Acquired Heart Conditions

Arrhythmia

The heart's rhythm must be carefully coordinated to pump blood efficiently throughout the body. To accomplish this coordination normally, the heart relies on an elaborate electrical system that stimulates the muscular walls of the atria and ventricles to contract and relax in sequence.

The heart's electrical system is composed of the Sinus Node, the AV (atrioventricular) Node, the Bundle Branch Conduction Pathways, and most of the muscle cells of the atria and ventricles. Each heartbeat starts with an electrical impulse from the Sinus Node, which is the normal pacemaker of the body. The impulse then travels along the other parts of the electrical system to activate the atria and the ventricles, thereby completing a single heartbeat. Damage to any part of the heart's electrical system can result in an abnormally slow heart rate, called ***bradycardia***, or an abnormally fast heart rate, called ***tachycardia***.

Children can have serious heart rhythm problems before or immediately after birth, most often with heart rates that are too fast. Rhythm problems can be congenitally acquired, but may not become troublesome until much later. They can result from viral infections or can occur after heart surgery.

Bradycardia

Bradycardia, or slow heart rate, may be caused by an abnormally slow activity of the Sinus Node or conduction blockage somewhere within the electrical system, a condition called heart block. Heart block can occur as only a delay in conduction of the impulses from the atria to the ventricles, called first degree block, or intermittently blocked impulses, called second degree block, or completely blocked impulses, called third degree block. Some patients may be born with heart block (congenital block). Other patients may develop heart block because of a medication, abnormal blood chemistries, inflammation, or after surgery. Bradycardia may cause symptoms such as fatigue, poor exercise tolerance, dizziness, and syncope (loss of consciousness). Rarely, profound bradycardia may lead to death.

Pacemakers

Bradycardia and its associated symptoms may be improved or prevented with insertion of a permanent pacemaker. This mechanical device provides an electrical trigger for a heart beat when and where it is needed.

A typical pacemaker consists of one or two pacing leads that connect to a pulse generator, which contains a battery and circuitry, including a computer chip. Some pacemakers have special sensors that measure how active a patient is, allowing for more normal range of heart rates (e.g., slower rate at rest and faster rate with exercise).

Patients who receive a pacemaker with only one pacing lead have a single chamber pacemaker; those with two leads have a dual chamber pacemaker. Epicardial pacing leads are implanted on the outer surface of the heart and the pulse generator is placed in the abdominal region. This is often the approach of choice for infants and small children. In contrast, transvenous, or endocardial, pacing leads are commonly used in adults and older children. These leads are passed through one of the upper veins leading into the heart using X-ray guidance and the pulse generator is placed in the shoulder region.

Pacemakers are usually implanted under general anesthesia, and the patient remains in the hospital for one to several days. Patients are typically seen in the Outpatient Clinic soon after hospital discharge, and then every 6 to 12 months thereafter. In addition to these visits, periodic telephone follow-up is accomplished using a special device called a transmitter. In general, a patient with a pacemaker can participate in most routine physical activities and have few restrictions to daily living.

Tachycardia

Tachycardia, or fast heart rate, may result from abnormal electrical activity from electrical "short circuits" or abnormal activity of a small group of heart cells. This may occur in the absence of obvious heart disease, or may be associated with heart disease such as those who have undergone cardiac surgery. Symptoms of tachycardia

include palpitations, dizziness, shortness of breath, and syncope (fainting). Serious forms of tachycardia can lead to sudden death. Depending upon clinical circumstances, tachycardia can be treated with medications, electrical therapy, radio-frequency ablation using catheters, or heart surgery.

Medications

A variety of medications are used to treat patients with tachycardia. No medicine can eliminate or permanently change the tissues of the heart causing the tachycardia, but many medicines may effectively stop tachycardia or prevent its recurrence. Although "antiarrhythmic" medications are generally well tolerated, most can cause side effects. Medications most commonly used in pediatric arrhythmia management include:

Adenosine is a medicine commonly used in the Emergency Room to abruptly stop tachycardia due to supraventricular tachycardia. Adenosine can only be given using an IV, and is usually very effective. Adenosine lasts only 10 seconds in the bloodstream, so it cannot be used as a long-term medicine to prevent recurrent tachycardia.

Digoxin is one of the most widely prescribed medications in children with heart disease. It is most often used to increase the squeezing strength (contractile force) of each heartbeat. Digoxin also slows down the heart rate during some types of tachycardia by increasing the activity of the parasympathetic nervous system. However, Digoxin is not a powerful way to treat arrhythmias and may be combined with other antiarrhythmic medications. Although generally well tolerated, possible side effects include nausea, vomiting, visual changes, and tiredness, especially if too much medicine is ingested. Some other medications alter the way Digoxin is absorbed in the body, so dosage may need to be adjusted.

