-defibrillators (ICDs) that have been developed to treat abnormally fast heart rhythms. They can be programmed to detect abnormal heart rhythms from the upper or lower chambers of the heart. They then either pace the heart out of the fast rhythm or deliver an electrical shock to convert the rhythm back to normal. Patients with these special pacers usually continue on medications to help prevent rhythm problems from starting.

Will I be able to tell if my heart is beating too slow or too fast?

Whatever the cause, heart rhythm problems can cause a range of symptoms. Sometimes there are no symptoms. If you have a heart defect, it is important that you know how to tell if you might be having a heart rhythm problem. Occasional skipped beats are common in everyone with or without heart defects. They are usually more of a nuisance than a danger. But if you have continual skipped beats or

a racing heart, you should call your ACHD cardiologist immediately. You should also let him/her know right away if you feel dizzy or faint.

Heart rhythm problems can occur at any time including at rest and during exercise. They can be fatal, so it is very important that you do not ignore symptoms. In general, atrial rhythm problems are less dangerous than ventricular problems. All rhythm problems should be evaluated to help protect your heart function and health.

How are heart rhythm problems diagnosed?

Be sure to continue with recommended visits to your ACHD cardiologist so your heart's rhythm is checked regularly. If you report heart rhythm symptoms, your ACHD cardiologist will look for possible causes. He/she might recommend that you have some tests done. These might include an electrocardiogram (ECG), 24-hour Holter monitor, 30-day event monitoring, exercise testing, or an intracardiac electrophysiology study. If there is a question about whether a change in your heart function is causing the problem, an imaging test such as an echocardiogram or a cardiac MRI might be ordered.

Do I need special care for my special beats?

There are many cardiologists trained to take care of heart rhythm problems. If you do have a congenital heart defect, it is important that you visit an ACHD cardiologist. Although congenital heart patients get similar heart rhythm problems as everyone else, they often need different treatments. Rhythm problems can sometimes be caused by new problems related to the underlying defects and/or previous operations. Heart rhythm problems should always be treated in the context of the “whole picture” of the defect and heart history. By getting checked, you will help ensure that your ACHD cardiologist recommends a treatment plan that takes into account your unique congenital heart anatomy. You can find an online directory of adult congenital heart programs at www.achaheart.org, or by calling the Adult Congenital Heart Association at (888) 921-ACHA.

Although some people born with heart defects have rhythm problems as a child, many patients develop new rhythm problems as they age. Heart rhythm disorders are a common heart problem in adults living with heart defects. The good news is that there are now many new medications, devices, and other treatments to help control heart rhythm problems. By working as a team with your healthcare providers and reporting any new problems promptly, you can help protect your heart health.

For his work on this article, ACHA thanks **Larry A. Rhodes, MD**, a pediatric cardiologist with special training in the management of patients with abnormal rhythms. He now spends a significant portion of his time taking care of adults with congenital heart disease in a rural state. He is very interested in the ongoing care of adults with congenital heart disease, especially those who live in areas remote from major centers that offer care for these patients.