Heart Conditions

Mitral Valve Prolapse

Mitral Valve Prolapse - is a condition in which the two valve flaps of the mitral valve do not close smoothly or evenly. Mitral valve prolapse is also known as click-murmur syndrome, Barlow's syndrome or floppy valve syndrome.

Symptoms, which in some cases may be related to leaking of the mitral valve, can include:

- Chest pain (not caused by coronary artery disease or a heart attack)
- Dizziness.
- Fatigue.
- Sensation of feeling the heart beat (palpitations)
- Shortness of breath with activity or when lying flat (orthopnea)

Mitral valve prolapse occurs in 2 percent to 3 percent of the population. A person can be born with the genetic risk of developing MVP or it can be caused by other health problems, such as some connective tissue diseases.

How Is Mitral Valve Prolapse Diagnosed?

Mitral valve prolapse (MVP) most often is detected during a routine physical exam. During the exam, your doctor will listen to your heart with a stethoscope.

Stretched valve flaps can make a clicking sound as they shut. If the mitral valve is leaking blood back into the left atrium, your doctor may heart a heart murmur or whooshing sound.

However, these abnormal heart sounds may come and go. Your doctor may not hear them at the time of an exam, even if you have MVP. Thus, you also may have tests and procedures to diagnose MVP.

Echocardiography (echo) is the most useful test for diagnosing MVP. This painless test uses sound waves to create a moving picture of your heart.

Long QT Syndrome

Long QT Syndrome (**LQTS**) - is a rare congenital and inherited or acquired heart condition in which delayed repolarization of the heart following a heartbeat increases the risk of episodes of *torsades de pointes* (TdP, a form of irregular heartbeat that originates from the ventricles). These episodes may lead to fainting and sudden death due to ventricular fibrillation. Episodes may be provoked by various stimuli, depending on the subtype of the condition.

Many people with long QT syndrome have no signs or symptoms.

Some people may experience the following symptoms:

- Fainting (or syncope). This may occur when the patient is emotionally or physically stressed. It is unusual in QT syndrome to have any signs before the person actually faints.
- Seizures
- Sudden death. If there is sudden death, and doctors suspect long QT syndrome as the cause, they may recommend that the family members of the deceased get tested for the disease.

Risk factors for long QT syndrome include the following:

- female sex
- increasing age
- liver or renal impairment
- family history of congenital long QT syndrome
- pre-existing cardiovascular disease
- electrolyte imbalance: especially hypokalemia, hypocalcemia, hypomagnesemia
- concurrent administration of interacting drugs

Anorexia nervosa has been associated with sudden death, possibly due to QT prolongation. It can lead a person to have dangerous electrolyte imbalances, leading to acquired long QT syndrome and can in turn result in sudden cardiac death. This can develop over a prolonged period of time, and the risk is further heightened when feeding resumes after a period of abstaining from consumption. Care must be taken under such circumstances to avoid complications of refeeding syndrome.[5]

Those diagnosed with LQTS are usually advised to avoid drugs that would prolong the QT interval further or lower the threshold for TDP. In addition to this, two intervention options are known for individuals with LQTS: arrhythmia prevention and arrhythmia termination.

Arrhythmia suppression involves the use of medications or surgical procedures that attack the underlying cause of the arrhythmias associated with LQTS. Since the cause of arrhythmias in LQTS is EADs, and they are increased in states of adrenergic stimulation, steps can be taken to blunt adrenergic stimulation in these individuals. These include administration of beta receptor blocking agents, which decreases the risk of stress-induced arrhythmias. Beta blockers are an effective treatment for LQTS caused by LQT1 and LQT2.[2]

Arrhythmia termination involves stopping a life-threatening arrhythmia once it has already occurred. One effective form of arrhythmia termination in individuals with LQTS is placement of an implantable cardioverter-defibrillator (ICD). Also, external defibrillation can be used to restore sinus rhythm. ICDs are commonly used in patients with fainting episodes despite beta blocker therapy, and in patients having experienced a cardiac arrest.

Marfan Syndrome

Marfan syndrome is a rare, inherited disorder of connective tissue and growth. The disease affects several parts of the body.

The disease is highly variable, which means that different people with the condition will have different parts of their body affected to a greater or lesser degree.

The features of Marfan syndrome result from changes in the body's connective tissue and in the control of body growth. Connective tissue holds our bodies together. It is found throughout the body, which is why it can affect many different parts of the body.