Beta Blockers, such as Propanolol (Inderal), Atenolol (Tenormin), and Metoprolol (Lopressor) are common medicines used to prevent tachycardia. These drugs block the sympathetic nervous system and slow activity in

the "beta receptors" in the heart, causing abnormal heart rate to slow down. The side effects of beta-blockers include bradycardia, hypotension, fatigue, and mood swings. Patients with asthma may have problems with breathing. Small children must eat or drink regularly, as beta-blockers may affect blood sugar levels. This is especially important in young children when they are sick and have a poor appetite.

Amiodarone (Cordarone) and **Sotalol** (Betapace) are medicines that are used when others have failed to treat tachycardia. These drugs have many actions, including the blockage of potassium, calcium channels and beta receptors in the heart. It is recommended that these medications be initiated in the hospital where the heart rhythm can be continuously monitored because of the potential for serious side effects, most notably serious arrhythmias. The medications may also require periodic blood tests to assess effects on other organs of the body. Some side effects seen with these medications are bradycardia, heart block, arrhythmias, skin sensitivity to the sun, vision disturbances, thyroid disorders, changes in lung function, liver or blood changes.

Electrical Therapy

When tachycardia is dangerous and cannot be controlled by medication, immediate electrical therapy may be needed to terminate the fast rhythm. Electrical therapy involves delivering electrical impulses directly to the atrial or ventricular tissue to stop tachycardia. These impulses can be delivered in several ways:

•A transesophageal lead or wire can be inserted through the nose and placed in the esophagus, which lies immediately behind the heart. This lead can be used to deliver electrical impulses to the heart, often stopping tachycardia. Mild sedation is often necessary to help the patient relax and decrease discomfort during this form of treatment. Occasionally, a transesophageal pacing study is used to determine which types of tachycardia a patient may have, or to evaluate the effectiveness of medication in prevent-

ing future episodes of tachycardia.

•Implantable antitachycardia devices are similar to pacemakers, but can also automatically deliver electrical therapy to stop tachycardia abruptly. These devices constantly monitor the patient's heart rhythm, and terminate tachycardia soon after it begins. Antitachycardia devices can decrease the need for medications and reduce emergency room visits for episodes of tachycardia. Most patients do not feel the therapies delivered by antitachycardia pacemakers. These devices may also prevent the heart from going too slow.

•Implantable cardioverter-defibrillators (ICDs) are another type of automatic device most commonly used to treat very dangerous forms of tachycardia, such as ventricular tachycardia. Most ICDs are implanted using the transvenous approach. Cardiac rhythms are constantly monitored and when a ventricular tachycardia occurs, the device can deliver a shock to the heart muscle. Most patients feel some discomfort during the therapy, but sudden death may occur without the shock. Patients with ICDs are followed in the Outpatient Clinic every 3 to 4 months. Patients generally have few restrictions to daily living, and can participate in many less strenuous physical activities, except as limited by underlying heart disease.

RF Catheter Ablation

This procedure is used to eliminate the source of the problem in some types of tachycardia. The cardiologist performs this procedure in the cardiac catheterization lab, usually with the patient under general anesthesia. Catheters or wires are placed into the patient's heart by way of large veins in the legs and neck. The cardiologist then "maps" the heart's electrical system to locate the electrical short-circuit or over-active cells. Once found, this small amount of abnormal tissue can be ablated, or destroyed, by direct heating of the tip of a special catheter using radio-frequency current. Preventing the tissue area from causing future episodes of tachycardia eliminates the need for medicines or other treatments.

Cardiomyopathy

Cardiomyopathy is a disease of the heart muscle, often without an identifiable cause. It can be associated with skeletal muscle diseases, such as Friedreich's ataxia, muscular dystrophy, or glycogen storage disease, or with viral infections. It is usually chronic and can result in dilated cardiomyopathy, which is a weakened heart muscle, or restrictive cardiomyopathy, which is a stiff heart muscle. Hypertrophic cardiomyopathy (also known as idiopathic hypertrophic subaortic stenosis, or IHSS, or as asymmetric septal hypertrophy or ASH) can occur in families. With these conditions, the left ventricular muscle, especially the septal wall separating the right and the left ventricles, becomes extremely thickened and can obstruct the exit of blood into the aorta. The thickened muscle is prone to heart rhythm irregularities.

Congestive Heart Failure

The heart has two pumping systems – the right side, which pumps blood to the lungs, and the left side, which pumps blood to the body. Congestive heart failure develops when the pumping action of the heart deteriorates to the point that the heart is unable to pump enough blood to meet the body's needs. This may occur as a result of an excessive workload on the heart muscle.