Diagnosis

There is not one simple test to make a diagnosis for Marfan syndrome. To find out if someone has Marfan syndrome, the patient needs to be checked for features of the condition. This may mean seeing a number of doctors and undergoing various special tests. The Marfan Clinic staff will determine which special evaluations or doctors a patient needs to see and will coordinate these evaluations. The genetics evaluation in the Marfan Clinic brings together the information from all these different doctors and tests to determine if the person has Marfan syndrome.

To be diagnosed with Marfan syndrome a person must either have several of the features or have just a few of the features but also have a family member with documented Marfan syndrome. Genetic testing can also help with the diagnosis. This is a blood test that looks for a mutation in the FBN1 gene, which is the cause of Marfan syndrome.

If the FBN1 mutation causing Marfan syndrome in a family is known, then other members of the family can be tested for that mutation. This will determine if they have inherited Marfan syndrome.

Treatment:

Care of people with Marfan syndrome has significantly improved over the years. Most people will live a normal life span and have a good quality of life, though they will need more medical attention and treatment than the average person. Because Marfan syndrome affects several different parts of the body, people with Marfan syndrome need to see doctors who specialize in genetics, cardiology, orthopedics, ophthalmology and other specialties as needed.

Most of the features of Marfan syndrome can be treated. As mentioned above, aortic dilation can be managed with medications, regular screening of the aorta by echocardiogram, and surgery to prevent aortic dissection. The eye features can be treated with glasses or, if needed, with surgery.

Hypertrophic Cardiomyopothy

Hypertrophic Cardiomyopothy – in this abnormality, the muscle wall of the left ventricle including muscle that separates the right and left ventricles (ventricular septum), is much thicker than normal. In some cases, the ventricular septum may be as much as 4 times the normal thickness. This is a rare abnormality that is often genetic in origin and may run in families. Girls and boys are equally affected. The thickened ventricular septum may obstruct the outlets to the aorta and/or pulmonary artery (PA), elevating left ventricular pressure and if severe, reducing coronary perfusion. However, there is no obstruction and therefore no murmur in the majority of cases. fainting spells, arrhythmia (irregular heart beat), heart failure, or even death. Patients with severe hypertrophic cardiomyopathy are at risk of dying suddenly from abnormal heart rhythms.

Symptoms - Many patients experience no symptoms except during periods of exertion, when chest pain and shortness of breath may occur. Hypertrophic Cardiomyopathy (HCM) may also cause.

Diagnosis - HCM may be readily diagnosed through a variety of tests, including electrocardiography (producing an ECG, or electrocardiogram) and echocardiography (producing an echocardiogram). The ECG will show abnormalities in 80-90% of HCM patients. About 25% of patients will develop arrhythmias (e.g. ventricular tachycardia, atrial fibrillation). A heart murmur may be present, characterized by changes in its nature depending on the position of the patient. Typically, the murmur will decrease in intensity as the patient goes from a standing to a squatting posture, and vice versa.

Treatments - There are various treatments for hypertrophic cardiomyopathy. These include medications such as beta blockers and calcium channel blockers, which slow the heart rate and minimize the symptoms. Children are restricted from vigorous exercise. For those patients with more serious symptoms or disease progression, hypertrophic cardiomyopathy may be treated with a pacemaker to optimize heart function. Another option to relieve obstruction is the surgical reduction (septal myectomy) of the ventricular septum during open heart surgery (see animation at left). Treatments to ward off sudden death involve medication with amiodarone and the implantation of ICDs (implantable cardioverter defibrillators, which will shock the heart to restore normal rhythm if necessary), both of which protect against fatal arrhythmias.

Bicuspid Aortic Valve

The aortic valve conducts oxygen-rich blood from the left ventricle into the aorta, which carries it to the body tissues. Normally, this valve has three leaflets. Occasionally, people are born with an aortic valve that has only two leaflets. This is known as a bicuspid aortic valve. A bicuspid aortic valve may occur in isolation in an otherwise normal heart, or it may occur with other heart defects. Isolated biscupid aortic valve is the most common of all congenital cardiac anomalies and usually has no effects on heart function. If a bicuspid aortic valve does not open normally, aortic valve stenosis can result. The aortic valve is generally bicuspid in cases of Aortic Stenosis, which accounts for 3-6% of all cases of congenital heart disease.

What Are the Effects of Bicuspid Aortic Valve?

Though bicuspid aortic valves usually cause no adverse symptoms, difficulties may arise as the patient grows older. For example, the valve opening may become narrowed (stenotic) because of calcification of the valve leaflets. Also, the valve might not open or close completely, resulting in leakage at the valve opening (regurgitation). Occasionally, patients with a bicuspid aortic valve will experience a progressive enlargement (dilatation) of the aorta. This can result in the formation of an ascending aorta aneurysm and, rarely, dissection (splitting open) of the aorta. Dissection is an up and down tear in the aortic wall with blood filling the inner and outer layers of the aorta.

How Is Bicuspid Aortic Valve Treated?

In most cases, a bicuspid aortic valve does not require any treatment in childhood. A pediatric cardiologist will make specific recommendations for activity restrictions for each patient if valve narrowing or leakage occurs. In general, no other special precautions are required other than regular follow up with a qualified cardiologist. To evaluate the bicuspid aortic valve, the cardiologist follows the patient every 1-5 years with non-invasive tests that might include an electrocardiogram (ECG), echocardiogram, and stress test. In cases where the narrowing of the aortic valve becomes severe, a balloon valvuloplasty or valve replacement operation (e.g. Ross, Ross-Konno Procedure, or prosthetic valve replacement) may become necessary.

Wolff-Parkinson-White Syndrome (WPW)

Wolff-Parkinson-White syndrome (WPW) is a type of abnormal heartbeat. If you have WPW, you may have episodes of tachycardia, when your heart beats very rapidly. WPW affects between one and three of every 1,000 people worldwide. Electrical signals going through your heart in an organized way control your heartbeat. This allows blood to pass from the upper chambers of your heart (the atria) to the lower chambers of your heart (the ventricles), and to then travel throughout your body. Normally, a structure in your heart called the sinoatrial node regulates how electricity passes from the upper chambers of your heart to the lower chambers. The sinoatrial node keeps your heartbeat at about 60 to 100 beats a minute. When you have WPW, you are born with an extra pathway that allows electrical signals to bypass the sinoatrial node. This can result in a very rapid heart rate — 200 beats per minute or more.

Symptoms

With WPW, you may not have any episodes of tachycardia for many years. Symptoms may also start and stop suddenly and occur at any age. Typical symptoms include:

- Shortness of breath
- A pounding in your chest
- Dizziness
- Passing out

WPW affects both men and women. In most cases, the cause of WPW isn't known, but doctors have identified mutations in a gene that may be responsible for WPW. A small number of people may be at risk because they inherited this gene from a parent. WPW is a common cause of tachycardia in China, and you may be at increased risk if you are of Chinese descent.

Diagnosis

If you have symptoms of tachycardia that come and go, your doctor will do a test called an electrocardiogram, or ECG. An ECG measures the electrical activity in your heart and your heart rate. If you are not having symptoms at the time of your ECG, results may look normal. Other tests may include:

- Conducting an ECG as you walk on a treadmill
- Wearing a type of recorder, called a Holter monitor, that takes an ECG over 24 hours
- Wearing a type of recorder, called an event recorder, that samples your heart rate over several days
- Electrophysiologic testing, a hospital procedure that involves threading catheters into your heart through a vein in your thigh

Treatment

You may not need any treatment if you do not have symptoms, or have infrequent symptoms. Also, symptoms sometimes go away as people get older. If you do need treatment, there are a number of options:

- You may be able to stop an episode of tachycardia by massaging your neck, coughing, or bearing down like you are having a bowel movement. This is called a Valsalva maneuver.
- You may be able to take medication to stop or prevent tachycardia.
- If medication and the Valsalva maneuver do not work, you may need to go to the hospital for cardioversion. This procedure restores your heartbeat to a normal rhythm, by passing an electric current through your chest into your heart.
- If you are having frequent or uncontrolled episodes of tachycardia, you may have a surgical procedure called **radiofrequency ablation**. Low-voltage, high-frequency electrical energy interrupts the extra pathway in your heart. Your doctor threads a catheter into your heart through a vein in your thigh. The treatment cures WPW about 95 percent of the time.

Link to addition heart conditionings.

https://www.hopkinsmedicine.org/heart_vascular_institute/conditions_treatments/conditions/pediatric_congenital_heart_disease.html

https://www.nicklauschildrens.org/medical-services/pediatric-cardiology

http://my.clevelandclinic.org/childrens-hospital/health-info/diseases-conditions/hic-pediatric-congenital-heart-defects