One way the heart responds to an excessive workload is by beating faster to meet the body's demand for blood and oxygen. While this may have a short-term benefit, over time the heart can become enlarged, causing the muscle walls of the heart to grow weak and inefficient. This can reduce blood volume in the body, which causes the body's arteries to constrict, forcing the heart to work even harder to pump blood. Eventually, blood will back up into the lungs and in other areas of the body, such as the liver and extremities. When blood backs up, it isn't delivered properly throughout the body, which places further demand on the heart to beat faster, which leads to further deterioration.

Congestive Heart Failure in Children

Congenital heart defects are the most common cause for heart failure in infancy and childhood. These include: hypoplastic left heart syndrome, coarctation of the aorta,

ventricular septal defects, patent ductus arteriosus, atrioventricular canal, total anomalous pulmonary venous connections, transposition of the great arteries and truncus arteriosus. Primary myocardial diseases, such as myocarditis or cardiomyopathies, may evoke heart failure at any time. In addition, heart failure may occur after open-heart repair of congenital heart defects. Other heart conditions that are acquired may lead to congestive heart failure after one year of age. These include: cardiomyopathies, rheumatic heart disease and severe cardiac dysrrhythmias. High blood pressure from any cause may also lead to cardiac failure due to prolonged resistance in the blood vessels.

Signs and Symptoms

Signs of heart failure will differ depending on the patient's age. In infants, a fast heart rate (greater than 160 beats per minute), difficulty breathing and/or sweating during feeding can be the first signs of heart failure. As the disease progresses, the infant may not grow. When severe, congestive heart failure causes the infant to have fast and labored breathing, sometimes with grunting.

In older children and adults, heart failure is recognized by limitations in physical activity. In mild failure, the patient is comfortable at rest, but moderate physical activity causes fatigue, shortness of breath and occasionally palpitations and chest pain. As symptoms progress, minimal physical activity causes fatigue and other symptoms. Finally, in its most severe stage, heart failure causes the patient to be unable to carry on daily activities without symptoms or discomfort, and shortness of breath or chest pain may occur even at rest.

Treatment

The goal of treatment is to reduce the workload of the heart by controlling excess salt and water, thereby improving the heart's ability to pump. Medications also can be very effective in working toward that goal. These medications include: ACE inhibitors, such as Captopril, to open the arteries and veins so more blood can reach the body's tissues; Diuretics, such as Lasix, to act on the kidneys to rid the body of extra salt and water, reducing the accumulation of fluid in the lungs to lower blood pressure and improve the efficiency of blood circulation; and Digoxin, which helps

treat congestive heart failure by increasing the strength of the heart's contractions.

New research studies are looking at medications to block the sympathetic activity of the heart, that part of the nervous system that causes an increase in heart rate and blood pressure. Beta blockers, such as Carvedilol, block the activity of the sympathetic nervous system and may be effective in slowing the progression of congestive heart failure.

Endocarditis

Endocarditis is a disease of the inner lining of the heart. It can be associated with generalized systemic inflammatory diseases, such as lupus erythematosus or Liebman-Sachs, and results in thickened and distorted heart valves. Rheumatic fever may cause similar valve damage. Infectious endocarditis is caused by bacterial bloodstream infections. It can develop rapidly, an acute condition, or slowly, a sub-acute condition. Children with certain congenital or acquired heart disease are sensitive to this infection. (For more information on Bacterial Endocarditis, see chapter on Family Living.)

Pericarditis

Pericarditis is an inflammatory disease of the outer lining of the heart, which normally has a sack-like double layer with a small amount of fluid inside for lubrication. When inflamed, the amount of fluid increases and can compress the heart to the point of malfunction. When infected, pus can accumulate in the pericardial sac, known as purulent pericarditis. Occasionally, there is not enough fluid between the layers and the smooth inner surfaces become rough, causing pain with each heartbeat, a condition known as fibrinous pericarditis.

Rheumatic Heart Disease

Rheumatic heart disease is caused by ***Streptococcus bacterium***, usually associated with a sore throat, commonly known as "strep throat." It starts as rheumatic fever and can involve the joints, the kidney, and the heart. It can affect the heart muscle, a condition known as myocarditis; the mitral and/or aortic valves, a condition called valvulitis; or the outer membrane of the heart, a condition known as pericarditis.

When the acute insult is over, it can leave behind leaky regurgitant valves. These conditions can be prevented by early treatment of streptococcal throat infections. Once a child develops heart disease from streptococcal infection, he or she must remain on antibiotics for life to prevent recurrence that can further damage the heart valves.

Viral Myocarditis

Viral myocarditis can be caused by a variety of viruses. Occasionally, it may damage the heart muscle permanently, a condition known as cardiomyopathy.